



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

### Poster Presentations

Tuesday, February 24, 2015 • 6:00 p.m. – 9:30 p.m.

*Authors will be standing by their posters during the following hours:*

*Odd-Numbered Posters: 6:45 p.m. – 7:30 p.m.*

*Even-Numbered Posters: 7:30 p.m. – 8:15 p.m.*

Poster #		Presenting Author
<b>Category: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm)</b>		
1	Normal Tension Glaucoma (NTG) is it a Type of Glaucoma or an Entity of a Compressive Optic Neuropathy (CON)?	Mohammed Areesh
2	Anterior Ischemic Optic Neuropathy Due to Biopsy-Proven Giant Cell Arteritis in Thai Patients	Taweevat Attaseth
3	Visual Outcomes Following Perioperative Vision Loss	Jasmina Bajric
4	Pituitary Apoplexy in Pregnancy	Matthew J. Benage
5	Bilateral, Sequential Anterior then Posterior Ischemic Optic Neuropathy in a Young Migraineur	Damian E. Berezovsky
6	Traumatic Optic Neuropathy. Our Experience	Mariana de Virgiliis
7	Sensitivity of Magnetic Resonance Imaging in Acute Demyelinating Optic Neuritis	Lulu LCD Bursztyn
8	Characterization of Leber's Hereditary Optic Neuropathy Patients Treated with Idebenone	Jasdeep S. Chahal
9	Gliosarcoma of the Optic Nerves 15 years After Radiation Treatment for Hypophyseal Adenoma	Ayse I. Colpak
10	Optic Nerve Compression by an Anomalous Internal Carotid Artery	Valerie I. Elmalem
11	Identifying Risk Factors for the Development of Radiation Optic Neuropathy at "Safe" Doses: A Review of Cases Seen 1994-2014	Pavle Doroslovački
12	Optic Neuritis After Refractive Surgery: Causal or Coincidence?	Shlomo Dotan
13	Long Term Treatment of Lebers Hereditary Optic Neuropathy with Idebenone	Jennifer I. Doyle
14	Macular Star Formation in Diabetic Patients with Non-Arteritic Anterior Ischemic Optic Neuropathy (NA-AION)	Alberto Galvez-Ruiz
15	Seasonal Influence on the Incidence of Biopsy-Proven Giant Cell Arteritis: The University of California Davis Institutional Experience	Kimberly K. Gokoffski
16	Possible Revatio (Sildenafil) Induced Optic Neuropathy in Mice	Nitza Goldenberg-Cohen
17	Cyclosporine Induced Papilledema without Elevated Intracranial Pressure	Poorav J. Patel
18	Asymptomatic Leukemic Optic Nerve Infiltration as Presentation of Acute Lymphoblastic Leukemia Relapse	Florin Grigorian
19	Progressive Visual Loss: An Unusual Presenting Symptom in Giant Cell Arteritis	Parima Hirunwiwatkul
20	Demyelinating Disorder with Optic Neuropathies in a Patient with Monoclonal Gammopathy: Case Report and Literature Review	Whitney B. Hough
21	Retrospective Review of Ophthalmic and Systemic Associations of Optic Nerve Hypoplasia	Imran Jivraj
22	Optic Nerve Morphology as Marker for Disease Severity in Cerebral Palsy of Perinatal Origin	Sachin Kedar



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm)</b>		<i>continued</i>
23	Clinical Characteristics of Optic Neuritis Associated with Viral Infection	Sa Kang Kim
24	Methanol Causes Highly Selective Retinal Ganglion Cell Layer Loss and Inner Nuclear Layer Microcysts	Kendra A. Klein
25	Parsing the Differences Between LHON Affected: Genetic Vs Environmental Triggered Disease	Valerio Carelli
26	Changes in Macular OCT Retinal Sublayers of Patients and Carriers in the Natural History Phase of the Leber Hereditary Optic Neuropathy G11778A Gene Therapy Clinical Trial	Byron L. Lam
27	Low Grade Glioma of the Pituitary Stalk, Case Report with Review of the Literature	Su Ann Lim
28	Visualisation of Nerve Fibre Orientation in the Human Optic Chiasm Using Photomicrographic Image Analysis	Neeranjali Jain
29	Optic Neuropathy in Chronic Lymphocytic Leukemia	Amina I. Malik
30	How is Eye Fixation Affected by Optic Neuropathy? Diagnostic Value of Precise Recording of Retina Movement During an OCT Scan	Robert M. Mallery
31	A Novel OPA1 Mutation in Autosomal Dominant Optic Atrophy (ADOA)	Jordan A. Margo
32	The Vegetative Aspects of Neuroprotective Action of High Corticosteroid Doses in Compressive Traumatic Optic Neuropathies (TON)	Yulya T. Maslyak
33	Optic Atrophy in a Large Specialist Hospital	Joyce N. Mbekeani
34	Neurofibromatosis 1 with Large Suprasellar and Bilateral Optic Nerve Pilocytic Astrocytoma	Joyce N. Mbekeani
35	Optic Neuropathy in Wolfram's Syndrome Imaged with High-Definition Spectral Domain OCT	Dan Milea
36	Retinal Oximetry (Oxygen Saturation) and Peripapillary Vascular Diameters in Normal Eyes and in Non-Arteritic Ischemic Optic Neuropathy (NAION)	Ashwin Mohan
37	Is Ishihara Color Plate Testing as Reliable on iPod/iPhone and iPad as on Paper Format?	Katherine Boudreault
38	Multifactorial Optic Neuropathy - When it isn't Always Glaucoma	Deborah C. Parish
39	Cobalt-Chromium Metallosis with Normal ERG	Huy V. Nguyen
40	Nonarteritic Anterior Ischemic Optic Neuropathy (NAION): A Misnomer. A Non-Ischemic Papillopathy Caused by Vitreous Separation	Cameron F. Parsa
41	Visual and Oculomotor Outcomes in Children with Posterior Fossa Tumors	Crandall E. Peeler
42	Radiation Optic Neuropathy and Retinopathy from Low Dose (20Gy) Radiation Treatment	Crandall E. Peeler
43	Bilateral Optic Neuropathy in Superficial Intracranial Siderosis	Thong D. Pham
44	Binocular Acuity Summation (BAS) in Multiple Sclerosis (MS): Relation to Retinal Architecture and Visual System Neurophysiology	Sara S. Qureshi
45	Nutritional Optic Neuropathy after Bariatric Surgery	Yuna Rapoport
46	Lyme Disease Mimicking Giant Cell Arteritis	Nailyn Rasool
47	Structural Analyses of the Anterior Visual Pathway in Compressive Neuropathy	Won Hyung A. Ryu



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm)</b>		<i>continued</i>
48	Changes in Thickness of Retinal Segments in the Early Course of Non-Arteritic Ischaemic Optic Neuropathy	Bernardo F. Sanchez-Dalmau
49	A First Case Report of Chordoid Glioma Invading Optic Nerve	Nicolae Sanda
50	Clinical Spectrum of Optic Neuritis in Indian Children at a Tertiary Care Centre	Swati Phuljhele
51	Prognosticating Factors in Nonarteritic Ischaemic Optic Neuropathy	Swati Phuljhele
52	Sweep My Blindness Away!	Salwa Abdel Aziz
53	Progressive Non-Arteritic Optic Neuropathy as the First Presentation of Anti-Phospholipid Antibody Syndrome	Tarek A. Shazly
54	Anterior Visual Pathway(AVP) Meningiomas: A Dosimetric Comparison of IMRT to Pencil-beam Proton Therapy	Scott L. Stafford
55	A Case of Lyme Neuroretinitis- An Elusive Entity	Padmaja Sudhakar
56	50% of Non-Arteritic Anterior Ischemic Optic Neuropathy Occurs Between 40-55 Years Old	Ming-Hui Sun
57	Evaluation of Retinal Nerve Fiber Layer and Ganglion Cell Complex in a Cohort of Chinese with Optic Neuritis or Neuromyelitis Optica Spectrum Disorders Using SD-OCT	Guohong Tian
58	Transient Monocular Vision Loss Upon Awakening: A Benign Phenomenon	Marc A. Bouffard
59	Autosomal Dominant Optic Atrophy In Singapore: Multiethnic Involvement And Report Of A New Gene Mutation Causing Optic Atrophy And Deafness	Sharon L. Tow
60	Optic Nerve Head and Macular Choroidal Vascularization in Patients Affected by Normal Tension Glaucoma and Non-Arteritic Ischemic Optic Neuropathy: What Do They Share? What Is Different?	Giacinto Triolo
61	Multifocal ERG Shows Pre-Ganglion Cells Dysfunction in Dominant Optic Atrophy: Genotype-Phenotype Correlation	Maria Lucia Cascavilla
62	Pediatric Primary Optic Nerve Sheath Meningioma	Kavin Vanikieti
63	Syphilitic Optic Neuropathy : Reemerging Cases Over A 2-Year Period	Anuchit Poonyathalang
64	Anterior Ischemic Optic Neuropathy as Sole Presenting Sign of Internal Carotid Artery Occlusion	Kimberly M. Wings
65	Challenges in the Management of Apoplexy of a Growth Hormone(GH)-Secreting Pituitary Adenoma During Pregnancy	John Wong
66	The Prevalence of Mitochondrial Disease in the Adult Population – Implications for the Prevention of Maternal Transmission	Patrick Yu-Wai-Man
67	Neuropathies in Children Involving Vision Caused by a Fall From Height	Alon Zahavi
68	Immunosuppressive Therapy of Chinese Isolated Non-MS Idiopathic Optic Neuritis	XiaoJun Zhang
69	How does Exogenous ROS Effect LHON Conversion?	Jeffrey Tran
<b>Category: Cranial Nerves (Paresis, etc.)</b>		
70	Chronic, Painful Abducens Palsy May Require Angiography: Lesson Re-Learned from a Case of a White-Eyed Shunt.	Aliaa H. Abdelhakim
71	Eight and a Half Syndrome-A Rare Neuro Ophthalmologic Manifestation Due to Pontine Infarction	Sanam Anwer



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Cranial Nerves (Paresis, etc.)</b>		<i>continued</i>
72	Where the Lung Meets the Eye	Nathan W. Blessing
73	Metastasis or Metamorphosis?	Krista Kinard
74	Acute Onset Internuclear Ophthalmoplegia and Migraine Headache	Oana M. Dumitrascu
75	Traumatic Avulsion of the Oculomotor Nerve: First Definitive Documentation on High Resolution MRI	Lauren C. Ditta
76	Ocular Motor Cranial Nerve Palsies in Pituitary Apoplexy	Rabih Hage
77	Later Life Decompensation of Congenital Trochlear Palsy due to Agenesis	Ji-Soo Kim
78	Presentation of Primary Hodgkin's Lymphoma as Multiple Cranial Neuropathies	Hreem N. Patel
79	Acute Angle Closure Glaucoma in a Patient with Miller Fisher Syndrome without Pupillary Dysfunction	Won Yeol Ryu
80	Congenital Oculomotor Nerve Paresis with Isolated Cyclic Pupillary Spasms	Michael Salman
81	Abducent Nerve Palsy in a Patient with Castleman Disease!	Islam Zaydan
82	Nivolumab Plus Ipilimumab Induced Aseptic Meningitis and Bilateral Sixth Nerve Palsy in Metastatic Melanoma Treatment	Mitchell Strominger
83	Combined Third Nerve Palsy and Vertical Gaze Palsy in Midbrain Thalamic Stroke	Kara Fister
84	A Case of Abducens Nerve Palsy Followed by Cyclic Esotropia: A Case Report and Review of the Literature	Takako Sugimoto
85	Functional MRI and MRI Tractography in Progressive Supranuclear Palsy-Like Syndrome	Michael Vaphiades
86	Abducens Palsy in a Patient with Pityriasis Rubra Pilaris	Amanda L. Way
<b>Category: CSF (Intracranial Hypertension, Intracranial Hypotension, etc.)</b>		
87	Characterization of Idiopathic Intracranial Hypertension (IIH) in Qatar	Mais N. Alkawaz
88	Osmometry of Cerebrospinal Fluid from Patients with Idiopathic Intracranial Hypertension (IIH)	Steffen E. Hamann
89	Corneal Biomechanics : A Journey Through Uncharted Territories	Ashwin Mohan
90	Clinical and Prognostic Significance of CSF Closing Pressure in Pediatric Pseudotumor Cerebri Syndrome	Shannon J. Beres
91	Subretinal Fluid in Idiopathic Intracranial Hypertension: Clinical Course and Outcome	John J. Chen
92	Overdiagnosis of Idiopathic Intracranial Hypertension (IIH)	Adeniyi Fisayo
93	An Update on Terson Syndrome: Prevalence and Prognosis	Philip S. Garza
94	Aberrant Presentation of Pseudotumor Cerebri	Sawyer B. Hall
95	Jugular Vein Thrombosis as a Cause of Intracranial Hypertension	Josepha Horowitz
96	Autism Spectrum Disorder in Pediatric Pseudotumor Cerebri Syndrome	Anne K. Jensen
97	Lumbar Drain in Fulminant Intracranial Hypertension	Mays A. El-dairi
98	Papilledema with Unilateral Occlusion of Internal Jugular Vein	Abhishek Thandra
99	Papilledema Secondary to Internal Jugular Vein Thrombosis in a Dialysis Patient	Shauna W. Berry
100	Ultramicroscopic Study of the Optic Nerve Sheath in Patients with Severe Vision Loss from Idiopathic Intracranial Hypertension- Methodology	Joshua W. Evans



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: CSF (Intracranial Hypertension, Intracranial Hypotension, etc.)</b>		<i>continued</i>
101	Ultramicroscopic Study of the Optic Nerve Sheath in Patients with Severe Vision Loss from Idiopathic Intracranial Hypertension- Results	Marla Davis
102	Exploratory Study of the Relationship Between the Levonorgestrel-Releasing Intrauterine System and Idiopathic Intracranial Hypertension	Reuben M. Valenzuela
103	Coning Following Lumbar Puncture in a Patient with Idiopathic Intracranial Hypertension	Christian J. Lueck
104	Long Term Follow-Up of PTC: Pre-Pubertal Children vs. Adolescents and Adults	Assaf Hilely
105	Hemodialysis Graft-Induced Intracranial Hypertension	Devin D. Mackay
106	Incidence of Idiopathic Intracranial Hypertension (IIH) Among Users of Tetracycline Antibiotics	Samuel F. Passi
107	CSF Characteristics in Patients with High Frisén Papilledema Grades from the IIHTT	John H. Pula
108	Visual Outcomes After Treatment of Venous Sinus Stenosis with Dural Venous Sinus Stenting	Ahmara G. Ross
109	Relationship between High Opening Pressure on Lumbar Puncture and Failure of Optic Nerve Sheath Decompression to Prevent Progressive Visual Loss in Patients with Idiopathic Intracranial Hypertension	Mark E. Robinson
110	Venous Sinus Stenting for Treatment of Increased Intracranial Pressure Secondary to Venous Sinus Stenosis	Nikisha Richards
111	Pediatric Primary Pseudotumor Cerebri Syndrome (PTCS): A Detailed, Retrospective, Multicenter Analysis of Anthropometrics	Claire A. Sheldon
112	Endovascular Venous Stenting in Treatment of Primary Idiopathic Intracranial Hypertension	Rajeev Sivasankar
113	Idiopathic Intracranial Hypertension Mimicking Foster Kennedy Syndrome	Chuan-Bin Sun
114	Going for the Jugular! Paragangliomas, Papilledema, and Vision Loss	Jonathan Trobe
115	Visual Outcomes Following Optic Nerve Sheath Fenestration	Neel S. Vaidya
116	New Side Effect of Acetazolamide: Palinopsia	Peggy H. Vogt
117	Endovascular Intervention in a Chronic Case of Idiopathic Intracranial Hypertension (IIH)	Muhammad-Atif Zubairi
<b>Category: Disorders of Vision Processing</b>		
118	Optical Aberrations- A Trigger Factor for Migraine?	Rohit Shetty
119	An Interesting Field of Study	Andrew S. Camp
120	Visual Field Assessment in the "Split-Brain" Patient: Effect of the Method/Laterality of Obtaining Patient Responses on Apparent Visual Field Defect Size	J. Alexander Fraser
121	Binasal Hemianopsia	Erica Ballard
122	Voice Processing in Developmental Prosopagnosia	Ran R. Liu
123	Building a Repository for Posterior Cortical Atrophy	Jennifer J. Olds
124	Subretinal Choroidal Neovascularization Associated with Unilateral Papilledema and IIH: Case Report	Jalpa M. Patel
125	The Initiation of Smooth Pursuit Eye Movements in Anisometropic Amblyopia	Rana A. Raashid
126	The Pulfrich Phenomenon: An Objective Signature-Biomarker of MS Pathophysiology	Millad J. Sobhanian
127	Distractibility in Multiple Sclerosis: A Potential Cause of Morbidity	Derek Sears



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Disorders of Vision Processing</b>		<i>continued</i>
128	Disconjugacy of eye alignment is greater with near fixation during binocular viewing in amblyopia	Vivian Xu
<b>Category: Misc. Motility Disorders (Nystagmus, etc.)</b>		
129	An Adjustable Magnetic Prism Carrier for Strabismus Evaluation	David S. Bardenstein
130	Patient Satisfaction with Prismatic Correction of Diplopia	Shauna E. Berry
131	Duane Retraction Syndrome in Duchenne Muscular Dystrophy	Thomas M. Bosley
132	Peribulbar Botulinum Toxin as a Treatment for Symptomatic Opsoclonus in Oculopalatal Myoclonus	Katherine Duncan
133	Thalamic Stroke in a Young Patient Presenting with Sudden Onset Large Skew Deviation	Oana M. Dumitrascu
134	Ophthalmological Spectrum of Locked-In Syndrome	Martin Graber
135	Cigarette Smoking and Activities of Daily Living in Ocular Myasthenia Gravis	Sean M. Gratton
136	An Evaluation of Educational Neurological Eye Movement Disorder Videos Posted on Internet Video Sharing Sites	Simon J. Hickman
137	How Do Patients with Strabismus Locate Visual Targets?	Jonathan C. Horton
138	Superior Cerebellar Peduncle Demyelination Causing Geotropic Central Positional Nystagmus	Cristina Duque
139	Normative Database for the King-Devick Test in Adults and Adolescents	Danielle Leong
140	Marcus Gunn Jaw Winking with Electronegative Cone-Rod Dystrophy: Case Report	Su Ann Lim
141	Levodopa-Induced Ocular Dyskinesias	Bryan V. Pham
142	Clinical Features, Diagnostic Findings and Treatment of Adult-Onset Opsoclonus-Myoclonus Syndrome: A Case Series	Olga Rosenvald
143	Divergence Palsy Due to Antiepileptic Drugs	Marc A. Bouffard
144	Quantifying the Vestibulo-ocular Reflex with Video-Oculography: Nature and Frequency of Artifacts	Ali S. Saber Tehrani
<b>Category: Miscellaneous</b>		
145	Neuroophthalmic Manifestations of Intracranial Tumours in South India	Kowsalya Akkayasamy
146	Sideline Testing in Youth and Collegiate Athletes: What Does Vision Add to the Concussion Puzzle?	Laura J. Balcer
147	Assessment of Recruitment Patterns in a Neuro-Ophthalmology Registry	Kimberly D. Blankshain
148	Prediction of Eye Position During General Anesthesia Using Bispectral Index Monitoring	Seung Ah Chung
149	Demographic Profile of the Patients Presenting to a Neuro-Ophthalmology Clinic of a Tertiary Eye Care Centre	Shiva Prasad Gantyal
150	Botulinum Toxin-Augmented Strabismus Surgery versus Conventional Surgery in the Treatment of Large-Angle Infantile Esotropia	Aubrey L. Gilbert
151	Heidenhain Variant of Creutzfeldt Jakob's Disease (CJD), In a Patient Who had Bovine Bioprosthetic Valve Implantation	Haneen Jabaly-Habib



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Miscellaneous</b>		<i>continued</i>
152	The Effects of Pediatric Primary Brain Tumors on Vision and Quality of Life	Supharat Jariyakosol
153	A Method for Quantifying off Chart Visual Acuities	Rustum Karanjia
154	Differential Functions Mediated by Melanopsin Assessed in Subjects with Healthy and Diseased Eyes	Aki Kawasaki
155	Familial Papillitis and Macular Cystoid Edema: A Genetic Autoimmune Disorder?	Chiara La Morgia
156	New Daily Persistent Headache Triggered by Cataract Extraction?	Julia E. Mallory
157	Ten Years of Temporal Artery Biopsies in Ontario, Canada: A Population-Based Study on Practice Patterns and the Incidence of Giant Cell Arteritis	Jonathan A. Micieli
158	A Method for Recognizing Colorblind Malingering	Andrew E. Pouw
159	Mycotic Aneurysms of Intracavernous Internal Carotid Artery	Kumudini Sharma
160	Platelet-Mediated Microvascular Ischemia Mimicking Migraine in Patients with Thrombocythemia	Melissa C. Tien
<b>Category: Nerve Fiber Layer and Retinal Testing (OCT, ERG, etc.)</b>		
161	Ganglion Cell Damage and Functional Recovery after Optic Neuritis	Alexander U. Brandt
162	To Evaluate the Retinal Nerve Fibre Layer Loss in Optic Neuritis Using Spectral Domain Optical Coherence Tomography	Kowsalya Akkayasamy
163	Retinal Ganglion Cell Injury in Early Pediatric Onset MS	Jennifer Graves
164	The Correlation of Critical Flicker Fusion Function and P100 Latency of Visual Evoked Potential with Luminance	Yanjun Chen
165	The Effects of Amblyopia on Visual Evoked Potentials	Florin Grigorian
166	Doubling Method for Papilledema and Pseudopapilledema	Bokkwan Jun
167	Correlation Between Structural and Functional Changes in Retina in Parkinson's Disease	Manpreet Kaur
168	Relationship Between Visual Acuity and Retinal Nerve Fiber Layer Thickness Measured by Spectral Domain Optical Coherence Tomography in Patients with Optic Neuropathy	Sungeun Kyung
169	Ganglion Cell Layer Thinning Detected by Optical Coherence Tomography as a Sign of Early Optic Atrophy in Pediatric Papilledema	Andrew R. Lee
170	Peripapillary Retinal Nerve Fiber Layer Thickness Corresponds to Drusen Location and Extent of Visual Field Defects in Patients with Optic Disc Drusen	Lasse Malmqvist
171	Progressive Retinal Structure Abnormalities in Multiple System Atrophy	Carlos E. Mendoza-Santiesteban
172	OCT Shows Consistent Relationships Between Macular Layers and Peripapillary RNFL in Chronic Demyelinating and Compressive Optic Neuropathies	Mark J. Morrow
173	OCT Shows Different Relationships Between Macular Layers and Peripapillary RNFL in Acute Versus Chronic Papillitis and Papilledema	Fawzi Abukhalil
174	The Photopic Negative Response in Idiopathic Intracranial Hypertension	Heather E. Moss
175	Attenuated Hunter Syndrome: A Rare Cause of Simultaneous Retinal and Optic Nerve Disease	Kannan Narayana
176	Ganglion Cell and Retinal Nerve Fiber Layer Analysis in a Case of PION	Joshua Pasol



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Nerve Fiber Layer and Retinal Testing (OCT, ERG, etc.)</b>		<i>continued</i>
177	Perimacular Ganglion Cell Complex Thinning Detected By Spectral-Domain OCT Useful In Detecting Optic Tract Syndrome	Fannie Petit
178	To Evaluate Changes in Retinal Nerve Fiber Layer and Ganglion Cell Layer on Cirrus HD-OCT in Cases of Multiple Sclerosis (with and without Optic Neuritis) and Optic Neuritis	Ganesh Pillay
179	Examination of Visual Evoked Potential (VEP) in a Pediatric Population with Newly Diagnosed Elevated Intracranial Hypertension	Ahmara G. Ross
180	Retinal Nerve Fiber Layer Thickness in a Population-Based Study of Elderly Subjects: The Alienor Study	Marie B. Rougier
181	OCT-Derived Retinal Capillary Density is Decreased in Corresponding Areas of Retinal Neuron Loss in Optic Neuropathy	Min Wang
182	Adaptive Optics Imaging with Histopathologic Correlation in Cancer-Associated Retinopathy	Zoë R. Williams
183	Robust Optic Nerve Head Analysis Based on 3D Optical Coherence Tomography	Ella M. Kadas
<b>Category: Neuro-Imaging (MRI, CT, etc.)</b>		
184	Ocular Motility Defects with Concordance Neuroimaging	Nagham Al-Zubidi
185	Mural Enhancement of the Intracranial Internal Carotid Artery in Giant Cell Arteritis	Sidney M. Gospe III
186	Morning Glory Disk Anomaly Associated with Absence of Intracranial Internal Carotid Artery	Adriana P. Grigorian
187	Measurement of Optic Nerve Sheath Diameter by CT, MRI and Ultrasound	Klara Landau
188	Occipital Partial Status Epilepticus with Abnormal MRI Imaging	Sonalee Kulkarni
189	Neuro-Imaging Characteristics of Common Extraocular Prosthetic Devices	Samantha F. Lu
190	A Lion in the Bush	Padmaja Sudhakar
<b>Category: Orbital and Eyelid Disorders</b>		
191	Why the Delay in Diagnosis? Increased Time From Symptom Onset to Diagnosis in Blepharospasm: A Prospective, Clinic-Based Study	Kristen E. Dunbar
192	Atypical Presentation of Orbital Lymphangioma	Haydée S. Martinez
193	Prospective Assessment of Peri-Oral Weakness Following Peri-Orbital Botulinum Toxin for Blepharospasm	Alexander J. Hartmann
194	Vertical Diplopia and Ptosis from Removal of the Orbital Roof in Pterional Craniotomy	Jonathan C. Horton
195	Treatment of rivaroxaban associated orbital hemorrhage	Marilyn C. Kay
196	Stonewalled: Bilateral Sequential Vision Loss In A Peritoneal Dialysis Patient Category: Systemic Disease	Angelina Espino Barros Palau
197	Allergic Fungal Sinusitis Mimicking Thyroid Orbitopathy	Lina Nagia
198	Optic Nerve Sheath Meningioma. No Longer Impossible but Still Difficult	T. Ben Ableman
199	An Advanced Pancoast Tumor Masquerading as Congenital Hereditary Ptosis	Deborah C. Parish
200	It's All Going Dark!	Tarek A. Shazly
201	Bartonella Henselae-Associated Neuroretinitis with Orbital Intraconal and Optic Nerve Sheath Involvement	Ann Shue
202	Impact of Injection Site on Dose Escalation in the Treatment of Blepharospasm	Rachel G. Simpson





# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Orbital and Eyelid Disorders</b>		<i>continued</i>
203	A Case of Nasopharyngeal Carcinoma Masquerading as Primary Orbital Tumour	Shantha Amrith
204	Visual Field and Graves Ophthalmopathy	Sabine Defoort
<b>Category: Posterior Afferent Visual Pathway (Post-Chiasmal)</b>		
205	VEPs to Lateralized Stimuli to Measure the Interhemispheric Transfer Time (IHTT)	Ilie P. Cretu
206	The Heidenhain Variant of Creutzfeldt-Jakob Disease	Kristen E. Dunbar
207	Six-Year Follow-Up of the Progression of Cortical Vision Loss in a Patient with HIV-Related Progressive Multifocal Leukoencephalopathy	Bahareh Hassanzadeh
208	Retrograde Degeneration of Retinal Ganglion Cells in Homonymous Hemianopia	Angela M. Herro
209	Lesions of the Optic Tract: A Review of 35 Cases	Kristopher M. Kowal
210	Hydrocephalus and More!	Islam Zaydan
<b>Category: Pupil</b>		
211	Ipratropium-Induced Mydriasis: A Possible Exception to the Current Anisocoria Diagnostic Pathway	Erica L. Archer
212	Automated Pupillograph as a Screening Tool in Ophthalmology Clinic	Ashwin Mohan
213	The Yield of Diagnostic Imaging in Patients with Isolated Horner's Syndrome	Johanna D. Beebe
214	Evaluation of Pupil Response as Ocular Marker for Pre-Clinical Alzheimer's Disease	Ling Bei
215	A Case of Horner's Syndrome After Clipping of the Internal Carotid-Posterior Cerebral Artery Aneurysm Associated with Subarachnoid Hemorrhage	Tamura Koichiro
216	Test-Retest Reliability of Hemifield, Central-Field and Full-Field Chromatic Pupillometry for Assessing the Function of Melanopsin-Containing Retinal Ganglion Cells	Shaobo Lei
217	Unique Presentation of Anti-GQ1b Antibody Syndrome	Tin Yan A. Liu
<b>Category: Retina</b>		
218	Novel Retinal Observations in Genetically Confirmed Kearns Sayre Syndrome	Thomas M. Bosley
219	Autoimmune Retinopathy and Optic Neuropathy in a patient with Anti-GAD and Other Retinal Antibodies	Sumayya J. Almarzouqi
220	GM2-gangliosidosis, AB variant: An Elusive Cause of Neurodegenerative Cherry Red Spots	Michael C. Brodsky
221	Malignant Optic Nerve Glioma Manifesting as Central Retinal Vein Occlusion and Papilledema	Ryan C. Burton
222	Comprehensive Postmarketing Review of Visual Defects Reported with Topiramate	Lisa Ford
223	Sequential Fundus Findings After Platelet Rich Plasma for Facial Rejuvenation	Emely Z. Karam
224	Contrast Sensitivity Visual Acuity is Degraded in REM Sleep Behavior Disorder	Matthew J. Khayata
225	Ophthalmoscopy Vs. MRI – Sometimes Less is More	Anton Kolomeyer
226	Retinal Vessel Oximetry and Vessel Diameter Measurements: A Novel Metabolic Marker of Multiple Sclerosis	Marlen Lucero
227	Acute Zonal Occult Outer Retinopathy Presenting as Retrobulbar Optic Neuritis	Sungeun E. Kyung
228	Long-term Follow-up of Laser Pointer Induced Macularopathy	Chuan-Bin Sun



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Retina</b>		<i>continued</i>
229	Use of Wide-field Retinography for Periphlebitis Detection in Patients with Multiple Sclerosis	Ruben Torres-Torres
230	An Atypical Presentation of Giant Cell Arteritis	Laurel N. Vuong
<b>Category: Systemic Disease</b>		
231	Discordance Rates in Healing/Healed Arteritis in Temporal Artery Biopsies: The Role for Bilateral Biopsies	Sangsu Han
232	Bilateral Intracranial Optic Nerve and Chiasmal Involvement in IgG-4 Related Disease	Raed Behbehani
233	Neuromyelitis Optica Preceded by Seizure Disorder	Iris Ben Bassat Mizrahi
234	Optic Nerve Meningioma Masquerading as Neurosarcoidosis	Ryan C. Burton
235	Comparison of the Clinical Characteristics of Patients with Active Arteritis and Healed Arteritis, the Two Histopathological Patterns Considered Positive in Giant Cell Arteritis	Michael W. Salter
236	Autologous Hematopoietic Stem Cell Transplantation in Neuromyelitis Optica - An Update	Fiona Costello
237	Social Media and Susac Syndrome	Robert A. Egan
238	Horner's Syndrome as Initial Manifestation of a Malignant Peripheral Nerve Sheath Tumor	Gerard L. Hershewé
239	Birefringence of Retinal Nerve Fiber Layer and Retinal Blood Flow Velocity in Multiple Sclerosis	Hong Jiang
240	Botox for Chronic intractable Headache among Veterans at VA Medical Center, Long Beach, California	Grace W. Kao
241	Thin-Film Optical Notch Filter Spectacle Coatings for the Treatment of Migraine and Photophobia	Bradley J. Katz
242	Monitoring Alzheimer's Disease with Retina Examination	Umur A. Kayabasi
243	Why is He Losing His Sight? He is Losing His Photoreceptor!	Ainat Klein
244	Extensive Macular Serous Retinal Detachment as the Initial Presentation of Bartonella Henselae Induced Neuroretinitis	Darrell Lewis
245	Visual Outcomes in Giant Cell Arteritis in Patients with Polymyalgia Rheumatica on Prior Corticosteroids	Nailyn Rasool
246	Progressive Supranuclear Palsy (PSP) After Ascending Aorta Dissection Surgery	Hyosook Ahn
247	Delineation of Natural History for Spinocerebellar Ataxia Type 7 (SCA-7) in Anticipation of Rnai Therapy	Steven F. Stasheff
248	Study of the Dynamics of Axonal Degeneration in Chemotherapy-Induced Neuropathy by in Vivo Corneal Confocal Microscopy	Ruben Torres-Torres

## Poster 1

### Normal Tension Glaucoma (NTG) is it a Type of Glaucoma or an Entity of a Compressive Optic Neuropathy (CON)?

Mohammed Areesh<sup>1</sup>, Seyed Mohammad Mirhosseini, Elahe Rajaei

*Zahedan Medical University, Zahedan Eye Hospital, Zahedan, Iran*

#### Introduction:

NTG is known to be a special type of glaucoma which is still a matter of dispute in its etiologic, diagnostic, therapeutic aspects and its terminology<sup>(1)</sup>. A lot of studies have been carried out, that indicated difference of POAG vs NTG such as: 1. Optic nerve head localized affection in NTG vs POAG. <sup>(2)</sup> 2. overall VF defect is less affected in NTG and both central and paracentral defect were seen vs PAOG. <sup>(2,3)</sup> 3. Genetic differences. <sup>(4)</sup> 4. Optic nerve head blood flow. <sup>(5)</sup> 5. The NTG patients were more younger. <sup>(1)</sup> In this study we propose that labeling these patients as NTG could be a misinterpretation and NTG could be really an entity of a CON.

#### Methods:

Total of 27 patient's eyes (10 men and 17 women) were included in this study, and Best corrected visual acuity (BCVA), IOP, vertical c/d ratio, VF test, Ishihara Red/Green test, Gonioscopy, Relative afferent pupillary defect were established on all cases.

#### Results:

Total of 20 (6 men and 14 women) patient's eye from 27 patient's eye were included in this study so the results are as below: 1. BCVA was achieved and 20/20 in all cases. 2. The mean IOP measured so no IOP greater than 21 mmHg was recorded. 3. The vertical c/d ratio was measured and significantly different in each patients. 4. The Humphrey VF study indicates central relative or absolutely defects. 5. Ishihara revealed relative Red-Green deficiency in both eyes according to plate's response were detected. 6. detectable RAPD recorded in the eye with larger c/d ratio in all patients.

#### Conclusions:

All these signs coincide with the compression of the optic nerve. So the authors of this article have come to the conclusion that NTG be considered under category of optic neuropathy. In fact we think that it may be a type other than glaucoma and maybe misnamed. According to this study all the clinical signs are in favor of the CON.

#### References:

1. Essentials in ophthalmology, Glaucoma edited by F. Grehn, R. Stamper, R.A. Hitchings, chapter 10. (page 147-155)
2. Hayamizu F, Yamazaki Y, Nakagami T, Mizuki K. Clin Ophthalmol. 2013;7:807-13. doi: 10.2147/OPTH.S42468. Epub 2013 May 3.
3. Lee J, Kong M, Kim J, Kee C. J Glaucoma. 2013 Jun 6.
4. Fraenkl SA, Golubnitschaja O, Yeghiazaryan K, Orgül S, Flammer J. Eur J Ophthalmol. 2013 May 31;23(6):841-849. doi: 10.5301/ejo.5000306.
5. Mamikonian VR, Galoian NS, Sheremet NL, Kazarian EE, Kharlap SI, Shmeleva-Demir OA, Andzhelova DV, Tatevosian AA. Vestn Oftalmol. 2013 Jul-Aug;129(4):3-8. Russian

**Keywords:** Normal Tension Glaucoma, Compressive Optic Neuropathy, Primary Open Angle Glaucoma, Visual Field Defect, Red-Green Deficiency

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Zahedan Medical University, Zahedan Eye Hospital

## Poster 2

### Anterior Ischemic Optic Neuropathy Due To Biopsy-Proven Giant Cell Arteritis In Thai Patients

Taweevat Attaseth<sup>1</sup>, Kavin Vanikiet<sup>1</sup>, Anuchit Poonyathalang<sup>1</sup>, Pisit Preechawat<sup>1</sup>, Panitha Jindahra<sup>2</sup>, Duangkamon Wattanatranon<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand, <sup>2</sup>Department of Medicine, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand, <sup>3</sup>Department of Pathology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand

#### Introduction:

Giant cell arteritis (GCA) is a vasculitis of medium- to large-sized artery which can interfere with arterial flow to the optic nerve head causing arteritic anterior ischemic optic neuropathy(AION). Little data has been published regarding arteritic AION in Asians, including Thais probably due to its rarity.

#### Methods:

The records of 236 patients with AION, seen during January 2005 to October 2014 at a tertiary care center in Bangkok, Thailand were reviewed. Only 6 patients with arteritic AION due to biopsy-proven giant cell arteritis were included in this study.

#### Results:

Of 6 patients, 4 (67%) were male and 2 (33%) were female. Mean age at onset was 72.5 years, ranged from 63 to 81 years. Two patients had bilateral simultaneous involvement and 4 patients had unilateral involvement. In unilateral AION, 2 patients developed contralateral visual loss from cilioretinal artery occlusion 2 weeks before the onset of AION. All eyes had severe visual loss (20/200 or worse) at initial presentation including 3 eyes with no light perception. Fluorescein angiography showed choroidal filling delays in 4 eyes. All patients had an elevated erythrocyte sedimentation rate (mean 95 mm/hr). High dose intravenous methylprednisolone and subsequent long-term oral corticosteroid were given in all cases. Final visual acuity remained unchanged in most of patients (5 eyes, 63%). Three eyes (37%) showed improvement of their visual acuity and 2 eyes achieved a final visual acuity of 20/50.

#### Conclusions:

Arteritic AION in GCA is a rare disease and represents 2.5 % of AION patients in a tertiary neuro-ophthalmologic service in Thailand. The clinical features and visual outcome in Thai patients are not different to those describe in white patients.

**References:** None.

**Keywords:** Neuro-Ophth & Systemic Disease ( Eg. MS, MG, Thyroid), Optic Neuropathy, Vascular Disorders, Retina, Orbit/Ocular Pathology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 3

### Visual Outcomes Following Perioperative Vision Loss

Jasmina Bajric<sup>1</sup>, Mohamed Hassan, Maria Garcia, Mark A Warner, Elizabeth A Bradley

*Mayo Clinic, Department of Ophthalmology, Rochester, MN, USA*

#### Introduction:

The aim of this study was to determine visual acuity outcomes following perioperative vision loss due to ischemic optic neuropathy, retinal artery or vein occlusion, and cortical blindness.

#### Methods:

In this retrospective chart review, all surgical procedures performed on adults and requiring general anesthetic between 2003 and 2012 at the Mayo Clinic were screened for visual complications. Patients who had new-onset vision loss within 30 days of surgery and a complete ophthalmic exam were included. Patients with surgery on the visual pathways or whose diagnosis was not related to surgery were excluded. Outcomes were determined up to 1 year after surgery.

#### Results:

A total of 41 cases of perioperative vision loss were identified after screening 352, 717 general anesthetic procedures (risk of 0.012%). Visual acuity was 20/490 for all patients at baseline and improved to 20/295 at 3 months ( $p = 0.27$ ), 20/132 at 6 months ( $p = 0.001$ ) and 20/53 at 1 year ( $p = 0.02$ ). A similar pattern of visual acuity improvement was seen in 14 patients who had complete 1-year follow-up: 20/289 at baseline, 20/118 at 3 months ( $p = 0.004$ ), 20/100 at 6 months ( $p = 0.006$ ), and 20/53 at 1 year ( $p = 0.02$ ). Visual field deficits were 73% at baseline and 83% at 1 year ( $p = 0.56$ ). Color vision deficits were 57% at baseline and 79% at 1 year ( $p = 0.63$ ). Ischemic optic neuropathy was the most common cause (57%), followed by retinal artery occlusion (19%), retinal vein occlusion (12%) and cortical blindness (12%).

#### Conclusions:

The risk of perioperative vision is low. Visual acuity improves throughout the first year following perioperative. Visual field and color vision deficits remain similar to baseline at one year following vision loss.

**References:** None.

**Keywords:** Optic Neuropathy, Vascular Disorders, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 4

### Pituitary Apoplexy in Pregnancy

Matthew J. Benage<sup>1</sup>, Erica E. Ballard<sup>2</sup>, Theodore E. Wills<sup>2</sup>

<sup>1</sup>University of Missouri - School of Medicine, Columbia, MO, USA, <sup>2</sup>Mason Eye Institute, Columbia, MO, USA

#### Introduction:

We present a case of pituitary apoplexy of a pregnant female with an initial presentation of mild headache and bitemporal hemianopia.

#### Methods:

This is a descriptive case report based on clinical records, patient observation and analysis of diagnostic studies.

#### Results:

An 18 year old G1P0 woman presented at 26 weeks gestation (Di/Di twins) with new onset visual disturbance and a mild headache. She described the headache as dull and diffuse. She noted trouble seeing peripherally to her right. On initial exam, she displayed visually significant temporal field loss to confrontation OD. Dilated fundoscopic exam was normal OU. Within 12 hours, the patient noted worsening of visual disturbance and new field loss temporally to confrontation was noted OS. Due to the bitemporal field loss as evident on CVF, she underwent an MRI brain for possible pituitary pathology. The MRI displayed a T2 and FLAIR rounded hyperintense focus measuring 7 x 11 x 6 mm located in the pituitary. GRE and flare sequences were notable for a fluid-filled level within the structure representing pituitary apoplexy. The patient was admitted for initial work-up. She was scheduled for follow-up MRI and visual field testing in four weeks. The clinical course is ongoing at this time.

#### Conclusions:

This is a case of pituitary apoplexy in a pregnant woman with mild headache and bitemporal visual disturbances. Pituitary apoplexy is a rare diagnosis, especially in pregnancy. Management of apoplexy includes urgent corticosteroid replacement if indicated with continuous monitoring of electrolytes. In life-threatening cases, trans-sphenoidal decompression may be indicated. Pituitary apoplexy in pregnancy is best managed by a multidisciplinary team including endocrinologists, obstetricians, ophthalmologists, and neurosurgeons.

#### References:

1. Nawar RN, AbdelMannan D, Selman WR, Arafah BM. Pituitary tumor apoplexy: a review. J Intensive Care Med. Mar-Apr 2008;23(2):75-90
2. Pituitary Apoplexy in Pregnancy: two case reports. Mangion Z, Rogers A, Mackillop L. Regional Clinical Cases 2012. Oxford
3. A Case of Pituitary Apoplexy in Pregnancy. Hayes A, O'Sullivan A, Davies, M. Endocrinology Diabetes Metabolic Case Report. 2014

#### Keywords:

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 5

### Bilateral, Sequential Anterior then Posterior Ischemic Optic Neuropathy in a Young Migraineur

Damian E. Berezovsky<sup>1</sup>, Timothy W. Winter<sup>2</sup>

<sup>1</sup>Dept. of Neurology, University of New Mexico, Albuquerque, NM, USA, <sup>2</sup>Dept. of Ophthalmology, University of New Mexico, Albuquerque, NM, USA

#### Introduction:

Non-arteritic anterior (NAION) and posterior (PION) ischemic optic neuropathy infrequently affect patients under 50; the two largest case series suggest that this group represents up to 23% of NAION patients.<sup>1,2</sup> Within this group of young NAION patients, 41-43% had eventual bilateral involvement. Risk factors in this population included crowded discs (82-88%), hyperlipidemia (23-47%), hypertension (32-35%), smoking (27-29%) and migraines (20%). Sequential NAION followed by PION in a young patient has not been previously reported.

#### Methods:

Case report and review of the literature.

#### Results:

A 37 year old female with frequent, severe headaches associated with photophobia and nausea noted the acute onset of right-sided painless visual loss upon awakening. Visual acuity was 20/70 in the right eye and 20/30 in the left eye, with an afferent pupillary defect (APD) on the right, disc edema on the right, and normal fundus on the left except for a small cup: disc ratio of 0.1. Visual field testing revealed an inferior altitudinal defect on the right. Contrasted brain MRI, lumbar puncture, inflammatory markers, ANA screen, and hypercoagulation panel were unremarkable. She was given intravitreal triamcinolone without improvement, and superior optic nerve pallor was observed at two-month follow-up. At this visit, she reported the recent sudden onset of blurry vision in her left eye, with unchanged visual acuity, no APD, and no disc edema. VF testing revealed a new superior and inferior nasal defect on the left, and a repeat contrasted brain MRI was normal.

#### Conclusions:

This is a unique case of sequential, anterior then posterior ischemic optic neuropathy occurring in a young patient with frequent migraines and small cup: disc ratio bilaterally. This case, as well as previous case series, highlights the frequent association between migraine headaches and ischemic optic neuropathy.<sup>1-3</sup> This suggests that vasospasm may play a role in ischemic optic neuropathy, especially in young patients.

#### References:

1. Arnold AC, Costa RM, Dumitrascu OM. The spectrum of optic disc ischemia in patients younger than 50 years (an American Ophthalmological Society thesis). *Trans Am Ophthalmol Soc.* 2013 Sep;111:93-118.
2. Preechawat P, Bruce BB, Newman NJ, Biousse V. Anterior ischemic optic neuropathy in patients younger than 50 years. *Am J Ophthalmol.* 2007 Dec;144(6):953-60.
3. Lee AG, Brazis PW, Miller NR. Posterior ischemic optic neuropathy associated with migraine. *Headache.* 1996 Sep;36(8):506-10.

**Keywords:** Optic Neuropathy, Headache, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 6

### Traumatic Optic Neuropathy. Our experience.

Mariana de Virgiliis<sup>1,2</sup>, Pablo I Perez Vega<sup>1,2</sup>, Maria L Braccia Gancedo<sup>1,2,3</sup>, Haydee S Martinez<sup>1,3</sup>, Luciana Iacono<sup>2</sup>, Mirta Arana<sup>1,3</sup>, Macarena Clementi<sup>1,3</sup>

<sup>1</sup>Universidad de Buenos Aires, Buenos Aires, Argentina, <sup>2</sup>Hospital Oftalmológico Dr. Pedro Lagleyze, Buenos Aires, Argentina, <sup>3</sup>Hospital de Clínicas José de San Martín, Buenos Aires,

#### Introduction:

Traumatic Optic Neuropathy (TON) is the optic nerve damage related with encephalo-cranial traumatism. Usually affects young people with potential severe visual loss. There is no proven effective treatment.

#### Methods:

Retrospective review of medical histories of patients treated in ophtalmology hospital between January 2012 and March 2014, selected those patients diagnosed with TON.

#### Results:

There were 52 patients with TON. 79% males younger than 39 years old (73%), only 3,8% were over 60 years. Ninety five percent of the TON were unilateral and all cases were indirect traumatic optic neuropathies, due to blunt head trauma Among the causes, Traffic collisions were the most frequent (46%), 67% had a motorcycle involvement (mostly without helmet); followed by direct blunt trauma 35% and due to falls from height 19%. Regarding the initial trauma: 60% of patients had loss of consciousness; 68% were hospitalized, 56% had abnormal neuroimagen and 19% required neuro surgical treatment. The first neuro ophthalmology visit, was within two months in 52 % of patients, but only 9% within first week from initial trauma. Initial Visual Acuity (VA) was worst than 0.1 en 57% of patients, 13,4 % had No light Perception. First Month control (23 patients): 63% visual acuity improvement. Last control (32 patients): 58% improvement in visual acuity; meanwhile only 15% had better color test result Initial ophthalmoscopic findings was: Difuse optic disc pallor 31% Normal optic nerve 29%, Temporal Pallor 25%, Optic nerve cupping 14%. All types of visual field defects were found, none of which were prevalent. OCT main findings was diffuse decreased retinal nerve fiber layer (RNFL).

#### Conclusions:

NOT is a significant cause of severe visual loss in young men, mainly related with motorcycle accidents without helmet protection and associated with other life threatening conditions secondary to head injuries. Consistent with other reports, spontaneous visual acuity improvement was seen.

#### References:

1. Yu-Wai-Man P, Griffiths PG. Steroids for traumatic optic neuropathy. Cochrane Database Syst Rev. CD006032. 2013. Warner, Eggenberger . Traumatic optic neuropathy: a review of the current literatura. Current Opinion in Ophthalmology, 21:459 – 462. 2010
2. Yu-Wai-Man P, Griffiths PG. Surgery for traumatic optic neuropathy (Review). Coch Collab; 1:15 – 28. 2009

**Keywords:** Optic Nerve Trauma And Treatment, Trauma, Optic Neuropathy, Neuroimaging, Visual Fields6

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 7

### Sensitivity of Magnetic Resonance Imaging in Acute Demyelinating Optic Neuritis

Lulu LCD Bursztyn<sup>1</sup>, Lindsey B De Lott<sup>1,2</sup>, Wayne C Cornblath<sup>1,2</sup>

<sup>1</sup>University of Michigan, Department of Ophthalmology and Visual Science, Ann Arbor, MI, USA, <sup>2</sup>University of Michigan, Department of Neurology, Ann Arbor, MI, USA

#### Introduction:

Magnetic resonance imaging (MRI) of the orbits is often used to confirm a diagnosis of optic neuritis when clinical findings are uncertain. The sensitivity of MRI in optic neuritis (ON) has not been well studied but has been estimated to be approximately 80-90%.<sup>1,2</sup> The aim of this study was to determine the sensitivity of MRI findings in acute demyelinating ON.

#### Methods:

We retrospectively reviewed the charts of all patients with ON diagnosed using ONTT criteria seen for a first or follow-up visit in the last 2 years in a single neuro-ophthalmology practice. Data abstracted included: age at diagnosis, sex, initial and final Snellen visual acuity, visual field mean deviation, optic disc edema, optic nerve abnormalities on MRI (none, high T2 signal, post-contrast enhancement, or enlargement), presence of 1 or more high T2-signal lesions on brain MRI and diagnosis of multiple sclerosis during the follow-up period. Descriptive statistics were used to describe the distribution of all variables and calculate the sensitivity of orbital MRI findings individually and in aggregate for ON.

#### Results:

Thirty patients met all inclusion criteria and 22 (73%) had at least one MRI feature consistent with ON. There was no difference in age, visual function, optic disc edema, or delay from symptom onset to MRI in patients with and without findings on MRI. Patients with MRI features of ON were more likely to have white matter lesions on MRI (55% vs 25%) and have an eventual diagnosis of multiple sclerosis (27% vs 0%) over a mean follow-up period of 3.1 years (range 0.4 to 8.7 years).

#### Conclusions:

The sensitivity of MRI in acute optic neuritis (73%) is lower than estimated. Optic neuritis without MRI features consistent with ON may have a lower risk of MS. We are reviewing additional cases to confirm these results.

#### References:

1. Miller, Newton, van der Poel et al. Magnetic resonance imaging of the optic nerve in optic neuritis. *Magn Reson Med*, 38,175–179, 1988.
2. McKinney, Lohman, Sarikaya, Benson, Lee and Benson. Accuracy of routine fat-suppressed FLAIR and diffusion-weighted images in detecting clinically evident acute optic neuritis. *Acta Radiol*, 54, 455-461, 2013.

**Keywords:** Optic Neuropathy, Neuroimaging, Demyelinating Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 8

### Characterization of Leber's Hereditary Optic Neuropathy Patients Treated with Idebenone

Jasdeep S Chahal<sup>1</sup>, Michael J Ammar<sup>1</sup>, Rustum Karanjia<sup>2</sup>, Alfredo Sadun<sup>2</sup>

<sup>1</sup>USC/Keck School of Medicine, Los Angeles, CA, USA, <sup>2</sup>UCLA/Doheny Eye Institute, Los Angeles, CA, USA

#### Introduction:

There is conflicting data regarding the efficacy of Idebenone as a treatment for Leber's Hereditary Optic Neuropathy (LHON) [1, 2]. Klopstock et al. did not find a significant increase in recovery of BCVA in patients treated with Idebenone while Carelli et al. did (from off-chart (<20/400) to on-chart), specifically in the mutation 11778. Our aim was to determine if there was an improvement in vision even if patients stayed off-chart.

#### Methods:

A retrospective chart review was done looking at all patients who visited a tertiary care eye clinic from 2009 until June 2014 and were diagnosed with LHON. Duration of disease, visual acuity (VA) and mean deviation (MD) were recorded for each visit. Counting fingers was estimated in decimal acuity by calculating the numbers of minutes of arc subtended by 3 fingers, which would represent the letter E. VA was assessed to be off-chart at the beginning of therapy, 6 months of therapy, 1 year, and at the end of therapy.

#### Results:

70 patients were identified with LHON--16 of these were treated with Idebenone for greater than a year and seen within 3 months of start of treatment (12 with 11778, 3 with 14484, 1 with 3460). Patients with 11778 showed a significant improvement in VA at the end of therapy, however this effect took longer than 1 year to manifest. There was no significant difference in VA and MD between right and left eyes at 1 year past conversion. VA improved from 20/1676  $\pm$  2105 to 20/829  $\pm$  920 ( $p=0.04$ ,  $n=24$  eyes) at end of therapy. 11/24 eyes were off-chart at the beginning of therapy, 13/24 eyes at 6 months, 14/24 eyes at 1 year and 16/24 eyes were off-chart at the end of therapy.

#### Conclusions:

Idebenone therapy can improve VA in mutation 11778, even if VA remains off-chart.

#### References:

1. Carelli, V., et al., Idebenone treatment in Leber's hereditary optic neuropathy. Brain, 2011. 134(Pt 9): p. e188.
2. Klopstock, T., et al., A randomized placebo-controlled trial of idebenone in Leber's hereditary optic neuropathy. Brain, 2011. 134(Pt 9): p. 2677-86.

**Keywords:** Genetic Disease, Optic Neuropathy, Visual Fields, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 9

### **Gliosarcoma of the Optic Nerves 15 years After Radiation Treatment For Hypophyseal Adenoma**

Ayşe I. Colpak<sup>1</sup>, Kamil Oge<sup>2</sup>, Kader Karlı-Oguz<sup>3</sup>, Ilkay Isikay<sup>2</sup>, Figen Soylemezoglu<sup>4</sup>, Tulay Kansu<sup>1</sup>

<sup>1</sup>Hacettepe University School of Medicine, Department of Neurology, Ankara, Turkey, <sup>2</sup>Hacettepe University School of Medicine, Department of Neurosurgery, Ankara, Turkey, <sup>3</sup>Hacettepe University School of Medicine, Department of Radiology, Ankara, Turkey, <sup>4</sup>Hacettepe University School of Medicine, Department of Pathology Ankara, Turkey

#### **Introduction:**

Gliosarcoma is a very rare mixed tumor in the central nervous system, consisting of glial and malignant mesenchymal elements. Gliosarcoma is considered a subtype of glioblastoma and termed as primary gliosarcoma. Secondary gliosarcoma is detected at subsequent surgery for previously resected and irradiated glioblastoma multiforme. We describe an unusual case with vision loss due to gliosarcoma, probably radiotherapy-induced, 15 years after radiation treatment for hypophyseal adenoma.

#### **Methods:**

Single case report

#### **Results:**

A 56 year old woman with a past history of pituitary adenoma and thyroid micropapillary carcinoma presented with bilateral progressive loss of vision over a year. She had undergone surgical resection of the hypophyseal adenoma followed by fractionated external beam radiation with a total dose of under 50 Gy, 15 years prior to her presentation. At the time of her first symptom visual acuities were 20/20 OD and counting fingers OS. Fundoscopy demonstrated an optic atrophy of OS with a normal fundus OD. Brain MRI revealed T2 hyperintensity, mild thickening with enhancement of the chiasm and left optic nerve. The patient was considered to have radiation induced optic neuropathy and underwent hyperbaric oxygen therapy, systemic corticosteroids and intravenous bevacizumab. Over the following months her vision is deteriorated to NLP OU. Repeated MRI showed progressive T2 hyperintensity and severely enlarged optic nerves, chiasm and optic tracts with patchy enhancement, suggestive of a high grade optic pathway glioma. Histopathological examination following excisional biopsy and debulking of the tumor revealed pleomorphic spindle cells with negative staining for glial fibrillary acidic protein (GFAP) and S-100 with GFAP and S-100 positive atypical cells consistent with gliosarcoma.

#### **Conclusions:**

Radiation induced gliosarcomas are very rare and to our knowledge this is the first report of gliosarcoma of the optic pathways. This case provides an insight for the carcinogenic effect of radiation therapy on intracranial tumors.

**References:** None.

**Keywords:** Optic Neuropathy, Radiation Injury, Gliosarcoma, Optic Pathway Glioma

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 10

### Optic Nerve Compression by an Anomalous Internal Carotid Artery

Valerie I Elmalem<sup>1</sup>, Michael Dattilo

*SUNY Downstate Medical Center, Department of Ophthalmology, Brooklyn, NY, USA*

#### Introduction:

Although there are several etiologies for optic atrophy, compression by intracranial masses or aneurysms is of particular concern. One infrequently reported cause of compressive optic neuropathy (CON) is compression of the intracranial portion of the optic nerve by an anomalous or ectatic internal carotid artery (ICA).

#### Methods:

Here we report three cases of CON caused by an anomalous ICA identified by magnetic resonance imaging (MRI).

#### Results:

The three patients presented in this series range in age from 27 to 71. All patients initially presented with decreased vision, a relative afferent pupillary defect (RAPD), and visual field deficits. Dilated funduscopic examination (DFE) revealed asymmetric cupping of the optic nerve with pallor in excess of cupping in the affected eye of all patients. Visual field deficits in the affected eye included constriction with a small central island, junctional scotoma, and arcuate defects. Two of the three patients were being treated for presumed glaucoma. Laboratory evaluation for other etiologies of optic neuropathy was negative. MRI brain and orbits was significant for close apposition with displacement or compression of the optic nerve by the intracranial portion of the ICA. The youngest of the three patients had progressive profound visual loss with chiasmal involvement and has been referred for neurosurgical evaluation.

#### Conclusions:

These cases add to the small number of reported patients with optic atrophy caused by close apposition/compression of the optic nerve by the internal carotid artery. Though a seemingly rare occurrence, this is an important, potentially treatable cause of optic atrophy. Often mistakenly presumed to have progressive glaucoma, this diagnosis should be considered in patients with marked asymmetry or in patients with progression of visual dysfunction despite adequate treatment with medications to lower the intraocular pressure. Although controversial, surgical decompression may be considered in selected cases.

#### References:

1. Colapinto EV, Cabeen MA, Johnson LN. Optic nerve compression by a dolichoectatic internal carotid artery: case report. *Neurosurgery*, 39(3), 604-606, 1996.
2. Fagen KM, Blackburn S. Surgical decompression for optic neuropathy from carotid artery ectasia: Case report with technical considerations. *World Neurosurg*, 82(1-2), 239, e9-e12, 2014.
3. Ishikawa T, Ito T, et al. Compressive optic nerve atrophy resulting from a distorted internal carotid artery. *Pediatr Neurol*, 22(4), 322-324, 2000.
4. Matsuo K., Kobayashi S., Sugita K. Bitemporal hemianopsia associated with sclerosis of the intracranial carotid arteries. Case report. *J Neurosurg*, 53(4), 566-569, 1980.
5. Storm R.G., Fouladvand M., et al. Progressive optic neuropathy caused by contact with the carotid artery: Improvement after microvascular decompression. *Clin Neurol Neurosurg*, 114(6), 812-815, 2012.

**Keywords:** Optic Neuropathy, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 11

### Identifying Risk Factors for the Development of Radiation Optic Neuropathy at “Safe” Doses: A Review of Cases Seen 1994-2014

Pavle Doroslovački<sup>1</sup>, Grant T. Liu<sup>2</sup>, Madhura A. Tamhankar<sup>1</sup>, Kenneth S. Shindler<sup>1</sup>, Michelle Alonso-Basanta<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, Scheie Eye Institute, University of Pennsylvania, Philadelphia, PA, USA, <sup>2</sup>Neuro-Ophthalmology Service, Department of Neurology, University of Pennsylvania, Philadelphia, PA, USA, <sup>3</sup>Department of Radiation Oncology, University of Pennsylvania, Philadelphia, PA, USA

#### Introduction:

Radiation optic neuropathy (RON) is an uncommon but often visually devastating complication caused by radiation therapy to structures near the anterior visual pathways. Published reports have identified a radiation dose of less than 55-60 Gy,<sup>1,2</sup> or 8 Gy for stereotactic radiosurgery,<sup>3</sup> below which RON is unlikely to occur. Having observed multiple cases of RON occurring at doses below these thresholds, we attempted to identify potential risk factors for the development of RON in these patients.

#### Methods:

Retrospective chart review of patients with the diagnosis of radiation optic neuropathy seen between 1994 and 2014.

#### Results:

Thirteen patients with 18 affected eyes were included in the study. The radiation modalities included external beam radiation (N=5), whole brain radiation (N=2), stereotactic radiosurgery (N=2), proton beam (N=3), and unknown (N=1). For patients with known target tissue doses (N=10), the mean dose ( $\pm$ standard deviation) was 50.2 $\pm$ 13 Gy, with only 2/10 patients receiving  $\geq$ 55 Gy. For eyes with available follow up (mean follow up: 15 months), 4/14 were NLP (29%), 6/14 (43%) were 20/200-HM, 2/14 (14%) were 20/50-20/200, and 2/14 (14%) were 20/40 or better, at the last visit. Of the patients who received a target tissue dose of  $<$ 55 Gy, 6/8 had vasculopathic risk factors, such as a past history of heavy smoking, hypertension, and hyperlipidemia. Multiple treatment modalities were attempted, including steroids, anticoagulation, hyperbaric oxygen, bevacizumab, and blood viscosity reduction, with no improvement in vision in the majority of eyes (10/14), although a minority showed relative stability in visual acuity (4/14).

#### Conclusions:

RON can occur with different radiation modalities and can also occur at target tissue doses often thought to be within “safe” levels ( $<$ 55 Gy). Vasculopathic risk factors may increase the risk of RON, leading to occurrence at lower doses. Prognosis continues to be poor, with no clearly effective treatment modality.

#### References:

1. Mayo C, et al. Radiation dose–volume effects of optic nerves and chiasm. *Int J Radiation Oncology Biol Physics*. 2010; 76(3): S28-S35.
2. Parsons JT, et al. Radiation optic neuropathy after megavoltage external-beam irradiation: analysis of time-dose factors. *Int J Radiation Oncology Biol Physics*. 1994; 30(4): 755-63.
3. Girkin, CA, et al. Radiation optic neuropathy after stereotactic radiosurgery. *Ophthalmology*. 1997; 104(10):1634-43.

**Keywords:** Chemotherapy And Radiation Injury, Optic Neuropathy, Neuro-Ophth & Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 12

### Optic Neuritis After Refractive Surgery: Causal Or Coincidence?

Shlomo Dotan<sup>1</sup>, Gala Beykin, Denise Wajnsztajn, Joseph Frucht-Pery

*Hadassah-Hebrew University Medical Center, Department of Ophthalmology, Jerusalem, Israel*

#### Introduction:

Although the number of laser-assisted in situ keratomileusis (LASIK) and photorefractive keratectomy (PRK) procedures performed is growing steadily, there are no reports in the literature on associated optic neuritis.

#### Methods:

In this retrospective observational case series, we report three patients who developed acute visual loss following refractive surgery. All three underwent complete eye examinations with detailed evaluation of the optic nerve, Humphrey 24-2 SITA visual field testing, retinal nerve fiber layer thickness using optical coherence tomography (OCT) instrument, serologic evaluation, and magnetic resonance imaging of the brain and orbits.

#### Results:

Two patients underwent LASIK and one PRK. All had clinical evidence of optic neuritis, manifested by a subjective decrease in visual acuity and visual field abnormality, and relative afferent pupillary defect. Two had normal-appearing optic discs and one had optic disc edema initially. Two of the patients had remarkable recovery of visual function within two weeks, while one showed no signs of recovery and developed optic atrophy in the affected eye.

#### Conclusions:

Optic neuropathy has been previously reported after LASIK surgery due to barotrauma or ischemia, related to extreme elevation of intraocular pressure that occurs during a portion of the procedure. There are no reports in the literature on other etiologies for optic nerve disorders associated with LASIK and PRK. Anxiety could be considered a plausible trigger of optic neuritis after refractive surgery.

#### References:

1. Cameron, Saffra, Strominger: Laser in situ keratomileusis-induced optic neuropathy. *Ophthalmology*, 108, 660, 2001.
2. Ackerman, Heyman, Rabin, Anderson, Houck et al.: Stressful life events precede exacerbations of multiple sclerosis. *Psychosomatic Medicine*, 64, 916, 2002.

**Keywords:** Optic Neuritis, Visual Field Testing, Diagnostic Tests, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 13

### Long Term Treatment of Lebers Hereditary Optic Neuropathy With Idebenone.

Jennifer I Doyle<sup>1</sup>, Brendan D Grondines<sup>1</sup>, Michael S Vaphiades<sup>1,2,3</sup>

<sup>1</sup>University of Alabama at Birmingham, Dept. of Ophthalmology, Birmingham, AL, USA, <sup>2</sup>University of Alabama at Birmingham, Dept. of Neurology, Birmingham, AL, USA, <sup>3</sup>University of Alabama at Birmingham, Dept. of Neurosurgery, Birmingham, AL, USA

#### Introduction:

Lebers hereditary optic neuropathy (LHON) has classically been viewed as a non-treatable disease. However, recently there have been several publications, including case reports, retrospective and prospective studies, which document visual improvement with the use of Idebenone. We report a case of Lebers hereditary optic neuropathy treated with Idebenone that showed significant visual recovery.

#### Methods:

A case report.

#### Results:

A 47 year old white male presented with visual loss of unknown etiology that was determined to be LHON mutation 11778. Vision on presentation was 20/400 OD and HM OS. Color vision was 4/8 OD and 0/8 OS. He was placed on 900mg of Idebenone QD in September 2012 and followed. At recent visit in October 2014, his best corrected vision had improved to 20/25 OD and 20/200 OS with 7.5/8 color vision OU. His VEP shows improvement consistent with exam.

#### Conclusions:

This case supports the treatment of long term Idebenone alone for LHON and documents VEP improvement in this setting. The retrospective case series on Idebenone treatment had variable treatment protocols. The prospective case series studied patients on treatment for only six months. Another case report shows concomitant use of Idebenone with IV steroids and oral coenzyme Q. Our patient received only oral Idebenone after diagnosis and has been on it for 24 months with significant improvement.

#### References:

1. Sabet-Peyman EJ1, Khaderi KR, Sadun AA. Is Leber hereditary optic neuropathy treatable? Encouraging results with idebenone in both prospective and retrospective trials and an illustrative case. *J Neuroophthalmol*. 2012 Mar;32(1):54-7.
2. Klopstock T1, Yu-Wai-Man P, Dimitriadis K, et al. A randomized placebo-controlled trial of idebenone in Leber's hereditary optic neuropathy. *Brain*. 2011 Sep;134(Pt 9):2677-86.
3. La Morgia C1, Carbonelli M2, Barboni P3, Sadun AA4, Carelli V1. Medical management of hereditary optic neuropathies. *Front Neurol*. 2014 Jul 31;5:141

**Keywords:** Genetic Disease, Visual Fields, Diagnostic Testing

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** The Eye Sight Foundation of Birmingham, AL

## Poster 14

### Macular star formation in diabetic patients with non-arteritic anterior ischemic optic neuropathy (NA-AION)

Alberto Galvez-Ruiz<sup>1,2</sup>, Oriol Franch<sup>2</sup>

<sup>1</sup>King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia, <sup>2</sup>Hospital Ruber Internacional, Madrid, Spain

#### Introduction:

NA-AION is a condition that exhibits a number of unique characteristics in diabetics compared with the rest of the population. In some diabetic patients with NA-AION, lipid deposits can be observed around the macula forming an incomplete macular star.

#### Methods:

We describe 12 case studies of patients with NA-AION observing the development of lipid deposits around the macula forming an incomplete macular star.

#### Results:

All our patients developed some level of lipid deposits around the macula in the form of a macular hemistar in the course of their illness.

#### Conclusions:

Some authors have suggested that the macular star is formed by transudation from capillaries deep in the optic disc through the intermediary tissue of Kuhnt, which is located between the retina and the anterior portion of the lamina retinalis. However, the development of the macular star is currently understood not as a simple transudation but as a multifactorial process involving the presence of vascular damage around the optic disc, which is considered one of the most important factors leading to its occurrence. Although some studies mention the presence of a macular star in patients with NA-AION, we believe that this phenomenon may be significantly more common than the current literature suggests.

#### References:

1. Hayreh SS, Zimmerman MB. Nonarteritic anterior ischemic optic neuropathy: clinical characteristics in diabetic patients versus nondiabetic patients. *Ophthalmology*. 2008 Oct; 115(10):1818-25.
2. Chen T, Song D, Shan G, Wang K, Wang Y, Ma J, Zhong Y. The association between diabetes mellitus and nonarteritic anterior ischemic optic neuropathy: a systematic review and meta-analysis. *PLoS One*. 2013 Sep 30;8(9):e76653.
3. Lee MS, Grossman D, Arnold AC, Sloan FA. Incidence of nonarteritic anterior ischemic optic neuropathy: increased risk among diabetic patients. *Ophthalmology*. 2011 May;118(5):959-63.
4. Hayreh SS. Ischemic optic neuropathy. *Prog Retin Eye Res*. 2009 Jan; 28(1):34-62.
5. Hayreh SS. Role of retinal hypoxia in diabetic macular edema: a new concept. *Graefes Arch Clin Exp Ophthalmol*. 2008 Mar; 246(3):353-61.

**Keywords:** Lipid Deposits, Neuroretinitis, Non-Arteritic Anterior Ischemic Optic Neuropathy, Macular Hemistar, Diabetes Mellitus

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 15

### Seasonal Influence on the Incidence of Biopsy-Proven Giant Cell Arteritis: the University of California Davis Institutional Experience

Kimberly K Gokoffski<sup>1</sup>, Syed K Khaderi

*University of California Davis Department of Ophthalmology and Vision Science, Sacramento, CA, USA*

#### **Introduction:**

Giant cell arteritis (GCA) is a vasculitis that affects large and medium sized arteries. The etiology of GCA is unknown and numerous risk factors have been proposed. We and other investigators have perceived an increased incidence over the summer months and hypothesized that this may represent an unrecognized risk factor for the disease.

#### **Methods:**

We performed a retrospective review based on billing codes of temporal artery biopsies performed at the University of California, Davis from January 2000 to June 2014. We recorded patient demographics, onset and nature of presenting symptoms, date of biopsy and, when possible, visual acuity and lab values.

#### **Results:**

We identified 175 biopsies (119 female, 56 male) during our time period. Of these, 21 positive biopsies were female while 9 were male. Although twice as many biopsies were performed on women, women were three times more likely to have a positive biopsy. Positive biopsies were significantly more likely to occur in the summer months (June through August) than the rest of the year ( $p=0.039$ ). Patients with a positive biopsy averaged  $76 \pm 9$  years of age.

#### **Conclusions:**

Our retrospective study is the first report of the seasonal incidence of biopsy-proven GCA in California. Our data suggests that increased age, female gender and summer months are risk factors for developing biopsy-proven GCA in our region. One plausible hypothesis to explain this finding is that GCA may be due to an infectious agent, such as CMV, parvovirus or Mycoplasma. Our data support this hypothesis, however, to date no infectious agents have consistently been isolated from positive biopsies.

**References:** None.

**Keywords:** Giant Cell Arteritis, Seasonal Incidence, Biopsy-Proven, California

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 16

### Possible Revatio (Sildenafil) Induced Optic Neuropathy in Mice

Nitza Goldenberg-Cohen<sup>1,3,4</sup>, Mark Vieyra<sup>4</sup>, Orkun Muhsinoglu<sup>1</sup>, Shirel Weiss<sup>1,4</sup>, David Zadok<sup>2,4</sup>, James D. Nicholson<sup>1,4</sup>

<sup>1</sup>The Krieger Eye Research Laboratory, Felsenstein Medical Research Center, Petach Tikva, Israel, <sup>2</sup>Ophthalmology Department, Assaf Harofeh, Zerifin, Israel, <sup>3</sup>Pediatric Unit, Ophthalmology Department, Schneider Children's Medical Center of Israel, Petach Tikva, Israel, <sup>4</sup>Sackler Faculty of Medicine, Tel Aviv University Tel Aviv, Israel

#### Introduction:

Purpose: To investigate possible Revatio (sildenafil) induced optic neuropathy in mice.

#### Methods:

Right optic nerve crush (ONC) was induced in 53 out of 134 mice used. Revatio was administered with and without ONC. In the ONC group, 27/53 received intravitreal (IVT) injection of Revatio (0.24µg/3µl) immediately before ONC induction and 26/53 mice received no treatment (saline). The left eyes served as a control. In remaining 81 mice without ONC induction, 21/81 were injected IVT and 60/81 intraperitoneally (IP, 24µg/300µl). FA was performed (day 0). Quantitative real-time PCR was used to quantify optic nerve apoptosis-related, oxidative-stress and ischemic-related gene expression on days 1 and 3. Retinal and optic nerve histology was examined on days 14 and 21 using flat mount, H&E, 2,3,5-Triphenyltetrazolium chloride (TTC) and luxol fast blue (LFB).

#### Results:

Maximal retinal vessels dilatation and increased choroidal effusion were detected by FA immediately after IVT Revatio injection, and 30 minutes after IP injection. At 21 days following ONC and IVT Revatio, RGCs were protected relative to ONC without treatment. In the Revatio injected mice without ONC induction, IVT produced no RGC loss on histology, or optic nerve stroke, at 14 or 21 days from the injection (n=10). However, molecular studies revealed 2 stroke candidates nerves (2/11) and increased Bcl-2 on day 1 which reverted to baseline at day 3. Following IP Revatio (n=40) 6 animals showed RGC loss and associated optic nerve damage histologically. All genes measured in the IP group without ONC (n=20), relating to apoptosis and oxidative-stress, decreased (<0.5 fold) on both days 1 and 3.

#### Conclusions:

Revatio increased choroidal perfusion and mildly dilated retinal vessels. Injection immediately before ONC, revealed a possible neuroprotective effect. Without ONC induction, possible occurrence of optic nerve stroke was observed, This might be associated with vessel dilatation and reperfusion, affecting optic nerve autoregulation.

**References:** None.

**Keywords:** Revatio (Sildenafil), Optic Neuropathy, Optic Nerve Crush, Mouse Model, Optic Nerve Stroke

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This study was supported in part by the Zanvyl and Isabelle Krieger Fund, Baltimore, MD (NGC).

## Poster 17

### Cyclosporine Induced Papilledema without Elevated Intracranial Pressure

Poorav J. Patel<sup>1</sup>, Sudha Mekala<sup>1</sup>, Frederick T. Fraunfelder<sup>2</sup>, William L. Hills<sup>2,3</sup>

<sup>1</sup>Oregon Health & Science University/School of Medicine, Portland, OR, USA, <sup>2</sup>Casey Eye Institute, Oregon Health & Science University/Ophthalmology, Portland, OR, USA, <sup>3</sup>Oregon Health & Science University/Neurology, Portland, OR, USA

#### Introduction:

Cyclosporine (CsA) is an immunosuppressant used to prevent renal transplant rejection. CsA has documented central nervous system toxicities including pseudotumor cerebri syndrome. We present a patient taking CsA for 20 years found to have papilledema without increased intracranial pressure, which resolved after discontinuation.

#### Methods:

Case report and literature review including data query of the World Health Organization Uppsala Monitoring Centre's medication related ocular toxicities.

#### Results:

A 48 year-old male with history of cadaveric kidney transplant August 26, 1991 due to chronic glomerulonephritis, was found to have papilledema on routine eye exam for blurred vision at near only. He denied transient visual obscurations, diplopia, pulse synchronous tinnitus, snoring, or weight change. Medications included azathioprine 150 mg daily and cyclosporine 175 mg twice daily. Neuro-ophthalmic examination revealed distance visual acuity without correction right eye 20/20-1, left eye pinhole 20/25+2. The remainder of his neuro-ophthalmic examination was normal with exception of bilaterally enlarged blind spots with superior arcuate depression both eyes and severe Frisén grade V papilledema. MRI brain was normal. Lumbar puncture found an opening pressure of 15 cm H<sub>2</sub>O and normal cerebral spinal fluid. Renal function, blood pressure and BMI were normal. Cyclosporine was discontinued with improved vision left eye and gradual resolution of bilateral disc edema over the subsequent 9 months. Query of the WHO database found 59 cases of papilledema with mean onset of 255 days and max of 4 years.

#### Conclusions:

Cyclosporine is a neutral, lipophilic, cyclic undecapeptide derived from the fungus *Trichoderma polysporin*, primarily used as an immunosuppressant metabolized by the hepatic cytochrome P-450 enzymes. Current research suggests that cyclosporine modifies mitochondrial structure and function through reactive oxygen species (ROS) which can induce obstructed axoplasmic transport and pseudo-papilledema. We present a rare case of papilledema without increased intracranial pressure due to CsA use after 20 yrs

#### References:

1. Avery, R, D A Jabs, J R Wingard, G Vogelsang, R Saral, and G Santos. 1991. "Optic Disc Edema after Bone Marrow Transplantation. Possible Role of Cyclosporine Toxicity." *Ophthalmology* 98 (8): 1294–1301.
2. Coskuncan, N M, D A Jabs, J P Dunn, J A Haller, W R Green, G B Vogelsang, and G W Santos. 1994. "The Eye in Bone Marrow Transplantation. VI. Retinal Complications." *Archives of Ophthalmology* 112 (3): 372–79.
3. Costa, Kellen Micheline A H, José Bruno de Almeida, Ricardo Humberto de M Félix, and Maurício Ferreira da Silva Júnior. 2010. "[Pseudotumor Cerebri Associated with Cyclosporin Use Following Renal Transplantation]." *Jornal Brasileiro de Nefrologia: 'órgão Oficial de Sociedades Brasileira E Latino-Americana de Nefrologia* 32 (1): 136–39.
4. De Arriba, Gabriel, Miryam Calvino, Selma Benito, and Trinidad Parra. 2013. "Cyclosporine A-Induced Apoptosis in Renal Tubular Cells Is Related to Oxidative Damage and Mitochondrial Fission." *Toxicology Letters* 218 (1): 30–38. doi:10.1016/j.toxlet.2013.01.007.
5. Katz, B. 1997. "Disk Edema Subsequent to Renal Transplantation." *Survey of Ophthalmology* 41 (4): 315–20. doi:10.1016/S0039-6257(96)00006-9.

**Keywords:** Cyclosporine, Toxicity, Papilledema, Pseudotumor Cerebri, Immunosuppression

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Research to Prevent Blindness, unrestricted grant

## Poster 18

### Asymptomatic Leukemic Optic Nerve Infiltration as Presentation of Acute Lymphoblastic Leukemia Relapse

Florin Grigorian<sup>1</sup>, Ang Li<sup>2</sup>, Adriana P Grigorian<sup>1</sup>

<sup>1</sup>University Hospitals Eye Institute, Cleveland, OH, USA, <sup>2</sup>Case Western Reserve Medical School, Cleveland, OH, USA

#### Introduction:

We present a 4 year old boy with history of CNS acute lymphoblastic leukemia (ALL) in remission, diagnosed with leukemic optic neuropathy at a routine eye exam.

#### Methods:

Vision was equal in both eyes, measuring 20/30 by Allen, considered normal given the age limitations. Color vision (HRR) was normal and equal in both eyes and pupils were equally reactive to light and accommodation without relative afferent papillary defect (RAPD). Fundus examination was normal in the right eye and revealed a white mass covering the left optic disk, completely obscuring it and peripapillar vascular sheathing. The rest of the fundus was normal.

#### Results:

The MRI showed no enhancement of the optic nerves or brain tissue. There was a questionable diffusion restriction of the left optic nerve head. The peripheral blood smear showed 1% blasts (5% diagnostic for relapse) and the CSF revealed rare cells suspicious of blasts. Chemotherapy was started (Methotrexate and Vincristine). No ocular radiation was performed due to the asymptomatic nature of the condition. At 3 weeks follow up the vision exam was the same and the mass covering the left optic nerve was replaced by mild gliosis with good visualization of the optic nerve head. The fluorescein angiography did not demonstrate any defects. The CSF pathology later revealed blasts as the dominant cells and the bone marrow biopsy revealed hypercellularity confirming the relapse.

#### Conclusions:

Tumoral optic nerve infiltration may be the first sign of ALL relapse. In our case the vision, color vision and pupillary reflexes were normal and maintained throughout the evolution of the disease. Of note, the absence of enhancement on MRI does not exclude neoplastic infiltration. We recommend routine ophthalmological exam for all patients with history of ALL to exclude optic nerve involvement without systemic symptoms or signs.

#### References:

1. Brown, G C, Shields, J A, Augsburger, J J, Serota, F T, K. P. (1981). Leukemic optic neuropathy. International ophthalmology, 3(2), 111–116
2. Kincaid MC, Green WR. Ocular and orbital involvement in leukemia. Surv. Ophthalmol. 1983;27(4):211–32
3. Rosenthal AR, Egbert PR, Wilbur JR, Probert JC. Leukemic involvement of the optic nerve. Journal of Pediatric Ophthalmology.:1975; 12(2):84-93.
3. Bandyopadhyay S, Das D, Das G, Gayen S. Unilateral optic nerve infiltration as an initial site of relapse of acute lymphoblastic leukemia in remission. Oman J Ophthalmol 2010;3:153-4

**Keywords:** Infiltrative Neuropathy, Leukemia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 19

### Progressive Visual Loss: An Unusual Presenting Symptom In Giant Cell Arteritis

Parima Hirunwiwatkul<sup>1,2</sup>, Supanut Apinyawasisuk<sup>2</sup>

<sup>1</sup>Department of Ophthalmology, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand, <sup>2</sup>Ophthalmology department, King Chulalongkorn Memorial Hospital, Bangkok, Thailand

#### Introduction:

Arteritic anterior ischemic optic neuropathy (AAION) due to Giant cell arteritis (GCA) is a rare cause of visual loss in Asian people. However, there are a few AAION cases in Asian that have been reported. In general, patient with AAION usually presented with acute severe visual loss with disc edema and systemic presentation of GCA.

#### Methods:

Case report

#### Results:

We herein report a case of 77-year-old Thai man who presented with gradual painless visual loss in his right eye for 2 weeks. His vision progressively deteriorated from 20/40 to counting finger. Fundus examination revealed chalky disc swelling at the same time as when the visual loss started. His associated systemic symptoms were weight loss and limb claudication without scalp tenderness or jaw claudication. On examination, a cord-like lesion of right temporal artery with absence of pulsation was found. An extensive investigation for a definite diagnosis was attempted. The diagnosis of GCA was finally made due to the pathognomonic histopathology result of the temporal artery biopsy. After treatment, his vision slightly improved and stabilized. Limb claudication also improved.

#### Conclusions:

Due to the unusual presentation, a misdiagnosis may be made in this case. The differences between Caucasian and Asian AAION was reviewed. The temporal artery pulsation, disc character and elevated levels of inflammatory markers are the clues for the diagnosis of GCA. The temporal artery biopsy was the definite investigation for his diagnosis.

**References:** None.

**Keywords:** Optic Neuropathy, Giant Cell Arteritis, Temporal Arteritis, Progressive Visual Loss, Ischemic Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 20

### Demyelinating Disorder with Optic Neuropathies in a Patient with Monoclonal Gammopathy: Case Report and Literature Review

Whitney B. Hough<sup>1</sup>, Matthew J. Khayata<sup>1</sup>, Charles G. Maitland<sup>1,2</sup>

<sup>1</sup>Florida State University College of Medicine, Tallahassee, FL, USA, <sup>2</sup>TMH Foundation Healthcare, Tallahassee, FL, USA

#### Introduction:

Neurologic involvement in monoclonal gammopathy typically consists of demyelinating/axonal peripheral neuropathies with many features in common with chronic inflammatory polyneuritis. Evidence of CNS involvement is occasionally present. Clinical findings reported include nystagmus, ataxia, dysarthria, and extensor plantar responses. MRI scans typically show multiple white matter lesions, similar findings are reported in Chronic Inflammatory Demyelinating Polyneuropathy (CIPD), and similar to MS (1). CSF findings are generally acellular with occasionally elevated protein, oligoclonal bands are typically absent (2).

#### Methods:

A 55 yo woman complained of blurred vision, slurred speech and unsteadiness with falling over several months. PMH was remarkable only for Benign Positional Vertigo, obesity and hypertension. Examination showed corrected VA of 20/30, 20/50; HRR 4/6 OU, mild disc head pallor on fundoscopy, binocular arcuate defects on Humphrey's 24.2 visual fields collaborated by spectral OCT scans. Neurologic revealed generalized hyperreflexia, wide based unsteady gait, positive Romberg with substandard limb coordination but preserved primary sensory modalities.

#### Results:

CBC and metabolic panels, VDRL, ACE and B12/folate levels unremarkable. ESR elevated at 68mm. MRI brain showed multiple subcortical white matter lesions; CSF analysis demonstrated acellular fluid, normal chemistries, but elevated monoclonal gamma spike matched by elevated IgM kappa light chain in serum. Oligoclonal bands were absent.

#### Conclusions:

In this case symptoms, signs, and MRI scans were consistent with CNS demyelination, as reported in other cases of monoclonal gammopathies. The absence of oligoclonal bands (present in 98.5% of MS patients) and matching monoclonal spikes in serum and in CNS suggest the gammopathy is causal. Monoclonal gammopathy should be added to the list of uncommon causes of afferent visual pathway defects.

#### References:

1. Hampel, Schneider, Hock, Muller-Spahn, Ackenheil, CNS demyelination in monoclonal gammopathy of undetermined significance (MUGUS): possible cause of a dementia syndrome, Eur Psychiatry, 11, 46-49, 1996.
2. Lehmann, Hoffmann, Meyer zu Horste, Hartung, Kieseier, Central nervous system involvement in patients with monoclonal gammopathy and polyneuropathy, European Journal of Neurology, 17, 1075-1081, 2010.

**Keywords:** Optic Neuropathy, Monoclonal Gammopathy Of Undetermined Significance (MGUS), CNS Demyelination

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 21

### Retrospective Review of Ophthalmic and Systemic Associations of Optic Nerve Hypoplasia

Imran Jivraj<sup>1</sup>, James Lewis

*University of Alberta Department of Ophthalmology, Edmonton, AB, Canada*

#### Introduction:

Optic Nerve Hypoplasia (ONH) is a leading cause of childhood blindness and visual impairment. ONH is characterized by an abnormally small or cupped optic disc with a subnormal number of axonal fibers. We sought to describe the prevalence of ophthalmic and non-ophthalmic associations of ONH in our practice.

#### Methods:

A retrospective review of an electronic database which spanned twenty years of a pediatric neuro-ophthalmologist's practice was completed and patients with ONH were identified. Charts were reviewed for sex, symmetry of ONH, coexisting ophthalmological and non-ophthalmological features, and neuroimaging findings.

#### Results:

Of the 340 patients with ONH, 46% were female and 54% were male. ONH was graded as mild (40%), moderate (31%), or severe (29%). ONH was bilateral and symmetrical in 43%, bilateral and asymmetrical in 33%, and unilateral in 24%. Atypical features of the optic nerve head were identified in 12.1% of patients which included cupping (5.3%), segmental truncation (2.9%), and a dysplastic appearance (3.8%). Other ophthalmological features included strabismus (68%) and nystagmus (51%). Coexisting ophthalmological conditions were identified, including cortical visual impairment (18.5%), high myopia (5.2%), and anisometropia (4.7%). Hypopituitarism was identified in 12.6% of patients. The most frequently identified structural brain abnormalities were absence of the septum pellucidum (15.3%), obstructive hydrocephalus (8.2%), and hypoplasia/aplasia of the corpus callosum (6.5%). There was a high prevalence of neurodevelopmental issues including developmental delay (66.8%), seizure disorder (23.2%), and cerebral palsy (19.1%). Genetic syndromes were identified in 12.1% of patients.

#### Conclusions:

While retrospective in nature, our study includes one of the largest groups of patients with ONH. Hypopituitarism was identified in 12.6% of cases of ONH which is lower than most reports [1, 2]. We identified a significant prevalence of strabismus, nystagmus, cortical visual impairment, and developmental delay among patients with ONH.

#### References:

1. Mohny, B.G., R.C. Young, and N. Diehl, Incidence and associated endocrine and neurologic abnormalities of optic nerve hypoplasia. *JAMA Ophthalmol*, 131(7): p. 898-902, 2013
2. Garcia-Filion, P. and M. Borchert, Optic nerve hypoplasia syndrome: a review of the epidemiology and clinical associations. *Curr Treat Options Neurol*, 15(1): p. 78-89, 2013

**Keywords:** Optic Nerve Hypoplasia, Neuro-Ophth & Systemic Disease, Pediatric Neuro-Ophthalmology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 22

### Optic Nerve Morphology as Marker for Disease Severity in Cerebral Palsy of Perinatal Origin

Sachin Kedar<sup>1</sup>, Deepta Ghate<sup>2</sup>, V Vedanarayanan<sup>3</sup>, James J Corbett<sup>3</sup>, Abdulbaset Kamour<sup>4</sup>

<sup>1</sup>University of Nebraska Medical Center and Truhlsen Eye Institute/Neurology and Ophthalmology, Omaha, NE, USA,

<sup>2</sup>University of Nebraska Medical Center and Truhlsen Eye Institute/Ophthalmology, Omaha, NE, USA, <sup>3</sup>University of Mississippi Medical Center/Neurology, Jackson, MS, USA, <sup>4</sup>University of Kentucky College of Public Health Lexington, KY, USA

#### Introduction:

Due to neuroplasticity, it is difficult to prognosticate the eventual neurologic outcome and ambulatory status from perinatal neurological insult till age 2-5 years. The objective of this study is to correlate ONH morphology (disc pallor and cupping) with period of gestation (POG) and severity of neurological damage in children with perinatal onset static encephalopathy (POSE).

#### Methods:

54 consecutive patients with POSE were enrolled. Exclusion criteria included genetic, metabolic or congenital structural brain abnormalities not related to perinatal complications; intraocular disease (ROP/glaucoma/cataract) and hydrocephalus. ONH morphology (pallor and cup to disc ratio-CDR) was assessed independently by 2 fellowship trained ophthalmologists by dilated direct and indirect ophthalmoscopic examination. ONH were labeled as pale or large cup (cup/disc ratio  $\geq 0.5$ ) only if the 2 ophthalmologists agreed. Inter-rater reliability was  $>0.8$  for all parameters. The pediatric neurologist determined eligibility, age of onset of POSE, neurological deficit and reviewed available neuroimaging.

#### Results:

Mean age was  $11.88 \pm 6.53$  years; period of gestation:  $33.26 \pm 4.78$  weeks. 33/54 (61%) showed ONH pallor or cupping. Of 17/54 patients with ONH pallor, 88% were quadriplegic and 82% non-ambulatory. Mean CDR was  $0.45 \pm 0.22$ ; 27/54 (50%) patients had large cup. Multivariate logistic regression models showed that disc pallor was significantly associated with non-ambulatory status (OR: 12.5;  $p=0.03$ ) and quadriplegia (OR: 21.7;  $p=0.0025$ ) and large cup was associated with age at examination (OR 1.15;  $p=0.025$ ). CDR and age at exam showed positive correlation ( $r=0.42$ ;  $p=0.002$ ). ONH parameters were not associated with POG.

#### Conclusions:

ONH changes are common in POSE and are not associated with POG as previously hypothesized. Optic disc pallor, a bedside clinical finding, can serve as a prognostic indicator for severe neurological insult in high risk children with perinatal complications that should prompt early referral for rehabilitation. Recognition of association of cupping with POSE will prevent unnecessary glaucoma examinations under anesthesia.

#### References:

1. Jacobson L, Hård AL, Svensson E, Flodmark O, Hellström A. Optic disc morphology may reveal timing of insult in children with periventricular leucomalacia and/or periventricular haemorrhage. Br J Ophthalmol. 2003;87(11):1345-9

**Keywords:** Optic Disc Pallor, Cerebral Palsy, Optic Neuropathy, Pediatric Neuro-Ophthalmology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 23

### Clinical Characteristics of Optic Neuritis Associated with Viral Infection

Sa Kang Kim<sup>1</sup>, Seong-Joon Kim

*Department of Ophthalmology, Seoul National University College of Medicine, Seoul, Korea*

#### Introduction:

Viral infection is well known to be associated with pediatric optic neuritis (ON) and some of adult ON. The optic neuritis study group reported that 26.1% of ON patients have a viral syndrome preceding visual loss. This study was conducted to characterize optic neuritis associated with viral infection.

#### Methods:

A retrospective chart review was performed on 124 patients with ON over 15 years old who visited a tertiary referral center in Korea from 2004 to 2014. Demographics, symptoms, signs and laboratory results were reviewed. Magnetic resonance images (MRI) and cerebrospinal fluid findings were also analyzed.

#### Results:

Seventeen out of 124 ON patients had medical history related to viral infection. There were 9 males (52.9%) and mean ( $\pm$ SD, range) age of onset was 36.07 ( $\pm$ 12.82, 18-64) years. Ten (76.9%) patients initially presented with poor visual acuity, which was defined by equal or worse than counting finger. Ocular pain or headache was accompanied in 82.4% of cases. Optic disc swelling was observed in 41.2% of the patients. Good visual outcome, equal or better than 20/40, was observed in 84.6%. Three patients had bilateral optic neuritis and three patients had recurrent optic neuritis. CSF analysis was evaluated in 12 patients and all patients had normal features. Antinuclear antibody was positive in 3 out of 12 patients. Among six patients who tested aquaporin-4 (AQP4) antibody, 2 patients showed positive result. Central scotoma was observed in 7 patients, and other variable field defects was also observed. Optic nerve enhancement in MRI was observed in 8 out of 15 cases.

#### Conclusions:

Optic neuritis associated with viral infection has distinct clinical profile compared with idiopathic optic neuritis.

**References:** None.

**Keywords:** Demyelinating Disease, Neuro-Ophth & Infectious Disease, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 24

### Methanol causes highly selective retinal ganglion cell layer loss and inner nuclear layer microcysts

Kendra A. Klein<sup>1</sup>, Thomas R. Hedges III, Carlos Medoza-Santiesteban

*New England Eye Center-Tufts Medical Center, Department of Ophthalmology, Boston, MA, USA*

#### Introduction:

Methanol in adulterated liquor can cause devastating metabolic acidosis, acute neurologic dysfunction, and vision loss by interfering with the mitochondrial respiratory chain.<sup>1,2,3,6</sup> Methanol differentially affects the small caliber axons of the papillomacular bundle.<sup>11</sup>

#### Methods:

A 19-year-old student developed severe systemic methanol toxicity and relatively mild optic neuropathy. Optical coherence tomography (OCT) showed unique changes.

#### Results:

A previously healthy woman developed ataxia, confusion, and difficulty breathing hours after drinking homemade alcohol in Indonesia. She was found to have a serum pH of 6.79 and elevated blood methanol which was treated with intravenous ethanol, sodium bicarbonate, and sodium bicarbonate. When she awoke she had blurred vision, photophobia, and central scotomas in both eyes. Her visual acuities were 20/30-1 OD and 20/15-1 OS. She had mild central depression on Humphrey visual field (HVF) testing and mild temporal pallor of the optic disc OS. OCT of the retinal nerve fiber layer (RNFL) was normal, but ganglion cell layer (GCL) analysis showed highly selective loss of the nasal fibers OU. Further, OCT of the maculas demonstrated inner nuclear layer (INL) microcysts OU in the corresponding areas of GCL loss.

#### Conclusions:

Selective involvement of the papillomacular bundle is common in toxic optic neuropathy. Prior OCT studies in methanol toxicity have shown diffuse RNFL thinning, typical of the widespread destruction of axons.<sup>5,12,6,7</sup> Retinal ganglion cell loss may be an early indicator of optic neuropathy.<sup>8</sup> The relative sparing of the optic nerve in this case shows the evolution of methanol toxicity with early segmental GCL involvement and preservation of the RNFL. INL microcysts have been observed in a variety of diseases, but have not been described in methanol optic neuropathy.<sup>4,9,10</sup> These findings support that INL microcysts are non-specific and may signify preferential loss of small caliber axons of the papillomacular bundle rich in mitochondria, which are most vulnerable in energy-depleted states.

#### References:

- (1) Brahmi, N., Blel, Y., Abidi, N., Kouraichi, N., Thabet, H., Hedhili, A., & Amamou, M. (2007). Methanol poisoning in Tunisia: report of 16 cases. *Clinical Toxicology (Philadelphia, Pa.)*, 45(6), 717–20. doi:10.1080/15563650701502600; (2) Desai, T., Sudhakar, A., Vyas, U., & Khamar, B. (2013). Methanol poisoning: predictors of visual outcomes. *JAMA Ophthalmology*, 131(3), 358–64. doi:10.1001/jamaophthalmol.2013.1463; (3) Gee, P., & Martin, E. (2012). Toxic cocktail: methanol poisoning in a tourist to Indonesia. *Emergency Medicine Australasia*: EMA, 24(4), 451–3. doi:10.1111/j.1742-6723.2012.01552.x; (4) Kaushik, M., Wang, C. Y., Barnett, M. H., Garrick, R., Parratt, J., Graham, S. L., ... Klistorner, A. (2013). Inner nuclear layer thickening is inversely proportional to retinal ganglion cell loss in optic neuritis. *PloS One*, 8(10), e78341. doi:10.1371/journal.pone.0078341; (5) Kim, Y.-K., & Hwang, J.-M. (2009). Serial retinal nerve fiber layer changes in patients with toxic optic neuropathy associated with antituberculosis pharmacotherapy. *Journal of Ocular Pharmacology and Therapeutics*: The Official Journal of the Association for Ocular Pharmacology and Therapeutics, 25(6), 531–5. doi:10.1089/jop.2009.0064; (6) Koehrer, P., Creuzot-Garcher, C., & Bron, A. M. (2011). Methanol poisoning: two case studies of blindness in Indonesia. *International Ophthalmology*, 31(6), 517–24. doi:10.1007/s10792-011-9492-2; (7) Martínez-López-Portillo, M. A., Martínez-Gamero, B. O., Mohamed-Noriega, J., Cavazos-Adame, M. H., & Mohamed-Hamsho, M. J. (2014). Behaviour of disc oedema during and after amiodarone optic neuropathy: case report. *Journal of Clinical and Diagnostic Research*: JCDR, 8(4), VD04–VD05. doi:10.7860/JCDR/2014/8254.4262; (8) Monteiro, M. L. R., Hokazono, K., Fernandes, D. B., Costa-Cunha, L. V. F., Sousa, R. M., Raza, A. S., ... Hood, D. C. (2014). Evaluation of inner retinal layers in eyes with temporal hemianopic visual loss from chiasmal compression using optical coherence tomography. *Investigative Ophthalmology & Visual Science*, 55(5), 3328–36. doi:10.1167/iops.14-14118; (9) Vanburen, J. M. (1963). Trans-Synaptic Retrograde Degeneration in the Visual System of Primates. *Journal of Neurology, Neurosurgery, and Psychiatry*, 26, 402–9. Retrieved from <http://www.pubmedcentral.nih.gov/articlerender.fcgi?artid=495606&tool=pmcentrez&rendertype=abstract>; (10) Wolff, B., Azar, G., Vasseur, V., Sahel, J.-A., Vignal, C., & Maugé-Fayssé, M. (2014). Microcystic changes in the retinal inner nuclear layer associated with optic atrophy: a prospective study. *Journal of Ophthalmology*, 2014, 395189. doi:10.1155/2014/395189; (11) Yu Wai Man, C. Y., Chinnery, P. F., & Griffiths, P. G. (2005). Optic neuropathies--importance of spatial distribution of mitochondria as well as function. *Medical Hypotheses*, 65(6), 1038–42. doi:10.1016/j.mehy.2004.10.021; (12) Zoumalan, Christoumalan, C. I., Agarwal, M., & Sadun, A. a. (2005). Optical coherence tomography can measure axonal loss in patients with ethambutol-induced optic neuropathy. *Graefes Archive for Clinical and Experimental Ophthalmology* = *Albrecht von Gr*, 410–6. doi:10.1007/s00417-004-1053-1; (13) Agarwal, M., & Sadun, A. a. (2005). Optical coherence tomography can measure axonal loss in patients with ethambutol-induced optic neuropathy. *Graefes Archive for Clinical and Experimental Ophthalmology* = *Albrecht von Graefes Archiv Für Klinische Und Experimentelle Ophthalmologie*, 243(5), 410–6. doi:10.1007/s00417-004-1053-1

**Keywords:** Methanol Toxicity, Microcysts, Ganglion Cell Layer, Optical Coherence Tomography, Papillomacular Bundle

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 25

### Parsing the differences between LHON affected: genetic vs environmental triggered disease

Valerio Carelli<sup>1,2</sup>, Pio d'Adamo<sup>3,4</sup>, Maria L Valentino<sup>1,2</sup>, Chiara La Morgia<sup>1,2</sup>, Fred N Ross-Cisneros<sup>5</sup>, Piero Barboni<sup>6,7</sup>, Rustum Karanjia<sup>5</sup>, Solange S Salomao<sup>8</sup>, Adriana Berezovsky<sup>8</sup>, Filipe Chicani<sup>8</sup>, Milton Moraes<sup>8</sup>, Milton Moraes-Filho<sup>8</sup>, Rubens Belford Jr<sup>8</sup>, Alfredo A Sadun<sup>5</sup>

<sup>1</sup>IRCCS Istituto delle Scienze Neurologiche di Bologna, Bologna, Italy, <sup>2</sup>Neurology unit, Department of Biomedical and NeuroMotor Sciences (DIBINEM), University of Bologna, Bologna, Italy, <sup>3</sup>Medical Genetics, Department of Reproductive Sciences, Development and Public Health, Trieste, Italy, <sup>4</sup>IRCCS-Burlo Garofolo Children Hospital, University of Trieste Trieste, Italy, <sup>5</sup>Doheny Eye Institute, UCLA Los Angeles, CA, USA, <sup>6</sup>Studio oculistico d'Azeglio Bologna, Italy, <sup>7</sup>IRCCS Istituto Scientifico San Raffaele Milano, Italy, <sup>8</sup>Department of Ophthalmology, Federal University of Sao Paulo (UNIFESP) Sao Paulo, Brazil

#### Introduction:

We aim to demonstrate that “purely genetic” and “tobacco-triggered” LHON may be different diseases.

#### Methods:

We present three LHON affected brothers: two cases strikingly resembled “tobacco-alcohol amblyopia”, with late-onset after decades of heavy smoking and strict temporal loss of fibers, whereas the third brother had classic severe LHON presentation in adolescence. In two cases (classic LHON and tobacco-related) we studied postmortem optic nerve cross-sections. We also retrieved the information on age of onset and environmental exposure in a large cohort of Italian LHON patients (n=134) and unaffected mutation carriers (n=126) and analyzed the data as in Kirkman et al., (Brain 2009).

#### Results:

At histology a devastating loss of axons involved the entire optic nerve with some peripheral sparing in the young-onset case, whereas the late-onset, tobacco-related case had a selective loss of temporal axons. In the Italian cohort, 2/3 of the affected individuals were smokers, compared with 1/2 of the unaffected carriers. Purely genetic cases (non-smokers) had a median age of onset of 16 yrs with a tight distribution, whereas smokers had a broader distribution with a median age of onset of 29 yrs. Gender stratification further highlighted tighter distribution of age of onset in males. The Kaplan-Meier curve corroborated the existence of two distinct subpopulations in affected individuals, related to age of onset and tobacco exposure.

#### Conclusions:

LHON can be distinguished in a “classic” early-onset form (15-30 years), with a prevalent genetic predisposition. A second late-onset form (>40 years) is strongly associated with long-term (decades) heavy smoking and most of these patients remained unaffected if avoided smoking. This distinction is crucial for identification of nuclear genetic modifiers of LHON penetrance. In fact, the genes most important for conversion in pure genetic cases may not be the same genes critical in tobacco-induced LHON.

#### References:

1. Kirkman MA, Yu-Wai-Man P, Korsten A, Leonhardt M, Dimitriadis K, De Coo IF, Klopstock T, Chinnery PF. Gene-environment interactions in Leber hereditary optic neuropathy. Brain. 2009 Sep;132(Pt 9):2317-26.

**Keywords:** Genetic Disease, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Telethon Grants GGP06233 and GGP11182 (V.C.) Research to Prevent Blindness, the International Foundation for Optic Nerve Diseases (IFOND), Struggling Within Leber's, The Poincenot Family, the Eierman Foundation, and a National Eye Institute grant EY03040 (F.N.R.-C and A.A.S.).

## Poster 26

### Changes in Macular OCT Retinal Sublayers of Patients and Carriers in the Natural History Phase of the Leber Hereditary Optic Neuropathy G11778A Gene Therapy Clinical Trial

Byron L. Lam<sup>1</sup>, Samuel P. Burke, Mindy Wang, Gloria Nadayil, Giovanni Gregori, William J. Feuer, Potyra R. Rosa, Sophia Cuprill-Nilson, Ruth Vandenbroucke, John Guy

*Bascom Palmer Eye Institute, University of Miami, Miami, FL, USA*

#### Introduction:

We determined macular retinal sublayer changes of patients and carriers with G11778A Leber hereditary optic neuropathy (LHON) by utilizing data from the natural history phase of the gene therapy trial where subjects underwent visual examination every 6 months from 2008 to 2013.

#### Methods:

Spectral-domain Cirrus OCT 512x128 macular cube scans were segmented for the baseline examination. Segmented retinal sublayers were the RNFL, GCL-IPL, INL-OPL, ONL-IS, and OS. Poor quality scans that cannot be segmented accurately were excluded. The thickness of the retinal sublayers of the LHON subjects and carriers were compared to a normal group.

#### Results:

Segmented macular OCT data were available from 20 LHON subjects (age  $31 \pm 14$  years, range 10-61), 31 LHON carriers (age  $38 \pm 18$ , 9-65), and 14 normal subjects (age  $39 \pm 13$ , 23-61). In general, parameters were not significantly correlated with age in any of the groups. There were no differences between carriers and normals for any sublayers but some layers were thinner or thicker in LHON subjects. Layers showing the most striking differences between LHON and the other two groups included the RNFL, GCL-IPL, and OS (p-values from 0.046 to  $<0.001$ ). LHON RNFL and GCL-IPL were thinner than carriers and normals while LHON OS was thicker than carriers and normals. Differences between groups were not significant in the INL+OPL. The ONL+IS layer of the temporal and inferior quadrants (inner and outer macular annular rings) was thicker in LHON ( $0.041 \geq p \geq 0.009$ ).

#### Conclusions:

LHON patients, compared to normal subjects, have thickened photoreceptor outer segment layer and some thickening of the ONL-IS in spite of having thinner RNFL and GCL-IPL layers. LHON carriers have retinal sublayer thickness similar to normal subjects. The findings indicate optic nerve degeneration in LHON also has an effect on the morphology of the outer retina.

**References:** None.

**Keywords:** Leber Hereditary Optic Neuropathy, OCT, Photoreceptor Outer Segment

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** NEI R24EY018600

## Poster 27

### Low Grade Glioma of the Pituitary Stalk, Case Report with Review of the Literature.

Su Ann Lim<sup>1</sup>, Hwei Yee Lee<sup>1</sup>, Wai Yung Yu<sup>2</sup>, Wai Hoe Ng<sup>2</sup>, Shaikh A K K Abdul S<sup>1</sup>

<sup>1</sup>Tan Tock Seng Hospital, Singapore, Singapore, <sup>2</sup>National Neuroscience Institute, Singapore, Singapore

#### Introduction:

In adults, thickening of the pituitary stalk can be the result of primary or secondary tumours, inflammatory or vascular lesions.<sup>1</sup> Glioma of the pituitary stalk is rare. Only two other cases have been reported in the English literature.<sup>2,3</sup> Clinical, imaging and pathological features of this rare tumour will be presented.

#### Methods:

Case report

#### Results:

A 39 year old female presented with diabetes insipidus (DI) and amenorrhea. Investigations revealed panhypopituitarism and as well as thickening of the pituitary stalk. She was followed up for a year during which the lesion enlarged causing visual field loss. Initial biopsy was inconclusive. Subsequent stereotactic biopsy revealed features compatible with a low grade glioma.

#### Conclusions:

To our knowledge, only two other cases of glioma affecting the pituitary stalk have been published. Although rare, this tumour should be considered, especially in cases presenting with DI and pituitary stalk thickening. Other glial tumours in this region, namely pituicytoma, spindle cell oncocytoma and granular cell tumour, do not usually present with DI, prolactinemia or galactorrhoea. These tumours usually present with visual disturbance and headache.<sup>4</sup> Stereotactic biopsy increases the accuracy of targeting the lesion and obtaining diagnostic yield<sup>2</sup>.

#### References:

1. Varan A, Atas E, Aydin B, Yalcin B, et. al. Evaluation of Patients with Intracranial Tumours and Central Diabetes Insipidus. *Pediatr Hematol Oncol* 2013;30:668-673.
2. Yap L, Crooks D, Warnke P. Low grade astrocytoma of the pituitary stalk. *Acta Neurochir* 2007;149:307-312.
3. Takeuchi J, Kikuchi K, Shibamoto Y, et. al. Radiation therapy for juvenile pilocytic astrocytoma of the pituitary stalk. *J Neurosurg* 1992;77:139-142.
4. Covington MF, Chin SS, Osborn AG. Pituicytoma, spindle cell oncocytoma, and granular cell tumour: Clarification and meta-analysis of the world literature since 1893. *AJNR* 2011;32:2067-72.

**Keywords:** Pituitary Stalk Enlargement, Glioma, Diabetes Insipidus, Tumors, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Visualisation of nerve fibre orientation in the human optic chiasm using photomicrographic image analysis

Neeranali Jain<sup>1,2</sup>, Swaranjali Jain<sup>1,2</sup>, Xiaofei Wang<sup>3</sup>, Andrew Neely<sup>3</sup>, Murat Tahtali<sup>3</sup>, Sanjiv Jain<sup>4,5</sup>, Christian J Lueck<sup>4,5</sup>

<sup>1</sup>Department of Neurology, The Canberra Hospital, Canberra, Australia, <sup>2</sup>Faculty of Medicine, University of New South Wales, Sydney, Australia, <sup>3</sup>School of Engineering and Information Technology, University of New South Wales, Canberra, Australia, <sup>4</sup>Department of Anatomical Pathology, the Canberra Hospital Canberra, Australia, <sup>5</sup>Australian National University Medical School Canberra, Australia

### Introduction:

Compression of the optic chiasm typically gives rise to bitemporal hemianopia due to selective damage to the decussating nasal fibres. It is unclear why nasal fibres are particularly vulnerable to the extent of generating a sharp vertical cut-off in the visual fields. One theory suggests that this is due to the geometry of individual fibres within the chiasm.<sup>1</sup> Unfortunately, detailed anatomical information about the precise arrangement and crossing of nerve fibres in the chiasm is limited. This study aimed to clarify the microscopic anatomy of the chiasm, looking particularly at nerve fibre distribution and the location of nerve fibre crossings.

### Methods:

A human optic chiasm obtained at autopsy was stained *en bloc* with silver stain and sectioned in the axial plane at 5 µm intervals. Photomicrographs were digitized and subdivided into smaller regions of interest (ROIs). Fibre orientation distribution data for each ROI were obtained and processed using *ImageJ* software and custom-written MATLAB code. The orientation data and crossing angles were then represented graphically.

### Results:

The central portion of the chiasm was found to contain fibres travelling predominantly in parallel in a medio-lateral direction. Nerve fibre crossings were located in the antero-inferior and supero-posterior portions of the para-central parts of the chiasm.

### Conclusions:

This study suggests that nerve fibre crossings are not located centrally in the chiasm but in the paracentral regions. The data from the study will be used to inform models of the optic chiasm which, in turn, will generate further insight into the pathophysiology of bitemporal hemianopia.

### References:

1. McIlwaine GG, Carrim ZI, Lueck CJ, Chrisp TM. A mechanical theory to account for bitemporal hemianopia from chiasmal compression. *Journal of Neuro-Ophthalmology*. 25(1), 40-3, 2005

**Keywords:** Optic Chiasm, Histology, Bitemporal Hemianopia, Photomicrographs

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Optic Neuropathy in Chronic Lymphocytic Leukemia

Amina I. Malik<sup>1</sup>, Michael Morgan<sup>2</sup>, Patricia Chevez-Barrios<sup>3</sup>, Andrew G. Lee<sup>4</sup>, Sumayya Almarzouqi<sup>5</sup>, Sushma Yalamanchili<sup>6</sup>

<sup>1</sup>Houston Methodist Hospital Department of Ophthalmology, Houston, TX, USA, <sup>2</sup>Houston Methodist Hospital Department of Ophthalmology, Houston, TX, USA, <sup>3</sup>Houston Methodist Hospital Department of Ophthalmology, Houston, TX, USA, <sup>4</sup>Houston Methodist Hospital Department of Ophthalmology Houston, TX, USA, <sup>5</sup>Houston Methodist Hospital Department of Ophthalmology Houston, TX, USA, <sup>6</sup>Houston Methodist Hospital Department of Ophthalmology Houston, TX, USA

### Introduction:

Central nervous system involvement in chronic lymphocytic leukemia (CLL) is rare. It may manifest with cognitive dysfunction, cerebellar signs, or cranial nerve palsies (1,5,6). Optic neuropathy (ON) in CLL is extremely rare with few previous reports described. Diagnosis is typically made from CSF analysis. Previous case studies have reported successful treatment with radiation or intrathecal methotrexate. We report an unusual case of CLL optic neuropathy.

### Methods:

A case report is described.

### Results:

A 42 year old African American male with history of CLL in remission presented with subacute onset of right sided blurry vision. Visual acuity was 20/30 OD, 20/20 OS with right afferent pupillary defect. Fundoscopy revealed 2+ disc edema OD, 1+ disc edema OS. MRI brain and orbits showed bilateral diffuse optic nerve enlargement. The patient underwent lumbar puncture (LP), which revealed 13 wbc/microL (95% lymphocytes) with negative cytology and flow cytometry. Patient's disc edema was re-evaluated at 1 month, and had increased to 3+ OD, 2+ OS. Biopsy of the tissue adjacent to the right optic nerve was performed, which revealed abnormal B cell lymphocytic proliferation. Repeat LP revealed 13 wbc/microL (100% lymphocytes) with cytology and flow cytometry showing clonal B cells co-expressing CD5 with kappa light chain restriction. This patient was treated with intrathecal methotrexate and chemotherapy (ibrutinib). At 3 month follow-up, his disc edema had improved to 1+ and visual acuity to 20/25 OD.

### Conclusions:

We report an unusual case of CLL ON, diagnosed by peri-optic nerve biopsy after negative initial CSF cytology, suggesting that multiple LPs may be necessary to diagnose CNS involvement in CLL. While previous reports have described ON development during hematologic relapse, this patient's symptoms developed while in remission. Additionally, this case describes successful use of intrathecal methotrexate with chemotherapy, without need for orbital radiation, in a patient with CLL ON.

### References:

1. Currie JN, Lessell S, Lessell IM, et al. Optic Neuropathy in Chronic Lymphocytic Leukemia. Arch Ophthalmol. 1988 May;106(5):564-560.
2. Cramer SC, Glaspy JA, Efird JT, et al. Chronic Lymphocytic Leukemia and the Central Nervous System: A Clinical and Pathological Study. Neurology 1996;46:19-25.
3. Barcos M, Lane W, Gomez GA et al. An Autopsy Study of 1206 Acute and Chronic Leukemias (1958 to 1982). Cancer 1987;60:827-837.
4. Cash J, Fehir KM, Pollack MS. Meningeal Involvement in Early Stage Chronic Lymphocytic Leukemia. Cancer 1987;59:798-800.
5. Takahashi T, Oda Y, Isayama Y. Leukemic Optic Neuropathy. Ophthalmologica. 1982;185(1):37-45.
6. Mowatt L, Matthews T, Anderson I. Sustained Visual Recovery after Treatment with Intrathecal Methotrexate in a Case of Optic Neuropathy Caused by Chronic Lymphocytic Leukemia. J Neuro-Ophthalmol. 2005;25(2):113-115.

**Keywords:** Chronic Lymphocytic Leukemia, Optic Neuropathy, Disc Edema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 30

### How is Eye Fixation Affected by Optic Neuropathy? Diagnostic Value of Precise Recording of Retina Movement During an OCT Scan

Robert M. Mallery<sup>1</sup>, Matthew J. Thurtell<sup>1,2</sup>, Pieter Poolman<sup>1,2</sup>, Jan M. Full<sup>1,2</sup>, Johannes Ledolter<sup>2,3</sup>, Enrique J. Rivera<sup>1</sup>, Randy H. Kardon<sup>1,2</sup>

<sup>1</sup>University of Iowa/Department of Ophthalmology and Visual Sciences, Iowa City, IA, USA, <sup>2</sup>VA Medical Center and the Center of Excellence for the Prevention and Treatment of Visual Loss, Iowa City, IA, USA, <sup>3</sup>University of Iowa/Department of Statistics and Actuarial Science, Iowa City, IA, USA

#### Introduction:

Instability of fixation and emergence of alternate preferred retinal loci (PRL) for fixation are well described in macular disease, but have not been well characterized in optic neuropathy. We evaluated the extent of retinal movements recorded during a standard OCT macular volume scan and explored its diagnostic utility.

#### Methods:

19 patients with central visual field loss from optic neuropathy (14 unilateral, 5 bilateral) and 11 normal subjects underwent OCT scanning with eye tracking (Spectralis, Heidelberg Engineering). Subjects fixated on an internal central blue target during OCT volume scans acquired with SLO eye tracking, lasting approximately 30 seconds. The distribution of fixation points on the retina with respect to the anatomic fovea during the OCT scan was quantified using Kernel Density Estimation (KDE). Each patient underwent EDTRS visual acuity testing and perimetry (Goldmann or Humphrey 24-2 SITA) to confirm central visual field loss.

#### Results:

Stability of fixation, measured by the area enclosed by the 68% isoline of the KDE of retina position, was significantly worse in the 22 eyes with optic neuropathy (mean=0.7 deg<sup>2</sup>) compared with the 33 unaffected eyes (mean=0.062 deg<sup>2</sup>) (two-sample t-test, p=0.005). Multiple PRL of fixation were identified in 8/22 eyes with optic neuropathy, and the degree of extra-foveal fixation was greater in eyes with optic neuropathy (two-sampled t-test, p=0.001). In patients with unilateral optic neuropathy, ANOVA identified a difference in fixation stability (p=0.013) and eccentricity of fixation (p=0.043) between the eyes with optic neuropathy and their paired, unaffected eyes.

#### Conclusions:

We introduce a readily accessible method of eye tracking recorded during OCT to provide an objective measure of fixation, which was significantly impaired in patients with optic neuropathy who have central visual field loss. Fixation stability assessed during standard OCT may help differentiate causes and severity of vision loss and facilitate monitoring of optic nerve function.

#### References: None.

**Keywords:** Optic Neuropathy, Fixational Eye Movements, Diagnostic Tests, Optic Coherence Tomography, Scanning Laser Ophthalmoscope

**Financial Disclosures:** R. Kardon Grants:Funding (grants) from NEI R009040554 R01 EY018853Funding (grants) Department of Defense TATRCFunding (grants) VA Rehabilitation Research and DevelopmentR. Kardon Consulting: Novartis steering committee OCTiMS

**Grant Support:** R. Kardon: Funding (grants) from NEI R009040554 R01 EY018853 Funding (grants) Department of Defense TATRC Funding (grants) VA Rehabilitation Research and Development



## Poster 31

### A Novel OPA1 Mutation in Autosomal Dominant Optic Atrophy (ADOA)

Jordan A. Margo<sup>1</sup>, Jana A. Bregman<sup>2</sup>, Vivian Rismondo<sup>3</sup>

<sup>1</sup>University of Maryland, Baltimore, MD, USA, <sup>2</sup>Vanderbilt University School of Medicine, Nashville, TN, USA, <sup>3</sup>Greater Baltimore Medical Center, Towson, MD, USA

#### Introduction:

Autosomal dominant optic atrophy (ADOA), or Kjer's disease, often overlaps clinically with other forms of optic atrophy making diagnosis challenging. Considering the potential for ADOA to cause significant visual impairment and its high degree of penetrance, it is important to make the correct diagnosis for both the individual patient and the family members.

#### Methods:

Here we describe a case of ADOA in a 61-year-old West African male who presented after several years of symptoms. Our investigation resulted in the detection of a novel mutation in OPA1 that had not been previously classified.

#### Results:

The patient presented with complaints of bilateral visual decline, beginning in his 30s, progressing over 10-15 years and then stabilizing. Examination was significant for the following findings: visual acuity of 20/100 OD, 20/80 OS and abnormal Ishihara testing in both eyes without APD. Fundoscopic examination revealed temporal disc pallor and thinning was confirmed on OCT. Previous testing showed reduced amplitudes and prolonged latencies on VEP and bilateral nasal visual field defect on microperimetry. Multifocal and full field electroretinography was normal and pedigree revealed similarly affected family members. Closer consideration of test results helped identify ADOA as the most likely cause of vision loss. Genetic testing identified a novel pathologic mutation in OPA1, variant c.748 (isoform 8) G>A; p.A1a250/Thr.

#### Conclusions:

Our patient was diagnosed with a rare cause of optic atrophy, years after symptom onset and fragmented care at multiple institutions. Genetic testing in concert with his clinical presentation revealed ADOA as the most likely diagnosis. This case helps further characterize ADOA by introducing a novel mutation and demonstrating its clinical significance. It also serves as a reminder that selective genetic testing can and should be performed after patient counseling and following initial laboratory and imaging work-up for progressive bilateral vision loss.

**References:** None.

**Keywords:** Genetic Disease, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 32

### **The vegetative aspects of neuroprotective action of high corticosteroid doses in compressive traumatic optic neuropathies (TON).**

Yulya T. Maslyak<sup>1</sup>, Nataliya M. Moyseyenko<sup>1</sup>, Halyna M. Leskiv<sup>2</sup>

<sup>1</sup>Ivano-Frankivsk National Medical University, Ivano-Frankivsk, Ukrenia, <sup>2</sup>Rohatyn Central District Hospital, Rohatyn, Ukrenia

#### **Introduction:**

Approaches to traumatic optic neuropathy treatment vary from country to country. High doses of corticosteroids is considered to be standard modality of management in many countries. However, there are several issues related to the possible side effects of this treatment option. The other limitation is inability to use this treatment modality in the first 48 years of the condition.

#### **Methods:**

Damage to right optic nerve in surgical setting has been modeled in 22 rabbits. 11 rabbits have been administered methylprednisolone subcutaneously (30mg per 1 kg) starting from the second day for three days. The dose was decreased after the third day of treatment. Control group consisted of 11 rabbits. Heartbeats and amplitude of pupil reaction (2 weeks and one month after trauma) have been measured in all 22 rabbits.

#### **Results:**

Methylprednisolone administration resulted in statistically significant elevation of heartbeat to  $219 \pm 31$  bpm one month post trauma in study group and  $182 \pm 23$  bpm in control group that proves normalization of the vegetative system functioning. Amplitude of pupil reaction on the affected side has increased from 0mm without treatment to  $1 \pm 0.5$ mm one month after treatment. This proves functional capability recovery of the optic nerve's afferent portion. Furthermore, this means beginning of optic nerve regeneration.

#### **Conclusions:**

Use of high doses of corticosteroids facilitates improved function of vegetative system. Therefore, it has neuroprotective action on neural tissue in compressive traumatic damage of the optic nerve in experiment.

**References:** None.

**Keywords:** Traumatic Optic Neuropathy, Pupil, Heartbeat, Corticosteroids, Rabbit

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 33

### Optic Atrophy in a Large Specialist Hospital

Joyce N Mbekeani<sup>1,3</sup>, Maaly Abdel Fattah<sup>1,4</sup>, Abdelmoneim Eldali<sup>2</sup>, Selwa A Hazzaa<sup>1,5</sup>

<sup>1</sup>Dept of Ophthalmology, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia, <sup>2</sup>Dept of Biostatistics, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia, <sup>3</sup>Dept of Ophthalmology and Visual Sciences, Albert Einstein College of Medicine, Bronx, NY, USA, <sup>4</sup>Dept of Ophthalmology, Cairo University Cairo, Egypt, <sup>5</sup>Al Faisal University Riyadh, Saudi Arabia

#### Introduction:

Optic atrophy is a frequent presentation in our clinics. It is not a diagnosis but represents permanent retinal ganglion cell loss that warrants investigations to establish etiology. Often the result of ischemic insult or neuritis in developed nations, we have noticed many other causes especially the effects of intracranial tumors. Our purpose was to elucidate the demographics and distribution of causes within our population.

#### Methods:

A prospective, descriptive study of patients presenting with optic atrophy was conducted with institutional IRB approval. Diagnosis was made based on visual acuities, ophthalmoscopic features and ancillary tests. Patient demographics, data from clinical exams and, where possible, VF, OCT, VEP and neuro-radiologic findings were tabulated. Decimal visual acuities were used to facilitate calculations. Sub-populations were compared and correlations determined by Chi-square, Mann-Whitney and Kruskal-Wallis tests. Atrophy secondary to glaucoma and primary retinal causes were excluded.

#### Results:

353 eyes of 204 patients were diagnosed with optic atrophy over nine months. The mean age was 30.8 years (range 0.25-77) with a median of 27 years (IQR=27). There were 111 (54.4%) females and 93 (45.6%) males. With statistical significance set at  $p=0.05$ , there was no difference in age of presentation between the genders. 155 cases (76%) were bilateral whilst 49 (24%) were unilateral. The commonest cause, intracranial tumors, accounted for 123 (60.3%) with the next cause, autoimmune diseases, accounting for 24 (11.8%). Neoplasia was more likely to cause bilateral atrophy than the other causes combined ( $p=0.007$ ) and occurred more in adults than pediatric (<20 years) patients when compared to other causes ( $p=0.019$ ). There was no significant correlation between gender or visual acuity and the etiology of optic atrophy.

#### Conclusions:

Neoplasia was the most common cause of optic atrophy. Although this may have resulted from mostly complicated, advanced case-referrals to a specialist hospital, it may also represent a high incidence of aggressive tumors and low awareness resulting in late presentations in the studied population.

**References:** None.

**Keywords:** Optic Nerves, Optic Atrophy, Neoplasia, Intracranial

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 34

### Neurofibromatosis 1 with Large Suprasellar and Bilateral Optic Nerve Pilomyxoid Astrocytoma

Joyce N Mbekeani<sup>1</sup>, Mohammed A Dababo<sup>2</sup>, Manzoor Ahmed<sup>3</sup>

<sup>1</sup>Dept of Ophthalmology and Visual Sciences, Albert Einstein College of Medicine, Bronx, NY, USA, <sup>2</sup>Dept of Anatomic Pathology, King Faisal Specialist Hospital & Research Center, Riyadh, Saudi Arabia, <sup>3</sup>Dept of Neuro-radiology, Cleveland Clinic Abu Dhabi, Abu Dhabi, United Arab Emirates

#### Introduction:

Bilateral optic nerve gliomas are suggestive of NF1 and typically are low grade (WHO Grade I) pilocytic astrocytoma (PA). Pilomyxoid astrocytoma (PMA) is a newly characterized, aggressive glioma (WHO grade II) with a propensity for the hypothalamic-chiasmal axis. It occurs in a younger age group and has a higher recurrence and lower survival rate. There are few reports associating PMA with NF1.

#### Methods:

We present a patient with genetically determined NF1 who presented with a large hypothalamic PMA extending to both optic nerves. Clinical, neuro-radiologic and histopathologic findings will be presented.

#### Results:

A previously healthy 2 year old girl with NF1 was noticed to have limited eye movements, strabismus and decreasing body mass. MRI revealed a large heterogeneous suprasellar and hypothalamic mass, involving the chiasm and extending into both optic nerves. Surgical debulking was performed and pathology revealed spindle-shaped, piloid astrocytes within a rich myxoid matrix. Classified as pilomyxoid astrocytoma, chemotherapy was given but radiation was avoided to avert inducing secondary tumors common in NF1. She has been followed regularly by ophthalmology, neurosurgery and pediatric neurology. On recent exam at 10years of age, she had multiple café au lait spots and axillary freckling. Vision was NPL OD and 20/200 OS. EOMs were full with a large angle right exotropia. Pupils were sluggishly reactive, anterior segments revealed few iris Lisch nodules but were otherwise normal and ophthalmoscopy revealed bilateral optic atrophy with severe loss of arcuate fibres. Recent MRI revealed post-operative changes but no evidence of residual tumor or recurrence.

#### Conclusions:

Although NF1 typically presents with pilocytic astrocytoma, the clinically and histopathologically distinct pilomyxoid astrocytoma should be considered. PMA is a more aggressive tumor occurring in a younger age group, and typically has a heterogeneous MRI pattern. Its manifestation in this setting likely represents the wide phenotypic spectrum of NF1.

**References:** None.

**Keywords:** Neurofibromatosis 1, Optic Nerve Glioma, Pilomyxoid Astrocytoma, Optic Atrophy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Optic neuropathy in Wolfram's syndrome imaged with high-definition spectral domain OCT

Dan Milea<sup>1,2,3,4</sup>, Mani Baskaran<sup>1,2</sup>, Marion Verschoore<sup>4</sup>, Stéphanie Leruez<sup>4</sup>, Xavier Zanlonghi<sup>5</sup>, Christophe Orssaud<sup>6</sup>, Pascal Reynier<sup>4</sup>, Amati-Bonneau Patrizia<sup>4</sup>, Dominique Bonneau<sup>4</sup>, Christophe Verny<sup>4</sup>, Régis Coutant<sup>4</sup>, Tin Aung<sup>1,2,3</sup>, Vincent Procaccio<sup>4</sup>

<sup>1</sup>Singapore National Eye Centre, Singapore, Singapore, <sup>2</sup>Singapore Eye Research Institute, Singapore, Singapore, <sup>3</sup>Duke\_NUS, Singapore, Singapore, <sup>4</sup>Angers University Hospital Angers, France, <sup>5</sup>Clinique Sourdis Nantes, France, <sup>6</sup>HGEP Paris, France

### Introduction:

Optic neuropathy is a cardinal finding in Wolfram syndrome (WS), which typically also associates at least one of the following findings: diabetes mellitus, diabetes insipidus and deafness. The aim of this study was to describe genetic, clinical and high definition OCT (HD-OCT) findings in patients with an optic neuropathy associated with genetically confirmed Wolfram syndrome.

### Methods:

This cross-sectional study included 19 patients with an optic neuropathy and genetically confirmed WS due to mutations in the *WFS1* gene, including three previously unreported mutations (c.1525\_1539del15, c.1153G>T and c.1592\_1593dup) and 39 age-matched healthy controls. Routine ophthalmic examinations, including HD-OCT (Cirrus, software version 6.0, Carl Zeiss Meditec, Dublin, CA), with segmentation of the peripapillary retinal nerve fiber layer (RNFL) and of the perifoveal retinal ganglion cell-inner plexiform layer (GC-IPL).

### Results:

The mean age at onset of optic neuropathy was 15.9 years (SD=14), significantly younger in patients with compound heterozygous mutations (n=13, mean age=9.1) than in patients with single allele mutations (n=6, mean age=33.3, p=0.001). HD-OCT imaging disclosed diffuse thinning of RNFL and IP-GCL in WS patients, most severely in the inferotemporal parapapillary region (54.15%, p=0.001). GC-IPL thinning was correlated with age in WS patients, in the inferior quadrant (r = -0.611, B = -0.4, p = 0.009) and in the temporal quadrant (r = -0.537, B = -0.412, p = 0.026). RNFL thickness in the temporal quadrant was significantly correlated with visual acuity (r = 0.644, B = 13.01, p = 0.004). Patients with WS had statistically significant larger vertical cup disc ratios, compared to controls (0.62 vs 0.43, p=0.001) and smaller rim areas (1.06 vs 1.45 mm, p=0.001).

### Conclusions:

Optic neuropathy in Wolfram's syndrome may present at later ages than previously reported, when associated with single allele mutations in the *WFS1* gene. HD-OCT allows anatomical phenotyping of the optic neuropathy, disclosing diffuse thinning of the retinal nerve fiber layers, with a predominant inferotemporal RNFL loss and associated optic disc cupping.

**References:** None.

**Keywords:** Wolfram Syndrome, Hereditary Optic Neuropathy, Optic Neuropathy, OCT, Genetics

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 36

### **Retinal oximetry (oxygen saturation) and peripapillary vascular diameters in normal eyes and in non-arteritic ischemic optic neuropathy (NAION)**

Ashwin Mohan<sup>1</sup>, Rohit Shetty

*Narayana Nethralaya, Bangalore, India*

#### **Introduction:**

Retinal oximetry is a non-invasive dual wavelength photo-spectrometric imaging of the retina which gives the oxygen saturation in the retinal vessels. Non-arteritic ischemic optic neuropathy results due to ischemia in the circulation of the optic nerve head. The central retinal artery enters the eye through the optic nerve. The disc swelling may directly affect the arterioles and venules, or the same pathology affecting the optic nerve head circulation may also affect the retinal circulation. We aim to study these alterations in the retinal vessels in cases of non-arteritic ischemic optic neuropathy

#### **Methods:**

Fifteen eyes diagnosed to have non-arteritic ischemic optic neuropathy on clinical examination underwent retinal oximetry (Oxymap T1, Oxymap hf, Iceland) to determine the oxygen saturation in the retinal vessels and their diameters. The values obtained from the affected eyes were compared to the other eye and 50 age matched normals.

#### **Results:**

Arteriolar diameters were reduced and venous diameters were increased in the affected eyes. Arteriolar saturations were increased, venous saturations were decreased and arterio-venous saturation difference (AVSD) was increased as compared to the other groups. The fellow eye showed alterations in oxygen saturation as compared to the age-matched normal, however the changes were subtle and not statistically significant.

#### **Conclusions:**

Decreased arteriolar diameters and increased venous diameters suggest stasis possibly due to the increased pressure due to optic nerve head edema. This may result in increased arterio-venous transit time thus allowing more time for oxygen exchange. This could possibly explain the increased arterio-venous saturation difference. Alterations in the fellow eye need to be confirmed with a larger case series, where favorable results imply that oximetry may be useful in the screening and prevention of ischemic optic neuropathy in patients with disk at risk or systemic diseases like diabetes mellitus or hypertension that place them at risk for NAION and possibly predict NAION in the fellow eye.

**References:** None.

**Keywords:** Oximetry, Optic Neuropathy, Ischemia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 37

### Is Ishihara Color Plate Testing As Reliable On iPod/iPhone and iPad as on paper format?

Katherine Boudreault<sup>1</sup>, Kinda Najem, Katie Luneau

*Notre-Dame Hospital CHUM, Ophthalmology department, Montreal, QC, Canada*

#### **Introduction:**

Ishihara testing is frequently used to evaluate dyschromatopsia in patients with optic nerve disease. Several electronic versions are available nowadays. We compared results on the electronic versus paper formats in patients with dyschromatopsia.

#### **Methods:**

Our study was comprised of 3 groups. Group 1 included 20 normal patients, group 2 had 20 patients with unilateral optic neuropathy and group 3, 21 patients with unilateral maculopathy. All patients performed 3 Ishihara tests (same color plates on paper, iPod and iPad) in an aleatory order. Results were reported on a total of 13. For each patient, the scores of the different versions were compared to verify if a systematic difference was detectable.

#### **Results:**

Compared with the normal group, patients in groups 2 and 3 had weaker performances with both paper and electronic formats ( $p < 0.0001$ ). Patients in the three groups had more difficulty with the electronic versions ( $p < 0.0001$ ). The iPod gave inferior results compared with the iPad ( $p < 0.0001$ ). When comparing both normal eyes, in group 1, results were constant for each eye, regardless of the testing version used ( $p = 0.42$ ). In groups 2 and 3, the difference between both eyes (normal versus diseased eye) was less pronounced with the paper format and the difference was worse with the iPod than with the iPad ( $p = 0.002$ ).

#### **Conclusions:**

Our study shows that Ishihara electronic versions are more difficult to perform than the paper format in all groups. Results were different even between electronic devices (iPod/iPad). Considering that other factors can influence the results, such as the versions of color vision tests on the Internet and the type of electronic devices used, we cannot extrapolate on the reliability of the electronic formats compared to the gold standard paper test. However, this study highlights the importance of not comparing results between different formats and even between different electronic devices.

#### **References:**

1. Swanson WH, Cohen JH, Color vision, Ophthalmol. Clin. N. Am., 16, 179-203, 2003.
2. Kuchenbecker H, Lindner W, Visual function tests on the Internet – sense or nonsense?, Strabismus, 12, 97-102, 2004.
3. Kuchenbecker H., Lindner W. et al., Options and limits of visual function tests available on the Internet, Ophthalmologie, 99, 866-871, 2002.

**Keywords:** Ishihara Color Plate Testing, iPod/iPhone/iPad, Optic Neuropathy, Dyschromatopsia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 38

### Multifactorial Optic Neuropathy - When it isn't always glaucoma.

Deborah C Parish<sup>1</sup>, Gabrielle R Bonhomme<sup>2</sup>, Ahmara G Ross<sup>3</sup>, Islam M Zaydan<sup>4</sup>

<sup>1</sup>UPMC / Neuro-Ophthalmology, Pittsburgh, PA, USA, <sup>2</sup>UPMC / Neuro-Ophthalmology, Pittsburgh, PA, USA, <sup>3</sup>UPMC / Neuro-Ophthalmology, Pittsburgh, PA, USA, <sup>4</sup>UPMC / Neuro-Ophthalmology Pittsburgh, PA, USA

#### Introduction:

We present a case of a rare HLA-B27 associated optic neuropathy masquerading as glaucomatous progression.

#### Methods:

A 38-year-old male with a history of HLA-B27 positive uveitis presented with an elevated intraocular pressure (IOP) of 60mmHg OD / 30mm Hg OS. He had a past ocular history of bilateral Retisert implants for persistent macular edema after cataract surgery. The patient subsequently underwent glaucoma device implant (GDI) placement and was started on topical therapy. Despite adequate IOP control, the patients vision in his right eye remained LP but the vision had deteriorated from 20/40 to 20/400 OS. Given the patients progressive vision loss and pain, despite adequate IOP, a neuro-ophthalmology consult was obtained. At the time of our exam, the patients vision had decreased to 20/800 OS and the patient was unable to perceive control Ishihara or contrast sensitivity plates. There was a right RAPD and an irregular surgical pupil OS. Ocular alignment revealed a sensory XT in primary gaze with normal motility. DFE revealed bilateral pallor of the optic nerves with C/D ratios of 0.99 OD and 0.5 OS.

#### Results:

MRI revealed an abnormal T2 signal with enhancement of the optic nerve and retrobulbar tissues. Goldmann visual fields were significant for generalized depression of the visual field sparing an island of vision nasally OD and global restriction OS. OCT and HRT were not consistent with glaucomatous damage. An extensive workup for suspected optic neuropathy included inflammatory, infectious, autoimmune and neoplastic etiologies. After all testing returned normal, the patient was admitted for IV steroids. Within 3 days of treatment, the patients had subjective improvement of his pain and his vision had improved to 20/80.

#### Conclusions:

Inflammatory optic neuropathy may initially be difficult to distinguish from glaucomatous damage, however, this case illustrates the importance of a thorough workup and evaluation in cases that masquerade themselves as pure glaucoma.

#### References:

1. Jakob, E., et al., Uveitis subtypes in a german interdisciplinary uveitis center--analysis of 1916 patients. J Rheumatol, 2009. 36(1): p. 127-36.
2. Chua, J., et al., Expression profile of inflammatory cytokines in aqueous from glaucomatous eyes. Mol Vis, 2012. 18: p. 431-8.
3. Rebolleda, G., et al., Optic disc cupping after optic neuritis evaluated with optic coherence tomography. Eye (Lond), 2009. 23(4): p. 890-4.

**Keywords:** Glaucoma, Optic Neuropathy, Hla-B27, Uveitis

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None



## Poster 39

### Cobalt-Chromium Metallosis with Normal ERG

Huy V. Nguyen<sup>1</sup>, Lola M. Grillo<sup>1</sup>, Stephen H. Tsang<sup>2</sup>, Donald C. Hood<sup>2</sup>, Jeffrey G. Odel<sup>2</sup>

<sup>1</sup>Columbia University College of Physicians and Surgeons, New York, NY, USA, <sup>2</sup>Columbia University Department of Ophthalmology, New York, NY, USA

#### Introduction:

Patients with a failed ceramic hip implant replaced by a cobalt-chromium implant may experience wear of the metallic implant from ceramic debris, resulting in elevations in serum cobalt and chromium. Within a year, patients may develop visual loss, sensory-neural hearing loss, hypothyroidism and cardiomyopathy. The visual loss has been assumed to be a toxic optic neuropathy. A previous case of cobalt/chromium toxicity suggests retinal involvement, with photopic ERG showing delayed and reduced amplitude and scotopic ERG showing reduced amplitude with an electronegative waveform.<sup>1</sup> Here, we present a patient with early cobalt/chromium metallosis.

#### Methods:

Case Report

#### Results:

A 66 year-old man with amblyopia OD presents with a two-month history of decreased color and acuity OU, difficulty hearing, cold intolerance, and cognitive decline. Seven months prior, he underwent a left cobalt-chromium head replacement for a failed ceramic hip. At time of presentation, the patient's serum cobalt was 1,076 ug/L (normal <0.9 ug/L) and his chromium level was 57 ug/L (normal <0.3 ug/L). On examination, his visual acuity was 20/30 OS and 20/80 OD with no RAPD. He saw 0/6 color plates OU. Fundi were normal. Humphrey 24-2 and 10-2 VF revealed central depressions OU. Multifocal VEP showed regions of central depressions consistent with defects seen on visual fields. OCT revealed retinal layers within normal limits, including the nerve fiber layer thickness OU. ISCEV Standard full-field ERG showed a normal latency and normal amplitude in both photopic and scotopic responses.

#### Conclusions:

Previous studies are divided as to whether cobalt/chromium toxicity causes damage to the retina or optic nerve.<sup>1-4</sup> Here, in our early case of cobalt-chromium metallosis, the results are not consistent with a retinal origin.

#### References:

1. Apel W, Stark D, Stark A, O'Hagan S, Ling J. Cobalt-chromium toxic retinopathy case study. *Doc Ophthalmology*. 2013;126(1):69–78.
2. Tower SS. Arthroprosthetic cobaltism: neurological and cardiac manifestations in two patients with metal-on-metal arthroplasty: a case report. *J Bone Joint Surg Am*. 2010;92(17):2847–2851.
3. Rizzetti MC, Catalani S, Apostoli P, Padovani A. Cobalt toxicity after total hip replacement: a neglected adverse effect? *Muscle Nerve*. 2011;43(1):146–7– author reply 147.
4. Steens W, Foerster von G, Katzer A. Severe cobalt poisoning with loss of sight after ceramic-metal pairing in a hip--a case report. *Acta Orthop*. 2006;77(5):830–832.

**Keywords:** Optic Neuropathy, Neuro-Ophth & Systemic Disease, Diagnostic Tests, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 40

### Nonarteritic Anterior Ischemic Optic Neuropathy (NAION): A Misnomer. A Non-Ischemic Papillopathy Caused By Vitreous Separation

Cameron F. Parsa<sup>1</sup>, William F. Hoyt<sup>2</sup>

<sup>1</sup>UPMC-Sorbonne Universities/Quinze-Vingts National Ophthalmology Hospital, Paris, France, <sup>2</sup>University of California San Francisco School of Medicine/Department of Ophthalmology, San Francisco, CA, USA

#### Introduction:

The fact that vascular abnormalities such as disc hemorrhages and swelling are present at the time of visual loss in NAION, followed by peripapillary vascular narrowing and ensuing disc pallor is enticing, but not etiologically conclusive for ischemia. There is both optic disc as well as retinal evidence that whiteness with disc swelling is indicative of axoplasmic stasis (cotton wool spots) that may also occur simply from anatomic distortion of axons rather than occlusion of vessels.<sup>1</sup> It may also occur from mechanical stretching and fracture of the axonal cytoskeleton<sup>2</sup> and frank membrane disruption with axoplasmic "leakage."

#### Methods:

Thirty-three patients with new-onset NAION and controls were examined for vitreous separation. Patients underwent OCT scanning to assess vitreous attachment to the peripapillary area. If no attachments were noted by OCT or by ophthalmoscopy, dynamic B-scan ultrasonography was performed to assess for total vitreous detachment.

#### Results:

Vitreous was fully detached from the optic disc in 40% of affected eyes while in the remaining 60% there was complete parapapillary detachment and only partial residual attachment to the disc. Findings were consistent with those reported in NAION showing complete peripapillary vitreous separation in all cases and elevated percentages of complete vitreous detachment from the disc (35-65%).<sup>3,4</sup> Teleangiectatic vessels on the disc surface in 10% correlated with visual field sparing and corresponded to areas of unseparated vitreous under tension. In age-matched individuals, only 10% had complete PVD, 70% partial PVD, and 20%, no PVD.

#### Conclusions:

Where ILM is absent over the disc and peripapillary retina and, most notably in cupless discs, vitreous adhesions are strongest, vitreous separation may momentarily stretch and elongate axons, breaking the cytoskeleton in more aged and less distensible axons,<sup>5</sup> leading to immediate axoplasmic accumulation and axonal atrophy in the prelaminar sites of separation. Ischemic pathophysiology need not be invoked in so-called NAION.

#### References:

1. McLeod D. Why cotton wool spots should not be regarded as retinal nerve fiber layer infarcts. *Br J Ophthalmol* 2005;89:229-37.
2. Tang-Schomer MD, Patel AR, Baas PW, Smith DH. Mechanical breaking of microtubules in axons during dynamic stretch injury underlies delayed elasticity, microtubule disassembly, and axon degeneration. *FASEB J* 2010;24:1401-10.
3. Sanjari MS, Falavarjani KG, Parvaresh MM, et al. Vitreopapillary traction in nonarteritic anterior ischemic optic neuropathy. *Iranian J Ophthalmic Res* 2006;1:110-2.
4. Lee MS, Foroozan R, Kosmorsky GS. Posterior vitreous detachment in AION. *Ophthalmology*. 2009;116:597.
5. Lamoureux PL, O'Toole MR, Heidemann SR, Miller KE. Slowing of axonal regeneration is correlated with increased axonal viscosity during aging. *BMC Neurosci* 2010;11:140.

**Keywords:** NAION, Optic Neuropathy, Diagnostic Tests, Vitreous Separation, Axonal Cytoskeleton Fracture

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 41

### Visual and Oculomotor Outcomes in Children with Posterior Fossa Tumors

Crandall E. Peeler<sup>1,2</sup>, Jeffrey Hollander<sup>3</sup>, Jane C. Edmond<sup>4,5</sup>, Gena Heidary<sup>1,2,3</sup>

<sup>1</sup>Massachusetts Eye and Ear Infirmary, Boston, MA, USA, <sup>2</sup>Harvard Medical School, Boston, MA, USA, <sup>3</sup>Boston Children's Hospital, Boston, MA, USA, <sup>4</sup>Baylor College of Medicine Houston, TX, USA, <sup>5</sup>Texas Children's Hospital Houston, TX, USA

#### Introduction:

As the survival rate from childhood tumors continues to improve, visual morbidity from childhood cancer is significant. Data are limited regarding visual and oculomotor outcomes in pediatric patients with posterior fossa tumors. We present these detailed outcomes in the largest pediatric cohort yet reported.

#### Methods:

A six year retrospective chart review of all patients with posterior fossa tumors was performed in the ophthalmology departments of two children's hospitals. Data including clinical presentation, tumor diagnosis and treatment, and ophthalmic findings from the most recent eye exam were recorded.

#### Results:

We identified 139 patients (42.4% female) with posterior fossa neoplasms. Mean age at diagnosis was 6.74 years, and mean ophthalmologic follow-up after tumor diagnosis was 20.5 months. Common ophthalmic signs and symptoms at presentation included esotropia (26.4%), diplopia (13.8%), and nystagmus (9.2%). The most frequent diagnoses were medulloblastoma in 65 patients (46.8%), juvenile pilocytic astrocytoma (JPA) in 45 patients (32.4%), and ependymoma in 14 patients (10.1%). Surgical resection occurred in 125 patients (89.9%) and 96 (69.0%) underwent radiation and/or chemotherapy. Visual outcomes were "good" (bilateral acuity of 20/20-20/40) in 101 patients (72.7%), "fair" (<20/40-20/200 in one or both eyes) in 12 patients (8.6%), and "poor" (<20/200 in one or both eyes) in 9 patients (6.5%). Medulloblastoma was the tumor type most commonly associated with optic nerve atrophy and a poor visual outcome. Thirty-two patients (23%) developed nystagmus and 59 (42.4%) developed strabismus after tumor resection, among whom 24 underwent eye muscle surgery for persistent diplopia.

#### Conclusions:

Although the majority of the patients in our cohort had good visual outcomes, 15.1% demonstrated significantly decreased visual acuity in one or both eyes, a finding most often associated with medulloblastoma. In addition, 17.3% of patients underwent eye muscle surgery to correct persistent diplopia as a result of their tumor or secondary to treatment.

**References:** None.

**Keywords:** Tumors, Nystagmus, Ocular Motility, Pediatric Neuro-Ophthalmology, Chemotherapy And Radiation Injury

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 42

### Radiation Optic Neuropathy and Retinopathy from Low Dose (20Gy) Radiation Treatment.

Crandall E. Peeler<sup>1</sup>, Dean M. Cestari

*Department of Neuro-ophthalmology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston, MA, USA*

#### Introduction:

The risk of optic neuropathy is thought to be low in patients receiving radiation to the anterior visual pathway in cumulative doses less than 50 Gy. We report a case of radiation optic neuropathy (RON) in a patient who received a total dose of 20 Gy, administered in two fractions.

#### Methods:

Case report.

#### Results:

A 44-year-old woman with a history of non-small cell lung cancer (NSCLC) presented with a three day history of painless vision loss OS. Visual acuity was 20/20 OD and 20/40 OS. There was dyschromatopsia and an afferent pupillary defect OS. Dilated fundus exam revealed swelling of the left optic nerve with adjacent nerve fiber layer hemorrhage. There was also a chorioretinal scar in the superior macula and scattered dot-blot retinal hemorrhages. MRI of the brain and orbits with gadolinium demonstrated enhancement of the left optic nerve but was otherwise normal. Five weeks following initial presentation, she developed sudden worsening of her vision OS and was noted to have neovascularization of the disc and a dense vitreous hemorrhage. Eighteen months prior to presentation, the patient underwent proton beam radiotherapy OS to treat a uveal metastasis of her NSCLC. The total radiation dose was 20 Gy, delivered in two 10 Gy fractions. Since that time, she had also been on maintenance chemotherapy for her underlying malignancy.

#### Conclusions:

Though radiation doses to the anterior visual pathway of less than 50 Gy are generally thought to be safe, the risk of RON increases when treatments are administered in fractions larger than 1.9 Gy<sup>1-2</sup>. Chemotherapy can also potentiate the effects of radiation<sup>3</sup>. Our patient developed radiation optic neuropathy, retinopathy, and neovascularization of the disc (a rare finding) following a relatively low total dose of proton beam radiation. This suggests that even low doses of radiation should be delivered in multiple, small fractions.

#### References:

1. Lessell S. Friendly fire: Neurogenic vision loss from radiation therapy. J Neuro-Ophthalmol 2004;4(3):243-250.
2. Levin LA, Gragoudas ES, Lessell S. Endothelial cell loss in irradiated optic nerves. Ophthalmology. 2000;107:370-4.
3. Fishman ML, Bean SC, Cogan DG. Optic atrophy following prophylactic chemotherapy and cranial radiation for acute lymphocytic leukemia. Am J Ophthalmol 1976;82:571-6.

**Keywords:** Chemotherapy And Radiation Injury, Optic Neuropathy, Tumors

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 43

### Bilateral Optic Neuropathy in Superficial Intracranial Siderosis.

Thong D Pham<sup>1</sup>, Mark J Morrow

*Harbor UCLA Medical Center/Neurology, Torrance, CA, USA*

#### Introduction:

Superficial intracranial siderosis (SIS) results from chronic subarachnoid hemorrhage. It can cause anosmia, sensorineural hearing loss, cerebellar dysfunction, pyramidal signs and dementia. Optic neuropathy is a rare complication of this disorder.

#### Methods:

A 60 year old man with hypertension and past alcohol and heroin abuse presented with three years of worsening bilateral visual and hearing loss, falls and cognitive problems. There was no history of severe headache or acute symptomatology. Examination showed acuities of 20/30 OD and 20/50 OS with flat, atrophic discs. There was moderate bilateral limb and gait ataxia and evidence of mild dementia.

#### Results:

Automated perimetry showed inferior altitudinal loss with superior arcuate scotoma OU (mean deviations: -8.31 db OD, -22.81 db OS). OCT demonstrated bilateral superior-predominant peripapillary RNFL loss (mean thickness: 77 microns OD, 58 microns OS). MRI showed evidence of over 30 cerebral and brainstem cavernomas, the largest of which abutted the right frontal horn. Extensive leptomeningeal hypodensity was seen on gradient echo (GrE) imaging, including the basilar cisterns, perichiasmatic region and optic nerve sheaths. Both intracranial optic nerves also showed hypodensity on GrE, implying intraparenchymal hemosiderin deposition. Ventricles were mildly enlarged. Lumbar picture showed acellular CSF with mildly elevated protein and a normal opening pressure of 11 cm H<sub>2</sub>O.

#### Conclusions:

We attributed our patient's optic neuropathy to SIS from recurrent intraventricular and subarachnoid hemorrhage due to periventricular cavernoma. It is important to suspect this diagnosis and order imaging accordingly in patients with an appropriate history.

#### References:

1. Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. *Brain* 1995;118:1051-66
2. Kumar N, Cohen-Gadol AA, Wright RA et al. Superficial siderosis. *Neurology* 2006;66:1144-521.
3. Painter SL, Mathew L, Quaghebeur G et al. Optic neuropathy in superficial intracranial siderosis. *J Neuroophthalmol* 2010;30:311-4

**Keywords:** Optic Neuropathy, Diagnostic Tests (OCT), Visual Fields, Neuroimaging, Vascular Disorders

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 44

### **Binocular Acuity Summation (BAS) in Multiple Sclerosis (MS): Relation to Retinal Architecture and Visual System Neurophysiology**

Sara S Qureshi<sup>1</sup>, Millad Sobhanian<sup>1</sup>, Amy Conger<sup>1</sup>, Darrel Conger<sup>1</sup>, Teresa C Frohman<sup>1</sup>, Laura J Balcer<sup>2</sup>, Elliot M Frohman<sup>1</sup>

<sup>1</sup>University of Texas Southwestern, Department of Neurology, Dallas, TX, USA, <sup>2</sup>NYU Lagone Medical Center, Department of Neurology, Population health and Ophthalmology, New York, NY, USA

#### **Introduction:**

The afferent visual pathway (AVP) is commonly involved in MS and neuromyelitis optica (NMO). Patients with/without history of acute optic neuritis (ON) can have inter-ocular differences (IOD) in visual acuity (VA), especially at low-contrast levels, which interfere with BAS. BAS is defined as the degree to which binocular vision is greater than VA of the better eye. Reductions in binocular visual functioning may impair depth/motion perception with important quality of life ramifications.<sup>(1,2,3)</sup> We determined the relation of BAS to scores for structural, functional and electrophysiological measures of the AVP in an MS/NMO cohort.

#### **Methods:**

High and low-contrast letter acuities (LCLA) were assessed binocularly and monocularly in MS/NMO patients and disease-free controls. Clinically significant BAS was defined as  $\geq 7$  letters (level beyond test-retest variability) improvement upon binocular versus monocular assessment. Spectral domain optical coherence tomography (OCT) and scanning laser polarimetry (GDx) were performed along with visual evoked potentials (VEP) and multifocal VEP (mfVEP).

#### **Results:**

Analyses at low-contrast levels (1.25% and 2.5%) in the MS and combined (MS+NMO+control) groups showed greater ability to summate with lower IOD in VA, retinal nerve fiber layer (RNFL) thickness as measured by OCT and GDx, and mfVEP1 ( $p < 0.05$  using logistic regression models accounting for age;  $p = 0.06$  for GDx in MS group at 1.25%). Frequencies of BAS were 4.8% for high-contrast, 38.7% for 2.5%LCLA and 56.5% for 1.25%LCLA. Among 'summators' maximum IOD for RNFL thickness was 8  $\mu\text{m}$  for high-contrast, 12  $\mu\text{m}$  for 2.5%LCLA, and 34  $\mu\text{m}$  for 1.25%LCLA.

#### **Conclusions:**

BAS, under low contrast conditions, occurs more frequently and despite greater IOD of RNFL thickness. This may reflect the superior sensitivity of LCLA compared to high-contrast VA to detect perceived visual disability among patients with unilateral ON. Conversely, this may suggest that under low contrast conditions binocular visual function is less sensitive to inter- eye differences.

#### **References:**

1. S.P. Azen, R. Varma, S. Preston-Martin, M. Ying-Lai, D. Globe, S. Hahn. Binocular visual acuity summation and inhibition in the ocular epidemiological study: the Los Angeles Latino eye study. Invest Ophthalmol Vis Sci, 43 (6) (2002), pp. 1742–1748
2. Stacy L., Pineles, Eileen E. birch, Lauren S. Talman et al. One Eye or Two: A Comparison of Binocular and Monocular Low-Contrast Acuity Testing in Multiple Sclerosis: Am J Ophthalmol. 2011 July; 152(1): 133–140
3. N.J. Newman, J.M. Wolfe, M.I. Steward, S. Lessell. Binocular visual function in patients with a history of monocular optic neuritis. Clin Vision Sci, 6 (2) (1991), pp. 95–107

**Keywords:** Binocular Acuity Summation, Low Contrast Visual Acuity, Optical Coherence Tomography, Scanning Laser Polarimetry, Visual Evoked Potentials

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 45

### Nutritional Optic Neuropathy after Bariatric Surgery

Yuna Rapoport<sup>1</sup>, Patrick Lavin

*Vanderbilt Medical Center/ Ophthalmology, Nashville, TN, USA*

#### Introduction:

We present a case of a patient with nutritional optic neuropathy attributed to copper deficiency as a result of bariatric surgery.

#### Methods:

Retrospective case report.

#### Results:

A 47-year-old woman developed progressive blurred central vision. Four weeks after onset of decreased vision, she developed formed, nonthreatening visual hallucinations. Her husband also noticed that she had mild cognitive dysfunction and some falls for 2 months prior to presentation. Her past surgical history was significant for vertical banded gastroplasty 24 years prior, Rouy-en-Y gastric bypass 4 years prior, and a recent C6-7 fusion complicated by pneumonia requiring intubation. Her exam was significant for visual acuity of 20/400 each eye, impaired color vision (Ishihara plates OD: 3/14, OS: 1/14), bilateral central scotomas, and mild temporal pallor of each optic disc. Optical coherence tomography was significant for thinning of the temporal retina bilaterally. Blood tests for vitamin B1, B6, and B12, methylmalonic acid, and red cell folate were sent. The B12 level was elevated but she had an elevated methylmalonic acid level, and a low serum copper level, confirming a nutritional optic neuropathy. Five weeks after beginning copper tablets and B12 injections, the patient reported improvement in her vision and five months after beginning treatment her best corrected visual acuity was 20/25 with each eye. We briefly review a differential diagnosis for bilateral blurred vision with central scotomas including other toxic and nutritional optic neuropathies to put this copper deficiency case in perspective.

#### Conclusions:

Gradual, progressive, painless, bilaterally symmetric visual loss in a patient with a history of a gastric bypass procedure should prompt evaluation for nutritional optic neuropathy to prevent permanent severe optic atrophy. The visual hallucinations likely were a variant of the Charles Bonnet syndrome as a result of relative visual deprivation.

#### References:

1. Becker, DA, Balcer, LJ, Galetta, SL. The Neurological Complications of Nutritional Deficiency following Bariatric Surgery. J Obesity, 608534: 1-8; 2012.
2. Epidemic optic neuropathy in Cuba: clinical characterization and risk factors. The Cuba Neuropathy Field Investigation Team. N Engl J Med, 333 (18):1176-1182; 1995.
3. Naismith RT, Shepherd, JB, Cross, AH. Acute and bilateral blindness due to optic neuropathy associated with copper deficiency. Archives of Neurology, 66 (8): 1025–1027; 2009.
4. Gratton, SM, Lam, BL. Visual loss and optic nerve head swelling in thiamine deficiency without prolonged dietary deficiency. Clin Ophthalmol, 8: 1021-1024; 2014.
5. Rovner, BW. The Charles Bonnet syndrome: A review of recent research. Curr Opin Ophthalmol, 17:275-7;2006.

**Keywords:** Optic Neuropathy, Visual Fields, Higher Visual Functions, Neuro-Ophth & Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 46

### Lyme Disease Mimicking Giant Cell Arteritis

Nailyn Rasool<sup>1</sup>, Lisa Cowan, Dean M. Cestari

*Neuro-ophthalmology, Massachusetts Eye & Ear Infirmary, Harvard Medical School, Boston, MA, USA*

#### Introduction:

Steere et al first described a patient who went blind from severe panophthalmitis from *Borrelia burgdorferi* in 1985<sup>1</sup>. Since then, a spectrum of afferent and efferent manifestations have been reported secondary to Lyme borreliosis including optic neuritis, peri-neuritis and cranial nerve palsies that have expanded our understanding of the neuro-ophthalmic manifestations of this treatable disease.

#### Methods:

Case Report

#### Results:

We report a case of a 74 year old Caucasian woman who developed severe headaches over the left forehead and temporal region. As part of her work up at an outside hospital, her brain MRI without contrast was normal. Two weeks later she developed a lower motor neuron facial palsy in association with scalp tenderness, shoulder and neck pain. ESR and CRP were reported to be elevated and she was started on prednisone 70mg/day for presumed GCA. A temporal artery biopsy was negative and prednisone was discontinued. One month later, she developed jaw claudication and an inferior altitudinal defect. ESR was 73mm/hr and CRP was 48.6 and prednisone was restarted for presumed GCA with complete resolution of her systemic symptoms. Two months later while taking prednisone 35mg/day, she developed iritis OS and a swollen optic nerve with an inferior altitudinal defect OD. MRI orbit/brain with gadolinium was normal. ESR was 34mm/hr and CRP 2. Blood work for Lyme disease showed 9/10 positive bands IgG and 3/3 positive bands IgM. CSF had 8 WBCs with normal protein and glucose. CSF Lyme IgM and IgG were positive.

#### Conclusions:

Ocular manifestations of Lyme disease can involve any ocular structure. Neuro-ophthalmologists often include Lyme disease in the differential diagnosis of optic neuritis and perineuritis in endemic regions. This case demonstrates that this infection can mimic the signs and symptoms of GCA and helps to broaden the clinical spectrum of neuro-ophthalmic manifestations of Lyme disease.

#### References:

1. Steere AC, Duray PH, Kauffmann DJ, Wormser GP. Unilateral blindness caused by infection with the Lyme disease spirochete, *Borrelia burgdorferi*. *Ann Intern Med*. 1985 Sep;103(3):382-4.

**Keywords:** Lyme Disease, Giant Cell Arteritis, Optic Neuritis, Headache

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 47

### Structural analyses of the anterior visual pathway in compressive neuropathy

Won Hyung A. Ryu<sup>1</sup>, Wesley Chan<sup>1</sup>, Jodie M Burton<sup>1</sup>, Yves Starreveld<sup>1</sup>, Fiona Costello<sup>1,2</sup>

<sup>1</sup>Department of Clinical Neurosciences, Calgary, AB, Canada, <sup>2</sup>Department of Surgery, Calgary, AB, Canada

#### Introduction:

Pituitary tumors are one of the most common types of brain tumors that can cause progressive blindness. Due to the indolent nature of the vision loss, these patients are often misdiagnosed which can turn a reversible cause of visual dysfunction into a permanent life-altering disability. Adding to the difficulty in treating patients with pituitary tumors is the lack of validated preoperative predictors of visual outcome. The purpose of this study was to determine whether structural changes in the anterior visual pathway differ in patients with good versus poor visual recovery.

#### Methods:

25 patients (15F: 10M) with radiographic diagnosis of pituitary macroadenomas underwent a neuro-ophthalmic evaluation and spectral-domain optical coherence tomography (OCT) testing pre-operatively and 6-months after surgery. Pre-operative retinal nerve fiber layer thickness (RNFLT) and macular volume (MV) were compared between patients with normalized visual function versus persistent visual deficits after surgery (visual acuity  $\leq 20/40$ ; visual field mean deviation  $\leq -5.0$  decibels).

#### Results:

There was a significant difference in pre-operative MV (9.9 vs 9.4mm<sup>3</sup>;  $p = 0.018$ ) when comparing patients with good versus poor visual outcome. Even after complete surgical decompression of the pituitary tumor, patients with persistent visual deficits had significantly greater thinning of the RNFLT at 6 months follow-up (-1.9 vs -7.9 $\mu$ m;  $p = 0.0002$ ).

#### Conclusions:

Structural changes seen in the anterior visual pathway may provide new predictive measures of visual outcomes after surgical resection of pituitary tumor. Specifically, visual recovery may be dependent on whether surgical decompression occurs before a critical threshold is reached in patients with compressive neuropathy.

**References:** None.

**Keywords:** Tumors, Neuroimaging, Diagnostic Tests, Visual fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Changes In Thickness Of Retinal Segments In The Early Course Of Non-Arteritic Ischaemic Optic Neuropathy

Bernardo F. Sanchez-Dalmau<sup>1,2</sup>, Johannes Keller<sup>1,2</sup>, Pablo Villoslada<sup>2</sup>

<sup>1</sup>Hospital Clinic. Ophthalmology Department, Barcelona, Spain, <sup>2</sup>idibaps. Center Of Neuroimmunology, Barcelona, Spain

### Introduction:

Purpose: To characterise changes of the retinal layers in patients with anterior ischaemic optic neuropathy (AION) aiming to identify imaging markers for predicting the residual visual function

### Methods:

Retrospective review of consecutive patients with unilateral AION from January 2010 to December 2013. Case-control analysis of affected eyes at baseline and 1 month later, and fellow eyes. Algorithmic segmentation in layers and division in ETDRS quadrants of optical coherence tomography images of the macula. Regression analysis of retinal layer thickness and best corrected visual acuity (BCVA) in logMAR units and mean deviation of the visual field (VF)

### Results:

20 eyes from 20 patients were included in the case-control analysis. At baseline we found significant thickening of the retinal nerve fibre layer (RNFL) at 42.2µm compared to 37.9µm in control eyes ( $p=0.002$ ). The outer nuclear layer (ONL) was also significantly thickened at 96.6µm compared to 90.8µm in the fellow eye ( $p<0.001$ ). After 1 month the RNFL and the ganglion cell layer (GCL) were thinned 17.7% (to 31.2µm,  $p<0.001$ ) and 19.3% (to 66.5µm,  $p<0.001$ ) compared to the contralateral eye. Additionally, the ONL remained thickened at 96.7µm ( $p<0.001$ ). At baseline, we found a significant correlation between the ONL thickness and the VF ( $r= -0.482$ ,  $p=0.005$ ) and the BCVA at discharge ( $r= 0.552$ ,  $p<0.001$ ), as well as between the GCL thickness and the BCVA at discharge ( $r=0.411$ ,  $p=0.02$ ). At the 1 month time point the GCL thinning was correlated with both the VF ( $r=0.471$ ,  $p=0.005$ ) and the BCVA ( $r=-0.456$ ,  $p=0.007$ )

### Conclusions:

Changes in thickness of different layer of the retina occur early in the course of AION and evolve in time resulting in the atrophy of the GCL and RNFL. Very early thickening of the OPL is the earliest predictor of residual visual dysfunction. Thinning of the GCL after 1 month correlates with poorer final outcomes.

**References:** None.

**Keywords:** Retinal Ganglion Cell, Ganglion Cell Layer, Ischemic Optic Neuropathy, Optical Coherence Tomography, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 49

### A First Case Report Of Chordoid Glioma Invading Optic Nerve

Nicolae Sanda<sup>1,2</sup>, Avinoam B Safran<sup>1,3</sup>, Claudiu N Mircea<sup>4</sup>, Michèle Bernier<sup>5</sup>, Sorin Aldea<sup>6</sup>

<sup>1</sup>Sorbonne Universités, UPMC Univ Paris 06, UMR\_S 968, Institut de la Vision, INSERM, U968, CNRS, UMR\_7210, Paris, France, <sup>2</sup>Neurology Department, Hôpital Foch, Suresnes, France, <sup>3</sup>Neurosciences, Geneva University School of Medicine, Geneva, Switzerland, <sup>4</sup>Radiology Departement, CHU de Versailles Versailles, France, <sup>5</sup>Pathology Departement, Hôpital Foch Suresnes, France, <sup>6</sup>Neurosurgery Departement, Hôpital Foch Suresnes, France

#### Introduction:

Chordoid glioma of the third 3<sup>rd</sup> is a rare, grade II glial tumor, that usually presents a poor outcome due to it's vicinity to hypothalamus and visual pathways. It is considered non-invasive.

#### Methods:

Case report.

#### Results:

A 39 yo male with unnoticeable medical history, presented with bi-temporal visual field defects, headache and sleep disorder. Right eye (RE) visual acuity was 1, left eye (LE) visual acuity 0.9; intraocular pressure was in normal range and pupils were isochoric. He had a right relative afferent pupillary defect and bilateral optic disc pallor. Humphrey perimetry showed bi-temporal hemianopia, more extensive in the RE. Except for a slightly diminished prolactine level and thyreotrop insufficiency all hypothalamic and pituitary hormones were in normal range. MRI revealed relatively homogenous mass developed in the anterior part of the 3<sup>rd</sup> ventricle with MRI signal, T1 isointense/ T2 hyperintense, exhibiting strong and uniform gadolinium enhancement that seemed to invade the right optic nerve. One T2\*GRE slice presented a hypointense spot suggestive of micro-hemorrhage or calcification. There was no sign of hydrocephalus. Surgery revealed a tumor inserted into the anterior part of the 3<sup>rd</sup> ventricle floor and adherent to the lateral walls of the 3<sup>rd</sup> ventricle and to anterior cerebral arteries. It invaded the chiasm and the right optic nerve dividing its fibers up to the lateral aspect. Excision was subtotal. The tumor exhibited typical features of chordoid glioma; GFAP, CD34 were highly positive, NF was negative and Ki67 was 2-3%. No other oncological treatment was delivered. Thyroid hormones deficit aggravated after surgery and the patient developed cortisonic deficiency requiring substitution. Residual tumor remained stable and visual field improved over a period of 18 months of follow-up.

#### Conclusions:

This is a first case report of a chordoid glioma invading optic nerve.

#### References:

1. Desouza R-M, Bodi I, Thomas N, Marsh H, Crocker M. Chordoid glioma: ten years of a low-grade tumor with high morbidity. Skull Base. 2010 Mar;20(2):125–138.
2. Al-Zubidi N, McGlynn MM, Chévez-Barrios P, Yalamanchili S, Lee AG. Neuro-ophthalmologic features of chordoid glioma. J Neuro-Ophthalmol. 2014 Mar;34(1):47–49.
3. Wilson JL, Ellis TL, Mott RT. Chordoid meningioma of the third ventricle: a case report and review of the literature. Clin Neuropathol. 2011 Apr;30(2):70–74.

**Keywords:** Chordoid Glioma, Optic Nerve

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Labex and Humanis Grants support for N. Sanda and A.B. Safran

## Poster 50

### Clinical spectrum of optic neuritis in Indian children at a tertiary care centre

Swati Phuljhele<sup>1</sup>, Reena Sharma<sup>1</sup>, Rohit Saxena<sup>1</sup>, Vimla Menon<sup>2</sup>

<sup>1</sup>All India Institute of medical Sciences, New Delhi, India, <sup>2</sup>Vision eye centre, New Delhi, India

#### Introduction:

To study the clinical spectrum of pediatric optic neuritis in Indian children

#### Methods:

A prospective study of patients diagnosed as optic neuritis below 20 yrs of age at a tertiary care centre. 28 cases (44 eyes) were reviewed. The ophthalmological examination findings, Goldmann perimetry, electrophysiological tests and MRI were analysed.

#### Results:

The study patients had a mean age of  $12.64 \pm 4.95$  yrs and were followed up for a mean period of  $6.38 \pm 4.99$  months. 32.1% were females (F: M – 0.47). Six patients (20.7%) had a viral prodrome and 12 (42.9%) complained of painful ocular movements. The mean baseline visual acuity was  $1.80 \pm 0.6$  logMAR units and the last best visual acuity was  $0.43 \pm 0.4$  log units. Thirty six eyes (81.8%) had a visual acuity of 3/60 or lower and 40 eyes (90.9%) had 6/60 or lower of the snellen chart at the time of presentation. Sixteen patients (57.1%) had a bilateral presentation and 20 (71.4%) had papillitis. All patients were given intravenous steroid treatment. At one wk, 21 out of 44 eyes (47.7%) had a visual acuity  $\geq 6/18$  of the snellen chart. Three patients showed demyelination changes on MR imaging of brain. Those below 10 yrs of age presented more commonly with papillitis (p value = 0.01).

#### Conclusions:

Pediatric optic neuritis patients have poorer initial visual acuity, but are associated with a faster recovery and a better visual outcome. There are no gender differences in either the presentation or the visual recovery. Papillitis was more often seen in children below 10 yrs of age.

#### References:

(1) Beck RW, Cleary PA. Optic neuritis treatment trial. One-yr follow-up results. Arch Ophthalmol. 1993;111(6):773-5. Berhman RE, Kliegman R, Arvin AM, Nelson WE. Nelson Textbook of Pediatrics, 15th Ed. Philadelphia: W.B. Saunders Company; 1996. (2) Rudolph AM, et al. Rudolph's Pediatrics, 21st Ed. New York: McGraw-Hill; 2002.; (3) Avery MD, First LR. Pediatric Medicine, 2nd Ed. Baltimore: Williams & Wilkins; 1994.; (4) Polman CH, Reingold SC, Banwell B, Clanet M, Cohen JA et al. Diagnostic criteria for multiple sclerosis: 2010 revisions to the McDonald criteria. Ann Neurol. 2011;69(2):292-302.; (5) Menon V, Mehrotra A, Saxena R, Jaffery NF. Comparative evaluation of megadose methylprednisolone with dexamethasone for treatment of primary typical optic neuritis. Indian J Ophthalmol. 2007;55(5):355-9.; (6) Visudhiphan P, Chiemchanya S, Santadusit S. Optic neuritis in children recurrence and subsequent development of multiple sclerosis. Pediatr Neurol. 1995 No;13(4):293-5.; (7) KENNEDY C, CARROLL FD. Optic neuritis in children. Arch Ophthalmol. 1960;63:747-55.; (8) Absoud M, Cummins C, Desai N et al. Childhood optic neuritis clinical features and outcome. Arch Dis Child. 2011;96(9):860-2.; (9) Wilejto M, Shroff M, Buncic JR, Kennedy J, Goia C, Banwell B. The clinical features, MRI findings, and outcome of optic neuritis in children. Neurology. 2006;67(2):258-62.; (10) Hwang JM, Lee YJ, Kim MK. Optic neuritis in Asian children. J Pediatr Ophthalmol Strabismus. 2002;39(1):26-32.; (11) Kriss A, Francis DA, Cuendet F, Halliday AM, Taylor DS et al. Recovery after optic neuritis in childhood. J Neurol Neurosurg Psychiatry. 1988;51(10):1253-8.; (12) Brady KM, Brar AS, Lee AG, Coats DK, Paysse EA, Steinkuller PG. Optic neuritis in children: clinical features and visual outcome. J AAPOS. 1999;3(2):98-103.; (13) Morales DS, Siatkowski RM, Howard CW, Warman R. Optic neuritis in children. J Pediatr Ophthalmol Strabismus. 2000 ;37(5):254-9.; (14) Jo DH, Kim SJ, Chae JH, Yu YS. The clinical characteristics of optic neuritis in Korean children. Korean J Ophthalmol. 2011;25(2):116-20.; (15) Cassidy L, Taylor D. Pediatric optic neuritis. J AAPOS. 1999;3(2):68-9.Review.; (16) Taylor D, Cuendet F. Optic neuritis in childhood. In: Hess RF, Plant GT, editors. Optic neuritis. Cambridge: Cambridge University Press; 1986. p. 73-85.; (17) Lucchinetti CF, Kiers L, O'Duffy A et al. Risk factors for developing multiple sclerosis after childhood optic neuritis. Neurology 1997;49: 1413-8.

**Keywords:** Optic Neuritis, Multiple Sclerosis, Vision, Papillitis, Dexamethasone

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 51

### Prognosticating factors in nonarteritic ischaemic optic neuropathy

Swati Phuljhele<sup>1</sup>, Reena Sharma<sup>1</sup>, Rini Sinha<sup>1</sup>, Rohit Saxena<sup>1</sup>, Vimala Menon<sup>2</sup>

<sup>1</sup>All India Institute of medical Sciences, New Delhi, India, <sup>2</sup>Vision eye centre, New Delhi, India

#### Introduction:

To review prognosticating factors in non arteritic ischaemic optic neuropathy (NAION)

#### Methods:

A prospective observational study of 27 NAION patients and 20 controls. Visual acuity, colour vision, contrast sensitivity, slitlamp biomicroscopy, visual evoked potential, Goldmann visual fields, optical coherence tomography, and fundus fluorescein angiography, were done and repeated at 1 and 3 months.

#### Results:

There were 16 males (59.3%) and 11 females (40.7%) with a mean age of 52.9 +/- 8.4 years. The right eye was affected in 20 (74.1%) patients and five patients (18.5%) had bilateral NAION. The mean presenting visual acuity was 1.5 ± 0.1 logMAR units with 14 patients (51.8%) above 50 years of age. 11 patients (40.7%) had no systemic associations. The remaining had diabetes (14.8%), hypertension (18.5%), both (18.5%) and hyperlipidemia (7.5 %). Visual parameters improved significantly at 1 month follow up (pvalue 0.002, 0.01 and 0.01 respectively) with no further improvement at 3 months (pvalue 0.08, 1.0 and 0.2 respectively). Thirteen patients (48.2%) showed spontaneous visual recovery on follow up. Mean baseline visual parameters were similar in patients with or without systemic disease (P value= 0.2), but the latter improved more on follow up (P value 0.03). Patients with a central scotoma had poorer vision (p value 0.01) and a larger number improved (P value 0.09). Presenting visual acuity and age did not affect the final visual outcome (pvalue 0.2 and 0.7 respectively). There was no significant difference in the VER and optic nerve head parameters at presentation or on follow up between the patients with and without visual recovery.

#### Conclusions:

NAION patients in Asia are younger with maximum visual recovery in the first month. Spontaneous visual improvement is seen in about half of the patients. Patients with systemic disease and central scotoma have a poorer visual prognosis. Presenting visual acuity, age and gender do not affect visual outcome.

**References:** None.

**Keywords:** Optic Neuropathy, NAION, Prognosticating, Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 52

### Sweep My Blindness Away!

Salwa Abdel Aziz<sup>1</sup>, Tarek A Shazly<sup>1</sup>, Islam M Zaydan<sup>1</sup>, Valeria Fu<sup>1,2</sup>, Gabrielle R Bonhomme<sup>1</sup>

<sup>1</sup>University of Pittsburgh, Department of Ophthalmology, Pittsburgh, PA, USA, <sup>2</sup>Children's Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

#### Introduction:

Optic neuropathy secondary to Antiphospholipid antibody syndrome (APS) can result in severe vision loss.

#### Methods:

Case report.

#### Results:

A 61-year-old female presented to our neuro-ophthalmology clinic with bilateral subacute painless vision loss. Best corrected acuity was 20/30 bilaterally with normal color vision and papillary examination. Fundus examination revealed normal optic nerves. Humphrey visual fields revealed bilateral central scotomas. Brain MRI with contrast was normal. Serologies revealed positive antiphospholipid antibodies which remained positive on repeat testing. Hematology started 81mg aspirin. Three months later, her acuity worsened to 20/40 bilaterally with inferior nasal field loss OS. CSF was normal and she received IV methylprednisolone for 3 days. Two weeks later she developed inferior nasal field loss in the right eye. Within 3 months her visual fields deteriorated, with bilateral dense inferior altitudinal field loss with central vision of 20/50 bilaterally. Hematology started Plasmapheresis. Her visual fields continued to worsen with peripheral constriction in both eyes with relative sparing of the central 10 degrees. Given her deteriorating vision, Hematology started IVIG despite the absence of systemic involvement of APS. Despite 3 IVIG infusions, central vision worsened to 20/70 bilaterally. Coumadin was contraindicated for the concern of fall risk with her poor vision. In spite of the progressive worsening of vision, no signs of optic nerve examination remained normal and Optical coherence Tomography revealed no RNFL, macular or ganglion cell thinning. Given the discrepancy between her visual testing and physical findings we obtained sweep and pattern Visual evoked potentials (VEPs). Pattern VEP was normal bilaterally and SVEP measured visual acuity of 20/16 OD and 20/17 OS.

#### Conclusions:

Early diagnosis of confounding functional visual loss allows proper patient management and avoidance of unnecessary and potentially harmful treatments. VEPs offer a means of objectively estimating acuity and therefore could assist with early and accurate diagnosis.

#### References:

1. Giorgi, D., and C. Balacco Gabrieli. "Optic neuropathy in systemic lupus erythematosus and antiphospholipid syndrome (APS): clinical features, pathogenesis, review of the literature and proposed ophthalmological criteria for APS diagnosis." *Clinical rheumatology* 18.2 (1999): 124-131.
2. Tyler, Christopher W., et al. "Rapid assessment of visual function: an electronic sweep technique for the pattern visual evoked potential." *Investigative ophthalmology & visual science* 18.7 (1979): 703-713.
3. Almoqbel, Fahad, Susan J. Leat, and Elizabeth Irving. "The technique, validity and clinical use of the sweep VEP." *Ophthalmic and Physiological Optics* 28.5 (2008): 393-403.
4. Norcia, Anthony M., et al. "Measurement of spatial contrast sensitivity with the swept contrast VEP." *Vision research* 29.5 (1989): 627-637.

**Keywords:** VEP, Sweep, Antiphospholipid, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 53

### Progressive Non-Arteritic Optic Neuropathy As The First Presentation Of Anti-Phospholipid Antibody Syndrome

Tarek A Shazly<sup>1</sup>, Gabrielle R Bonhomme

*University of Pittsburgh, Department of Ophthalmology, Pittsburgh, PA, USA*

#### **Introduction:**

To report the clinical presentation and visual outcomes of non-arteritic anterior ischemic optic neuropathy (NAION) with anti-phospholipid antibody syndrome (APS).

#### **Methods:**

A non-comparative consecutive case series. Electronic medical records were retrospectively reviewed for all patients with NAION seen at our institution with progressive vision loss with positive APS antibodies between 2001 and 2014. The corresponding clinical records were then reviewed to evaluate the clinical features, laboratory results and visual outcomes.

#### **Results:**

Four patients were identified, two of the patients were females and 2 males with a mean age of 63.3 +/- 7.9 years. All patients initially presented with classic altitudinal visual field loss above or below the horizontal midline, with swelling and hemorrhage of the corresponding portion of the optic nerve typical of NAION. Presenting Snellen visual acuity in the affected eye at the time of the first episode of NAION ranged from 20/20 to 20/30, while visual acuity at the time of the second episode ranged from 20/20 to counting fingers. Examination at the time of the second episode revealed ipsilateral visual field loss in their previously unaffected hemi-field, with new, acute optic nerve hemorrhages and disk edema corresponding to the new visual field loss. The interval between the first and the second episodes in the same eye ranged from 15 to 71 days (mean 37 days, SD 25.37 days).

#### **Conclusions:**

Patients with NAION, particularly in the setting of progressive or recurrent vision loss, should be tested for APS and promptly treated to avoid progressive severe vision loss. NAION secondary to APS leads to severe progressive vision loss and requires close monitoring and prompt control vs treatment of the underlying hematological disorder.

#### **References:**

1. Giorgi, D., and C. Balacco Gabrieli. "Optic neuropathy in systemic lupus erythematosus and antiphospholipid syndrome (APS): clinical features, pathogenesis, review of the literature and proposed ophthalmological criteria for APS diagnosis." *Clinical rheumatology* 18.2 (1999): 124-131.
2. Watts, M. T., et al. "Antiphospholipid antibodies in the aetiology of ischaemic optic neuropathy." *Eye* 5.1 (1991): 75-79.
3. Reino, S., et al. "Optic neuropathy in the "primary" antiphospholipid syndrome: report of a case and review of the literature." *Clinical rheumatology* 16.6 (1997): 629-631.
4. Srinivasan, Sathish, et al. "Reversal of nonarteritic anterior ischemic optic neuropathy associated with coexisting primary antiphospholipid syndrome and Factor V Leiden mutation." *American journal of ophthalmology* 131.5 (2001): 671-673.
5. Cordeiro, M. F., et al. "Ischaemic optic neuropathy, transverse myelitis, and epilepsy in an anti-phospholipid positive patient with systemic lupus erythematosus." *Journal of neurology, neurosurgery, and psychiatry* 57.9 (1994): 1142.

**Keywords:** Antiphospholipid, Optic, Neuropathy, NAION, Visual

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 54

### Anterior Visual Pathway(AVP) Meningiomas: A Dosimetric comparison of IMRT to Pencil-beam Proton Therapy

Scott L Stafford<sup>1</sup>, Kevin L Kisrow, Charles S Mayo

*Mayo Rochester /Radiation Oncology, Rochester, MN, USA*

#### Introduction:

Introduction: Intensity Modulated Radiation Therapy (IMRT) is the standard technique to minimize dose to nearby organs at risk (OAR) when treating tumors involving or abutting the Optic Nerve. Proton therapy may further decrease the dose to OAR while maintaining primary target coverage. A dosimetry study compared IMRT photons to pencil-beam protons to evaluate the utility of protons in these tumors. Methods: Four representative tumors involving the AVP, treated with IMRT were planned with pencil-beam proton planning. Equivalent target coverage and OAR dose- volume restrictions were applied to both modalities, and dosimetric outcomes compared. Patient 1: large orbital schwannoma. Patients 2,3,4: orbital optic nerve sheath, clinoid, and cavernous sinus meningiomas. Results: Patient 1: ipsilateral retina V20, and mean dose were significantly less with protons. Chiasm dose was substantially less with protons. Pituitary mean dose was substantially less with protons Patient 2 and 3: Ipsilateral retina, pituitary doses, and brain V5Gy, all received significantly less dose with protons. Chiasm dose was no different in patient 2 (due to proximity to target volume), but markedly reduced in patient 3. Pt four: Chiasm V40Gy, Pituitary mean and maximum dose, and ON V40 are all less with proton therapy. Brain V5 was less for protons (50%-70%) in all patients. Conclusions: Pencil beam protons decrease dose to OAR in all but small apical tumors. OAR 1cm to 5cm distant to target volumes receive substantially less dose with protons but clinically relevant differences with IMRT is variable with cavernous sinus and para-clinoid tumors benefiting for more OAR. Brain dose is much less with protons (V5Gy). Protons should be considered for large benign tumors involving the AVP where OAR dose differences impact long term toxicity risk. Small apical tumors can be treated with IMRT photons. Reduced brain dose theoretically may reduce second malignancies.

**References:** None.

**Keywords:** IMRT, Proton Therapy, Dosimetric Comparisons, Toxicity, AVP Tumors

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 55

### A Case Of Lyme Neuroretinitis- An Elusive Entity

Padmaja Sudhakar<sup>1</sup>, Muhammad Zafar<sup>1</sup>, Flavius Raslau<sup>2</sup>, Joshua Chalkley<sup>1</sup>, Kimberly Jones<sup>1</sup>, Sachin Kedar<sup>3</sup>

<sup>1</sup>University Of Kentucky, Dept Of Neurology, Lexington, KY, USA, <sup>2</sup>University Of Kentucky, Dept of Neuroradiology, Lexington, KY, USA, <sup>3</sup>University Of Nebraska Medical Center, Dept of Neurology, Omaha, NE, USA

#### Introduction:

Lyme disease is a multi-organ systemic infectious disease caused by *Borrelia burgdorferi*. Neuro-ophthalmic manifestations include papilledema, papillitis, retrobulbar neuritis and neuroretinitis. Using strict diagnostic criteria modified from the CDC surveillance criteria, there are very few cases of "Lyme" optic neuropathy (other than papilledema) in literature. We present the clinical, serological and radiological characteristics of a patient with Lyme neuroretinitis.

#### Methods:

Single case report and review of literature

#### Results:

A 10-year-old boy presented with bilateral progressive loss of vision. 2 months prior to presentation, he had suffered multiple tick bites that resulted in several "ring" rashes on his body. 1 month later he developed headaches, vomiting, neck stiffness, bilateral blurred vision and binocular horizontal diplopia. At presentation, exam revealed visual acuity of 20/200 in both eyes, left 6th nerve palsy and bilateral disc edema with macular exudation. He had constricted visual fields and central scotoma on confrontation testing. Contrast enhanced MRI orbit showed bilateral prominent focal nodular enhancement of the optic discs. CSF opening pressure was > 55mm Hg; he had lymphocytic pleocytosis (17 cells) with normal glucose and protein. Western blot of CSF and serum confirmed Lyme. Testing for all other infections including serum and CSF RPR were negative. He received a course of ceftriaxone with acetazolamide following which his visual acuity and fundus exam improved but developed bilateral disc pallor.

#### Conclusions:

Our patient met Category II criteria (Sibony et al) for optic neuropathy from Lyme's disease. Using these criteria, only 11 cases of Lyme optic neuropathy were reported in a recent review on the topic (Traisk et al). This patient likely suffered from papilledema secondary to meningitis as well as neuroretinitis, both resulting from Lyme disease. Focal nodular optic disc enhancement on MRI orbits was suggestive of optic disc inflammation

#### References:

1. Bhatti et al, Optic Neuropathy Secondary to Cat Scratch Disease: Distinguishing MR Imaging Features from Other Types of Optic Neuropathies, AJNR26:1310–1316, June/July 2005
2. Traiska F, Lindquist L. Optic nerve involvement in Lyme disease Curr Opin Ophthalmol 2012, 23:485–490
3. Sibony P, Halperin J, Coyle PK, Patel K. Reactive Lyme serology in optic neuritis. J Neuroophthalmol. 2005;25(2):71-82

**Keywords:** Lyme Disease, Neuroretinitis, Encephalitis, Papilledema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 56

### 50% of Non-Arteritic Anterior Ischemic Optic Neuropathy Occurs between 40-55 Years Old

Ming-Hui Sun<sup>1,2</sup>, Mohammad Ali Shariati<sup>1</sup>, Yaping Joyce Liao<sup>1</sup>

<sup>1</sup>Byers Eye Institute at Stanford University/department of ophthalmology, Palo Alto, CA, USA, <sup>2</sup>Chang Gung Memorial Hospital/department of ophthalmology, Taoyuan, Taiwan

#### Introduction:

Non-arteritic ischemic optic neuropathy (NAION) is the most common acute optic neuropathy in those older than 50.<sup>1-2</sup> and young onset NAION has been reported in 4-23% of patients.<sup>1-4</sup> Aging is a risk factor for NAION, and the normal rate of retinal ganglion cell axonal loss triples after 50.<sup>5</sup> The aim of our study is to investigate the age of onset of NAION and correlation with risk factors.

#### Methods:

We performed a retrospective review at a single institution of patients with NAION (2009-2014) and analyzed patient characteristics, visual field, and optical coherence tomography (OCT).

#### Results:

We studied 57 NAION eyes in 30 patients (32 male, 8 female). NAION led to significant visual field loss (mean deviation AION:  $-14.1 \pm 1.5$  dB; control:  $-1.6 \pm 0.5$  dB;  $P < 0.001$ , Mann-Whitney) and thinning of OCT mean retinal nerve fiber layer (AION  $63 \pm 2$   $\mu$ m; control:  $88 \pm 5$   $\mu$ m,  $P < 0.001$ ) and ganglion cell complex (AION:  $61 \pm 2$   $\mu$ m; control:  $80 \pm 4$   $\mu$ m;  $P < 0.001$ ). Although 50 is commonly considered the cut-off for young onset AION, 50% patients had their first event between age 40-55, with onset at  $\leq 50$  in 11 (33.7%) and  $> 50$  in 19 (66.3%). We examined the NAION risk factors and found that optic disc drusen was associated with earlier onset by 2 decades ( $N=2$ ). Optic disc-at-risk was common in all ages (mean optic disc diameter  $1.60 \pm 0.04$  mm,  $N = 45$  eyes). Analysis of many risk factors revealed that the  $\leq 50$  and  $> 50$  groups were indistinguishable. Obstructive sleep apnea was the most common risk factor ( $\leq 50$ : 82%,  $> 50$ : 79%), and vascular risk factors are common in all age groups. Younger patients did have better prognosis, with final visual acuity better than 20/40 in 75% of those  $\leq 50$  vs. 21% in  $> 50$  group.

#### Conclusions:

Since 50% of NAION patients had onset at 40-55,  $> 50$  should not be considered the common presentation of NAION.

#### References:

1. Hayreh SS. Anterior ischaemic optic neuropathy. I. Terminology and pathogenesis. Br J Ophthalmol 1974;58:955-963.
2. Arnold AC, Costa RM, Dumitrascu OM. The spectrum of optic disc ischemia in patients younger than 50 years. Trans Am Ophthalmol Soc. 2013 Sep;111:93-118.
3. Preechawat P1, Bruce BB, Newman NJ, Biousse V. Anterior ischemic optic neuropathy in patients younger than 50 years. Am J Ophthalmol. 2007 Dec;144(6):953-60.
4. Deramo VA1, Sergott RC, Augsburger JJ, Foroozan R, Savino PJ. Ischemic optic neuropathy as the first manifestation of elevated cholesterol levels in young patients. Ophthalmology. 2003 May;110(5):1041-6.
5. Balazsi AG, Rootman J, Drance SM, et al. The effect of age on the nerve fiber population of the human optic nerve. Am J Ophthalmol 1984;97:760-766.

**Keywords:** NAION, Age, Risk Factors, OCT, Visual Field

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 57

### Evaluation of Retinal Nerve Fiber Layer and Ganglion Cell Complex in a Cohort of Chinese with Optic Neuritis or Neuromyelitis Optica Spectrum Disorders Using SD-OCT

Guohong Tian<sup>1</sup>, Chaoyi Feng<sup>1</sup>, Mengwei Li<sup>1</sup>, Min Wang<sup>1</sup>, Xinghuai Sun<sup>1</sup>, Yan Sha<sup>2</sup>

<sup>1</sup>Department of Ophthalmology, Eye Ear Nose and Throat Hospital of Fudan University, Shanghai, China, <sup>2</sup>Department of Radiology, Eye Ear Nose and Throat Hospital of Fudan University, Shanghai, China

#### Introduction:

To evaluate the retinal nerve fiber layer (RNFL) in eyes of patients with typical multiple sclerosis related optic neuritis (MS-ON), relapsing optic neuritis (R-ON) or neuromyelitis optica spectrum disorders (NMO-SD) using SD-OCT.

#### Methods:

Retrospective and prospective cross sectional study. Patients diagnosed of optic neuritis were recruited between May 2013 and July 2014 in neuro-ophthalmology division. Demographics and clinical features as well as optic disc SD-OCT at least 3 months after the acute attack were analyzed.

#### Results:

Fifty-seven patients with 79 eyes were evaluated. Three groups included MS-ON (32 cases, 39 eyes), R-ON (8 cases, 13 eyes) and NMO-SD (17 cases, 27 eyes). The mean age in three groups was  $32.97 \pm 13.8y$ ,  $44.00 \pm 13.37y$  and  $42.41 \pm 12.64y$ ,  $p=0.026$  between groups. The RNFL in three groups showed  $p=0.019$  between groups. The age was unrelated with the RNFL thickness. The GCC volume in MS-ON group was about  $5\mu m$  and  $10\mu m$  thicker than R-ON and NMO-SD group respectively.

#### Conclusions:

OCT retinal nerve fiber layer and GCC measurement will be helpful in distinguishing NMO spectrum disorders from other forms of MS disease.

**References:** None.

**Keywords:** Optic Neuritis, Optic Neuritis, Optical Coherence Tomography, Retinal Nerve Fiber Layer, Retinal Nerve Fiber Layer

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Initial Grant for introduction of talent plan of Fudan University, Shanghai 上海市

## Poster 58

### Transient Monocular Vision Loss Upon Awakening: a Benign Phenomenon

Marc A. Bouffard<sup>1</sup>, Nurhan Torun<sup>2</sup>

<sup>1</sup>*Beth Israel Deaconess Medical Center, Department of Neurology, Boston, MA, USA*, <sup>2</sup>*Beth Israel Deaconess Medical Center, Division of Ophthalmology, Department of Surgery, Boston, MA, USA*

#### Introduction:

Transient monocular vision loss is often an alarming symptom, as the differential diagnosis includes thromboembolism and giant cell arteritis in the right age group. However, when patients describe waking up with dim or blurred vision in one eye, which resolves quickly, we found workup to be uniformly negative.

#### Methods:

We describe a group of 16 patients who were referred for transient monocular vision loss upon awakening and review patient characteristics and results of their workup.

#### Results:

Medical records of 16 patients (14 women and 2 men), ages 20 to 67, who had been seen in the Neuroophthalmology Clinic for transient monocular vision loss upon awakening were reviewed. All except 3 patients had workup. 11 patients had imaging studies and various combinations of carotid dopplers, echocardiograms and hypercoagulable studies. Workup uniformly failed to explain the patients' symptom.

#### Conclusions:

While transient monocular vision loss is an alarming symptom in most patients, waking up with blurred or dim vision in one eye with subsequent improvement in minutes seems to be an exception. Literature search did not find any description of this phenomenon. The majority of our patients underwent extensive testing for their episodes of visual disturbance. To our knowledge, this is the first report describing a uniformly negative workup in patients presenting with this symptom.

**References:** None.

**Keywords:** Transient Vision Loss

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 59

### Autosomal Dominant Optic Atrophy In Singapore: Multiethnic Involvement And Report Of A New Gene Mutation Causing Optic Atrophy And Deafness

Sharon L Tow<sup>1,2,3</sup>, Jing-Liang Loo<sup>1,2,3</sup>, P Amati-Bonneau<sup>4</sup>, D Bonneau<sup>4</sup>, V Procaccio<sup>4</sup>, P Reynier<sup>4</sup>, Dan Milea<sup>1,2,3,4</sup>

<sup>1</sup>*Singapore National Eye, Centre, Singapore, Singapore*, <sup>2</sup>*Singapore Eye Research Institute, Singapore, Singapore*, <sup>3</sup>*Duke-National University of Singapore, Singapore, Singapore*, <sup>4</sup>*Angers University Hospital Angers, France*

#### Introduction:

Autosomal dominant optic atrophy (ADOA) is a ubiquitous condition causing bilateral visual loss, most commonly related to mutations in the *OPA1* gene, mapped on the chromosome 3q28-q29. Recent data suggests a minimum prevalence of 4.07/100 000 in northern England while ADOA is more rarely reported in Asia. Our study aimed to detect patients with genetically confirmed ADOA in Singapore.

#### Methods:

We conducted a preliminary cross-sectional study at our institution, testing patients (using direct sequencing of the *OPA1* gene) who had a clinical picture compatible with ADOA, seen at our neuro-ophthalmology clinics between January and August 2013.

#### Results:

We tested 6 patients after ruling out other causes of bilateral optic neuropathy including glaucoma, Leber's hereditary optic neuropathy, compressive, nutritional and toxic causes. We found 4 patients with genetically confirmed ADOA on exons 8, and 9 and 17: c.869G>A (p.Arg290Glu), c.871-1G>A, and c.892A>G (p.Ser298Gly) and c.1669C>T (p.Arg557X). Each patient belonged to a different family and 3 different ethnic groups were represented, namely Chinese, Malay and Indian. All patients had bilateral visual loss associated with paracentral scotomas, color vision loss and optic atrophy. A positive family history of visual loss was found in only one of the patients. One proband harboured a novel heterozygous *OPA1* pathogenic variant c.892A>G (p.Ser298Gly) that has not been previously catalogued in the eOPA1 database, causing optic atrophy and deafness in early childhood.

#### Conclusions:

We report the first 4 families with genetically confirmed ADOA in Singapore, belonging to the three main ethnic groups of the country. Among them, we report a new, previously undescribed mutation causing "dominant optic atrophy plus", adding deafness to the classical phenotype of optic atrophy. Further epidemiological studies are needed in order to determine the prevalence of ADOA in Singapore.

**References:** None.

**Keywords:** Genetic Disease, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 60

### **Optic Nerve Head and Macular Choroidal Vascularization in Patients Affected by Normal Tension Glaucoma and Non-Arteritic Ischemic Optic Neuropathy: What do they share? What is different?**

Giacinto Triolo<sup>1</sup>, Enrico Borrelli, Maria Lucia Cascavilla, Federico Di Matteo, Paolo Bettin, Francesco Bandello, Piero Barboni

*University Vita-Salute, San Raffaele Scientific Institute, Milan, Italy*

#### **Introduction:**

To compare peripapillary and macular choroidal vascularization in patients affected by Normal Tension Glaucoma (NTG) and Non-Arteritic Ischemic Optic Neuropathy (N-AION).

#### **Methods:**

Two consecutive series of patients affected by chronic optic neuropathy due to NTG and N-AION, as well as a cohort of healthy subjects, underwent complete ophthalmological examination, Humphrey Visual Field (HVF), Enhanced Depth Imaging Optical Coherence Tomography (EDI OCT). Peripapillary choroidal thickness has been measured on peripapillary circular scans at 3, 6, 9, 12 clocks' hours. Macular choroidal thickness was evaluated on line scans below the subfoveal region (SF), and at 750- and 1000- $\mu$ m intervals, nasally, inferiorly, temporally and superiorly to the fovea. Ganglion Cell Complex (GCC) thickness was also considered in the study. T-test and ANOVA were used to perform statistical analysis.

#### **Results:**

Twelve and 17 patients diagnosed with chronic optic neuropathy due to NTG and monolateral N-AION, respectively, and 14 healthy subjects were enrolled in the analysis. Non statistically significant difference was found in the macular choroidal thickness of N-AION group compared with controls, neither between N-AION affected eyes and the fellow ones. Statistically significant difference was found in the macular choroidal thickness of NTG group in the SF region ( $p<0.01$ ), at the all 750- $\mu$ m intervals ( $p<0.01$ ), and at superior ( $p<0.01$ ), nasal, and inferior ( $p=0.01$ ) 1500- $\mu$ m intervals from the fovea. Non statistically significant difference was found in the peripapillary choroidal thickness among the 2 groups of patients, compared with healthy subjects. Statistically significant GCC thickness' reduction was also found in both NTG and N-AION groups ( $p<0.01$ ), compared with controls.

#### **Conclusions:**

The present study demonstrates no changes in the peripapillary choroidal vascularization in patients affected by NTG and N-AION. Interestingly, macular choroidal vascularization resulted markedly affected in NTG patients, but apparently unmodified in both N-AION affected and fellow eyes.

**References:** None.

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 61

### Multifocal ERG Shows Pre-Ganglion Cells Dysfunction in Dominant Optic Atrophy: Genotype-Phenotype Correlation.

Maria Lucia Cascavilla<sup>1</sup>, Giacinto Triolo<sup>1</sup>, Vincenzo Parisi<sup>2</sup>, Enrico Borrelli<sup>1</sup>, Chiara La Morgia<sup>3,4</sup>, Alfredo Sadun<sup>5</sup>, Francesco Bandello<sup>1</sup>, Valerio Carelli<sup>3,4</sup>, Piero Barboni<sup>1,6</sup>

<sup>1</sup>University Vita-Salute, San Raffaele Scientific Institute, Milan, Italy, <sup>2</sup>G.B. Bietti Foundation, Rome, Italy, <sup>3</sup>IRCCS, Istituto delle Scienze Neurologiche di Bologna, Bologna, Italy, <sup>4</sup>Department of Biomedical and Neuromotor Sciences (DIBINEM), University of Bologna Bologna, Italy, <sup>5</sup>Ophthalmology, UCLA, Doheny Eye Institute Los Angeles, CA, USA, <sup>6</sup>Studio Oculistico d'Azeglio Bologna, Italy

#### Introduction:

To assess the macular function by means multifocal electroretinogram (mfERG) in patients with dominant optic atrophy (DOA) stratified by *OPA1* mutation type.

#### Methods:

A consecutive series of DOA patients, and a group of healthy controls, underwent complete ophthalmological examination and mfERG. Patient's eyes were stratified by *OPA1* mutation type. Two types of analysis have been performed on the mfERG responses: (i) "rings analysis" consisted on analyzing the averaged response obtained from 5 concentric rings centered on the fovea; (ii) "sectors analysis" considered the averaged response obtained from 4 sectors: temporal-superior (TS), temporal-inferior (TI), nasal-superior (NS), and nasal-inferior (NI). For each obtained averaged response we evaluated the response amplitude densities (RAD) between the first negative peak - N1 - and the first positive peak - P1 - (N1-P1 RAD, expressed in nanoVolt/degree<sup>2</sup>).

#### Results:

Eighteen patients from 14 pedigrees with DOA harboring heterozygous mutations in the *OPA1* gene (12 patients with *OPA1* missense mutations (DOA-M) and 6 patients with haploinsufficiency mutations (DOA-H), providing 11 eyes), and 29 age-matched healthy subjects were enrolled in the study. "Rings analysis": DOA-M Group showed Ring 1, Ring 2, Ring 3 and Ring 4 N1-P1 RADs values significantly reduced ( $p<0.01$ ) compared to controls. In DOA-H Group, only the Ring 1 N1-P1 RADs values were significantly reduced ( $p<0.01$ ) compared to controls. "Sectors analysis": DOA-M Group showed a significant reduction ( $p<0.01$ ) of N1-P1 RADs values observed in TI, NS, and NI sectors, compared to controls. In DOA-H Group, the N1-P1 RADs values detected in all sectors (TS, TI, NS, NI) were not significantly different ( $p>0.01$ ) compared to controls.

#### Conclusions:

A clear genotype/phenotype correlation emerged stratifying mfERG responses by *OPA1* mutation type, being missense mutations the most severe. In addition, we focused our attention on the possible detection of the retinal areas that should present the main pre-ganglionic impairment.

**References:** None.

**Keywords:** Optic Neuropathy, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 62

### Pediatric Primary Optic Nerve Sheath Meningioma

Kavin Vanikiet<sup>1</sup>, Pisit Preechawat, Anuchit Poonyathalang

*Department of Ophthalmology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand*

#### Introduction:

Primary optic nerve sheath meningioma (PONSMS) is a proliferation of meningothelial cells within the nerve sheath of the orbital or intracanalicular portion of the optic nerve. It is not common in patients younger than 20 years of age, represent less than 5% of all cases of PONSMS<sup>1</sup>.

#### Methods:

Two cases reports of pediatric PONSMS

#### Results:

Case 1: A 12 year-old-boy gradually lost his right vision over 2 months. Ocular examination showed visual acuity of counting finger, limitation of ocular motility and generalized optic disc edema with opticociliary shunts in his right eye. Systemic evaluation revealed multiple plexiform neurofibromas in both hands and 1 café-au-lait spot on his trunk. MRI of orbit and brain revealed right PONSMS and multiple schwannomas of cranial nerves V, VI, VII, VIII, IX, and XI, and spinal nerve roots. The child was diagnosed with definite neurofibromatosis type 2. He received stereotactic radiotherapy. Case 2: A 10 year-old –boy developed gradually progressive proptosis in his left eye for 2 years. Left ocular examination showed visual acuity of 20/25, limitation of ocular motility and generalized optic disc edema with opticociliary shunts. Systemic evaluation was unremarkable. MRI of orbit and brain revealed PONSMS in the left eye without any abnormalities of his brain. Incisional biopsy of the tumor confirmed the diagnosis. Six months later, his left vision deteriorated to 20/200 and then stereotactic radiotherapy was performed. His vision remained stable over 6 years of follow-up.

#### Conclusions:

Pediatric primary optic nerve sheath meningioma seems to have aggressive clinical behavior with rapid visual decline and poor visual outcome. Stereotactic radiotherapy was shown to be effective in long-term tumor growth control and have no serious complications.

#### References:

1. Harold Lee HB, Garrity JA, Cameron JD, Strianese D, Bonavolonta G, Primary Optic Nerve Sheath Meningioma in Children, *Surv Ophthalmol*, 53, 543-58, 2008

**Keywords:** Optic Neuropathy, Orbit, Pediatric Neuro-Ophthalmology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 63

### Syphilitic Optic Neuropathy : Reemerging Cases Over A 2-Year Period

Anuchit Poonyathalang<sup>1</sup>, Supanut Apinyawasisuk, Pisit Preechawat, Kavin Vanikieti

*Department of Ophthalmology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand*

#### Introduction:

Although the ocular manifestation of syphilis can affect any structures of the eye, optic nerve involvement is not a common presentation. We report a series of 7 new cases of syphilitic optic neuropathy in a tertiary center.

#### Methods:

Retrospective case series

#### Results:

During a 2-year period (2013-2014), 11 eyes of 7 patients were diagnosed with syphilitic optic neuropathy by serologic and cerebrospinal fluid tests. All patients were male, aged between 35 and 45 years. All cases were newly diagnosed with HIV infection, the CD4 T-cell count ranged from 116 to 613 cells/microliter. Three cases (43%) presented with bilateral disc edema, 3 (43%) with unilateral disc edema, and 1 (14%) with bilateral retrobulbar optic neuropathy. Anterior chamber and vitreous reaction were found in 3 patients. Presenting visual acuity ranged from 20/20 to 20/400. Most of patients (6 eyes, 54%) had mild visual loss ( $\geq 20/40$ ), 3 eyes (27%) had moderate visual loss (20/50-20/200), and 2 eyes (11%) had severe visual loss ( $<20/200$ ). Blind spot enlargement was the most common type of visual field defect (7 eyes, 64%). All patients had favorable visual outcome ( $\geq 20/25$ ) after treatment with intravenous penicillin G (6 patients) or ceftriaxone (1 patients).

#### Conclusions:

Syphilitic optic neuropathy has reemerged in our institution in the past two years. Visual loss from optic nerve involvement can be the first manifestation of syphilitic and HIV coinfection. Patients usually have optic disc edema and mild visual dysfunction. Blind spot enlargement syndrome is the common presentation. Syphilitic optic neuropathy has an excellent prognosis if the disease is diagnosed rapidly and treated properly.

**References:** None.

**Keywords:** Neuro-Ophth & Infectious Disease (Eg, AIDS, Prion), Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 64

### Anterior Ischemic Optic Neuropathy as Sole Presenting Sign of Internal Carotid Artery Occlusion

Kimberly M. Winges<sup>1,2</sup>, Rebecca B. Bittner<sup>3</sup>

<sup>1</sup>VA Portland Health System, Department of Ophthalmology, Portland, OR, USA, <sup>2</sup>Casey Eye Institute, Oregon Health and Science University, Portland, OR, USA, <sup>3</sup>VA Southern Oregon Rehabilitation Center and Clinics, Department of Radiology, White City, OR, USA

#### Introduction:

Anterior ischemic optic neuropathy with ophthalmic artery occlusion usually results from giant cell arteritis (GCA), but can rarely occur due to dissection or atherosclerotic occlusion of the internal carotid artery (ICA). Here we present a rare case of severe vision loss in one eye as the sole presenting symptom of ICA occlusion causing cerebral infarction.

#### Methods:

Case Report

#### Results:

A 60 year-old male presented with two days of acute, painless vision loss in the right eye, described as a "blur with kaleidoscope" effect. He denied any headache, weakness, transient vision loss, diplopia, or GCA symptoms. Exam revealed hand motion vision OD and 20/20 OS, with a 2.1 logunit RAPD and pallid optic disc edema OD. Confrontation visual field showed a denser inferior than superior global depression OD and was full OS. Sedimentation rate and c-reactive protein were normal for age. A fluorescein angiogram revealed delayed filling of the nasal and peripapillary choroid and optic disc leakage. Temporal artery biopsy was negative. The patient was lost to followup but was later found to have an inferior temporal field defect on static automated perimetry OS. Subsequent brain MRI revealed an occluded right internal carotid artery and encephalomalacia of the right temporo-parietal-occipital watershed region. CTA head and neck showed 100% occlusion of the right ICA from its origin to the cavernous sinus, with collateral reconstitution of the supraclinoid portion. The patient remained completely asymptomatic apart from vision loss and was followed on anticoagulation by the vascular surgery service.

#### Conclusions:

While neuroimaging is usually reserved for atypical cases, this report highlights the importance of considering intracranial vascular disease in the differential diagnosis of anterior ischemic optic neuropathy and ophthalmic artery occlusion.

#### References:

1. Kleinberg TT, Uretsky S, Flanders AE, Bilyk JR, Murchison AP. Black as night. *Surv Ophthalmol*. 2014 Aug 13.
2. Biousse V, Schaison M, Touboul PJ, D'Anglejan-Chatillon J, Bousser MG. Ischemic optic neuropathy associated with internal carotid artery dissection. *Arch Neurol*. 1998 May;55(5):715-9.
3. Lamirel C, Newman NJ, Biousse V. Vascular neuro-ophthalmology. *Neurol Clin*. 2010 Aug;28(3):701-27.
4. Hayreh SS. Non-arteritic anterior ischemic optic neuropathy versus cerebral ischemic stroke. *Graefes Arch Clin Exp Ophthalmol*. 2012 Sep;250(9):1255-60.

**Keywords:** Ophthalmic Artery Occlusion, Internal Carotid Artery, Stroke, Anterior Ischemic Optic Neuropathy, Infarct

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 65

### Challenges In The Management Of Apoplexy Of A Growth Hormone(GH)-Secreting Pituitary Adenoma During Pregnancy

John Wong<sup>1</sup>, Su Ann Lim

*National Healthcare Group Eye institute, Tan Tock Seng Hospital, Singapore, Singapore*

#### Introduction:

The occurrence of pituitary apoplexy during pregnancy is rare, especially with GH-secreting pituitary adenomas. The management of such a patient is challenging.

#### Methods:

We present a patient who had pituitary apoplexy during early pregnancy.

#### Results:

A 29 year old female presented with symptoms of left sided headache for 1 week. She experienced gradual visual loss in her left eye for 3 years duration. She was 8 weeks pregnant. Her visual acuity was 6/9 in her right eye, and no light perception (NPL) in her left eye with a relative afferent pupillary defect. Her left optic disc was pale. Confrontation visual field testing revealed a temporal hemifield defect of her right eye and complete loss in her left visual field. She had facial features suggestive of acromegaly and spoke in a relatively deep voice. A chiasmal lesion was suspected on account of her visual field and apoplexy considered due to the acute onset of headache. Magnetic resonance imaging (MRI) of her brain was performed and revealed a large pituitary mass, with features suggestive of hemorrhage; causing compression of the optic chiasm and optic nerves. Her serum hormonal profile showed elevated GH and insulin-like growth-factor 1 (IGF-1) levels. She underwent emergent trans-sphenoidal resection of the tumour. Improvement of visual function in both eyes was noted 2 weeks after surgery. The discussion is focused on the literature regarding the effects of MRI with and without contrast, the interpretation of the hormonal profile and the options of treating pituitary apoplexy in pregnancy.

#### Conclusions:

This case illustrates the challenges in the management of a rare occurrence of apoplexy of a GH-secreting pituitary adenoma during pregnancy which was managed surgically, and led to improvement of visual function. All clinicians should be mindful of the usage of MRI during pregnancy.

#### References:

1. Molitch ME. Prolactinoma in pregnancy. Best Pract Res Clin Endocrinol Metab. 2011;25:885- 896.
2. Couture N, Aris-Jilwan N, Serri O. Apoplexy of a microprolactinoma during pregnancy. Case report and review of literature. Endocr Pract. 2012;18:e147-e150
3. Lunardi P, Rizzo A, Missori P, et al. Pituitary apoplexy in an acromegalic woman operated on during pregnancy by transsphenoidal approach. Int J Gynaecol Obstet. 1991;34:71-74.
4. Expert panel on MR safety. ACR guidance document on MR safe practices:2013. Journal of Magnetic Resonance Imaging 2013;37:501-530
5. Webb JA, Thomsen HS, Morcos SK et al. The use of iodinated and gadolinium contrast media during pregnancy and lactation. Eur Radiol. 2005;15:1234–1240
6. Beckers A, Stevenaert A, Foidart JM, Hennen G, Frankenne F. Placental and pituitary growth hormone secretion during pregnancy in acromegalic women. J Clin Endocrinol Metab. 1990;71:725–31.
7. Nawar RN, AbdelMannan D, Selman WR, et al. Pituitary Tumor Apoplexy: A review. J J Intensive Care Med 2008;23:75–90
8. Stalldecker G, Mallea-Gil MS, Guitelman M, et al. Effects of cabergoline on pregnancy and embryo-fetal development: retrospective study on 103 pregnancies and a review of the literature. Pituitary. 2010;13:345-350

**Keywords:** Apoplexy, Pregnancy, MRI, Pituitary

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## **The Prevalence of Mitochondrial Disease in the Adult Population – Implications for the Prevention of Maternal Transmission**

Patrick Yu-Wai-Man<sup>1,2</sup>, Grainne S Gorman<sup>1,3</sup>, Andrew M Schaefer<sup>1,3</sup>, John P Grady<sup>1</sup>, Yi Ng<sup>1,3</sup>, Patrick F Chinnery<sup>1</sup>, Robert W Taylor<sup>1</sup>, Robert McFarland<sup>1,3</sup>, Douglass M Turnbull<sup>1,3</sup>

<sup>1</sup>1. Wellcome Trust Centre for Mitochondrial Research, Newcastle University, Newcastle upon Tyne, United Kingdom, <sup>2</sup>2. Newcastle Eye Centre, Royal Victoria Infirmary, Newcastle upon Tyne, United Kingdom, <sup>3</sup>3. Department of Neurology, Royal Victoria Infirmary, Newcastle upon Tyne, United Kingdom

### **Introduction:**

Establishing the prevalence of mitochondrial disease in the population is challenging due to the extensive clinical and genetic heterogeneity observed in this group of disorders. Innovative IVF techniques to prevent the maternal transmission of pathogenic mitochondrial DNA (mtDNA) mutations have been developed recently and the number of women of childbearing age who could potentially benefit from these breakthroughs in reproductive medicine need to be evaluated.

### **Methods:**

In this national epidemiological study, we comprehensively assessed the prevalence of mitochondrial disease in the adult population secondary to pathogenic mutations involving both the nuclear and mitochondrial genomes. Patients were identified from the MRC Mitochondrial Disease Patient UK Cohort, which is a national database set up to collect extensive clinical and family history information on patients with proven mitochondrial disease throughout the United Kingdom.

### **Results:**

The minimum prevalence figure for pathogenic mtDNA mutations was 20.4 per 100,000 compared with 2.9 per 100,000 for nuclear mutations associated with mitochondrial disease. Leber hereditary optic neuropathy secondary to the m.3460G>A, m.11778G>A and m.14484T>C mutations affected 3.7 per 100,000 of the UK population (~ 1 in 27,000). The most common pathogenic mtDNA mutation was the MELAS m.3243A>G mutation with a minimum carrier rate of 7.9 per 100,000. There are ~ 2,300 women of childbearing age in the UK harbouring pathogenic mtDNA mutations. By using the national fertility rate, this equates to ~ 150 pregnancies per year that could result in the birth of a child at high risk of developing severe mitochondrial disease.

### **Conclusions:**

Mitochondrial disease is common affecting ~ 1 in 4,300 people in the UK. Our findings have major implications for the evaluation of interventions and for the provision of an integrated clinical service. The lack of effective treatments emphasises the importance of developing safe IVF techniques to prevent the maternal transmission of pathogenic mtDNA mutations.

### **References:**

1. Tachibana M, Sparman M, Sritanaudomchai H, et al. Mitochondrial gene replacement in primate offspring and embryonic stem cells. *Nature*. 2009;461:367-72.
2. Craven L, Tuppen H, Greggains GD, et al. Pronuclear transfer in human embryos to prevent transmission of mitochondrial DNA disease. *Nature*. 2010;465:82-5.
3. DiMauro S, Schon EA, Carelli V, Hirano M. The clinical maze of mitochondrial neurology. *Nature reviews Neurology*. 2013;9:429-44.

**Keywords:** Mitochondrial Disease, Genetic Disease, Leber Hereditary Optic Neuropathy, Epidemiology, Treatment

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 67

### Neuropathies In Children Involving Vision Caused By A Fall From Height

Alon Zahavi<sup>1,3</sup>, Judith Luckman<sup>2</sup>, Iftach Yassur<sup>1</sup>, Nitza Goldenberg-Cohen<sup>3,4,5</sup>

<sup>1</sup>Ophthalmology, Rabin Medical Center, Petah Tiqwa, Israel, <sup>2</sup>Radiology, Rabin Medical Center, Petah Tiqwa, Israel, <sup>3</sup>Sackler School of Medicine, Tel Aviv University, Tel Aviv, Israel, <sup>4</sup>Pediatric Ophthalmology Unit, Schneider Children's Medical Center of Israel Petah Tiqwa, Israel, <sup>5</sup>Krieger Eye Research Laboratory, Felsenstein Medical Research Center, Tel Aviv University Petah Tiqwa, Israel

#### Introduction:

Fall from heights can result in severe injuries, and account for the most common pediatric trauma cause for an emergency department visit. We report the incidence and clinical course of neuropathies in children caused by fall from heights.

#### Methods:

Data was obtained from the computerized medical records of all patients admitted to the emergency department of a large tertiary medical center due to fall from height between 2004-2014. Associated optic neuropathies which were assessed during the acute event and the follow-up period were analyzed, including imaging studies and associated clinical findings and outcome.

#### Results:

An estimated 5400 cases of pediatric falls with head trauma presented to the emergency department during the study period. Only 11 children (7 boys, 4 girls, mean age 6.4 years) were diagnosed with neuropathies causing visual disturbance and were followed. Eight were diagnosed with traumatic optic neuropathy, one of which after a 6 months delay and two with accompanying cranial nerve (CN) III injuries. Five of the patients had anisocoria or abnormal pupillary response to light upon initial presentation. The remaining two had CN VI paralysis and temporary visual loss, respectively. Visual improvement varied among all these patients.

#### Conclusions:

Neuropathies due to a fall from heights in children are rare. Neuropathies involving vision or ocular motility impairment are usually missed in the acute intake of emergency patients, especially in cases of monocular loss of vision. Such neuropathies might cause severe morbidity with limited improvement, and prompt interpretation with a high index of suspicion might improve the outcome. Assessment of pupillary response to light is simple and essential in these cases. Due to the potentially devastating visual outcomes of traumatic optic neuropathy, pediatric trauma centers must make an effort to rule out optic neuropathies in all patients visiting the emergency department after a fall from height.

**References:** None.

**Keywords:** Pediatric Neuro-Ophthalmology, Optic Nerve Trauma And Treatment, Orbit/Ocular Pathology, Trauma

**Financial Disclosures:** The Zanyvl and Isabelle Krieger Fund, Baltimore, MD (NGC).

**Grant Support:** The Zanyvl and Isabelle Krieger Fund, Baltimore, MD (NGC).

### Immunosuppressive Therapy of Chinese Isolated Non-MS Idiopathic Optic Neuritis

Xiaojun Zhang<sup>1</sup>, Yunqing Wu<sup>1</sup>, Hanqiu Jiang<sup>1</sup>, Hengri Cong<sup>1</sup>, Lijuan Liu<sup>2</sup>, Jingting Peng<sup>1</sup>, Xiuyun Kong<sup>1</sup>, Rong Yan<sup>1</sup>, Wenbin Wei<sup>3</sup>

<sup>1</sup>Department of Neurology, Beijing Tongren Hospital, Capital Medical University, Beijing, China, <sup>2</sup>Beijing Eye Institute, Beijing, China, <sup>3</sup>Eye Center, Beijing Tongren Hospital, Capital Medical University, Beijing, China

#### Introduction:

Immunosuppressive therapy is recommended for preventing the relapse of neuromyelitis optica(NMO) and NMO-spectrum diseases, but very few studies focused on isolated Non-MS idiopathic optic neuritis(Non-MS-ION).

#### Methods:

Non-MS-ION patients met the inclusive criteria and treated with our standard immunosuppressive therapy were retrospectively studied. Annual relapsing rate(ARR) before and after treatment were compared for all patients, recurrent Non-MS-ION and AQP4-Ab(+) subgroups. Conversion rate to NMO between patients with persistent and non-persistent were also compared.

#### Results:

In 165 cases with averaged age 41.2yrs, 105(63.6%) were female. NMO-like brain lesions was found on 7 cases(4.2%). AQP4-Ab was positive on 30(44.1%) of 68 cases examined. During averaged 29 months of follow-up, the differences of ARR before and after treatment were statistically significant for all 165 cases( $0.46 \pm 0.84$  vs  $0.16 \pm 0.45$ ,  $P=0.000$ ), also in 25 cases with persistent therapy( $0.72 \pm 1.00$  Vs  $0.13 \pm 0.29$ ,  $P=0.005$ ) and 140 cases with non-persistent therapy( $0.41 \pm 0.81$  Vs  $0.17 \pm 0.47$ ,  $P=0.001$ ). For 64 recurrent cases including 14 cases with persistent therapy and 50 cases with non-persistent therapy, the differences were also significant ( $1.18 \pm 0.99$  Vs  $0.22 \pm 0.53$ ,  $P=0.000$ ;  $1.29 \pm 1.02$  Vs  $0.19 \pm 0.33$ ,  $P=0.002$ ;  $1.15 \pm 0.99$  Vs  $0.23 \pm 0.57$ ,  $P=0.000$ ). In 30 cases of AQP4-Ab(+) subgroup, ARR was significantly decreased after treatment for the 11 patients with persistent therapy( $0.67 \pm 0.8$  Vs  $0.08 \pm 0.21$ ,  $P=0.046$ ) but not for the 19 cases with non-persistent therapy ( $0.5 \pm 0.88$  Vs  $0.55 \pm 0.85$ ,  $P=0.80$ ). Fourteen cases(14/165, 8.5%) converted into NMO during follow-up, all of which came from the 140 cases with non-persistent therapy. In AQP4-Ab(+) Non-MS-ION subgroup, none of 11 cases with persistent therapy but 9 of 19(47.3%) patients with non-persistent therapy developed NMO and the difference was statistically significant( $0\%$  Vs  $47.3\%$ ,  $P=0.01$ ).

#### Conclusions:

Immunosuppressive therapy decreased ARR of Chinese Non-MS ION. Persistent immunosuppressive therapy is needed to reduce the ARR and conversion rate to NMO for AQP4-Ab positive patients.

#### References:

1. Kowarik MC, Soltys John, Bennett JL. The treatment of neuromyelitis Optica. J Neuro-ophthalmology, 2014, 34: 70-82.

**Keywords:** Optic Neuritis, Neuromyelitis Optica, Preventative, Immunosuppressive, Treatment

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** 1. Beijing Educational Committee Research Funding, 2011 2. Chinese National Science-Tech Supportanting Plan, Clinical study of Optic Neuritis. Grant No. 2012BAI08B06

## Poster 69

### How does Exogenous ROS Effect LHON Conversion?

Jeffrey Tran<sup>1</sup>, Youning Zhang<sup>1</sup>, Rustum Karanjia<sup>2</sup>, Chiara La Morgia<sup>3,4</sup>, Solange R. Salomao<sup>5</sup>, Adriana Berezovsky<sup>5</sup>, Filipe Chicani<sup>5</sup>, Milton Moraes<sup>5</sup>, Milton M Filho<sup>5</sup>, Rubens Belford Jr.<sup>5</sup>, Alfredo A. Sadun<sup>2</sup>, Valerio Carelli<sup>3,4</sup>

<sup>1</sup>Keck School of Medicine of University of Southern California, Los Angeles, CA, USA, <sup>2</sup>Doheny Eye Institute, UCLA, Los Angeles, CA, USA, <sup>3</sup>IRCCS Istituto delle Scienze Neurologiche di Bologna, Bellaria Hospital, Bologna, Italy, <sup>4</sup>Neurology unit, Department of Biomedical and NeuroMotor Sciences (DIBINEM), University of Bologna Bologna, Italy, <sup>5</sup>Department of Ophthalmology, Federal University of Sao Paulo (UNIFESP) Sao Paulo, Brazil

#### Introduction:

Mitochondrial DNA mutations associated with Leber's hereditary optic neuropathy (LHON) are associated with endogenous reactive oxygen species (ROS) production. Research has demonstrated that exogenous ROS generators, like smoking, are associated with an earlier age of conversion from carrier to affected state. However, the exact role of exogenous ROS in LHON is ill defined. To understand the role of exogenous ROS in LHON, we hypothesize dividing LHON into two subtypes. I: conversion is predominantly genetically determined and largely independent of exogenous ROS and II: the genetic component is necessary but insufficient for conversion and exogenous ROS exposure is required.

#### Methods:

To explore this theory we conducted a retrospective case-control study of 19 LHON-affected patients from a single pedigree, looking at their ages of conversion and smokers versus non-smokers. Because type II individuals require higher ROS exposure, we expect the mean of their ages of conversion to be higher and the distribution to be wider. We thus separated our study population at the 75<sup>th</sup> percentile of age of conversion and looked at the smoker to non-smoker ratio in each population using a Pearson's X<sup>2</sup>-test.

#### Results:

The 75<sup>th</sup> percentile for age was 36 years and 32% patients studied were found to have an age of conversion equal to or greater than the 75<sup>th</sup> percentile. Of these patients, 83% were found to have smoked in contrast to 46% in the population with ages of conversion less than 36. Comparison of smoker to non-smoker ratio via Pearson's X<sup>2</sup>-test revealed  $X^2(1, N=19)=1.53$ ,  $p=0.22$ .

#### Conclusions:

While our analysis did not yield a significant result ( $p>0.05$ ), the sample size was small and rough approximations were made to determine type I versus type II patients. However, the descriptive statistics are in line with our hypothesis suggesting there may be two different manifestations of LHON even in a single pedigree.

**References:** None.

**Keywords:** Genetic Disease, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 70

### Chronic, Painful Abducens Palsy May Require Angiography: Lesson Re-Learned From A Case Of A White-Eyed Shunt

Aliaa H. Abdelhakim<sup>1</sup>, Jeffrey G. Odel<sup>2</sup>, Philip M. Meyers<sup>3</sup>, Angela Lignelli<sup>4</sup>, Alexander G. Khandji<sup>4</sup>

<sup>1</sup>Columbia University College of Physicians and Surgeons, New York, NY, USA, <sup>2</sup>Columbia University, Edward S. Harkness Eye Institute, Department of Ophthalmology, New York, NY, USA, <sup>3</sup>Columbia University Medical Center, Department of Neurological Surgery, New York, NY, USA, <sup>4</sup>Columbia University Medical Center, Department of Radiology New York, NY, USA

#### Introduction:

Posteriorly-draining cavernous-carotid fistulas (CCFs) ("white-eyed shunts") can present with painful cranial nerve palsies. We report a case of posteriorly-draining CCF presenting as a persistently painful, chronic left abducens palsy.

#### Methods:

Case Report.

#### Results:

A 58 year-old woman presented with left periorbital pain and diplopia, both worsened on left gaze and preceded by several months of headache. Vision, fields and pupils were normal. Her eyes were white and without proptosis. Tensions were 19/20 mmHg. She had 2-3 mm of left abduction. Cranial nerves V, VII and VIII were intact, and tear production was normal. Gonioscopy was wide open, with no blood in Schlemm's canals. No bruits were heard, and fundi were normal. Contrast-enhanced MRI of the brain and orbits were normal. On review, we suspected a slightly large left superior ophthalmic vein of questionable significance. CT angiogram revealed congestion of the vessels in the cavernous sinus on the left with early contrast opacification of the left cavernous sinus in comparison to the right, consistent with a posteriorly-draining CCF. This was confirmed by diagnostic angiogram, which showed a Barrow type D fistula with retrograde intracortical venous drainage. She was treated with endovascular coil occlusion of the fistula. At her three-month follow-up, her pain had significantly improved, with minimal improvement in her abducens palsy.

#### Conclusions:

Posteriorly-draining CCFs should be considered in patients presenting with chronic, painful ocular muscle palsy (Hawke et al., 1989; Acierno et al., 1995). If MRI of cavernous sinus and skull base is negative, the clinician should proceed with CT angiography or MRI angiography with contrast; if these are unrevealing, conventional catheter-based angiography should be considered.

#### References:

1. Acierno, MD et al. Arch Ophthalmol., 1995; 113(8):1045-9.
2. Hawke, SH et al. Arch Neurol., 1989; 46(11):1252-5.

**Keywords:** Vascular Disorders, Neuroimaging, Interventional Neuroradiology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 71

### Eight And A Half Syndrome-A Rare Neuro Ophthalmologic Manifestation Due To Pontine Infarction

Sanam Anwer<sup>1</sup>, Apoorv Prasad<sup>2</sup>

<sup>1</sup>SUNY upstate university, Syracuse, NY, USA, <sup>2</sup>SUNY upstate university, Syracuse, NY, USA

#### Introduction:

Gaze hemiparesis with INO and facial nerve palsy, the aforementioned triad commonly known as the “Eight and a half syndrome” is a very rare presentation of the insults to pontine tegmentum abducent nucleus, PPRF and the ipsilateral MLF. In this case report we describe eight and a half syndrome in setting of basilar artery aneurysm.

#### Methods:

62 yr old gentleman with history of uncontrolled hypertension, dyslipidemia and diabetes type II presented with sudden onset of right hemiparalysis, left facial droop with decreased closure of the left palpebral fissure, scanning speech and double vision. On presentation his NIHSS was 15. He had left INO, right horizontal gaze paresis and LMN type left facial nerve palsy. Accompanying these symptoms he also had complete paralysis of the right upper and lower extremities, strength 0/5. DWI showed acute ischemic infarction of the left midbrain, pons and cerebellum in the distribution of the superior and inferior cerebellar arteries. Four vessel angiogram further revealed fusiform dilation of the basilar artery as a result of a dissecting aneurysm with no impending stenosis. Extreme tortuosity of the left cervical vertebral artery at the origin was also seen which made intervention almost impossible. The patient was discharged to sub acute rehab.

#### Results:

#### Conclusions:

One and a half syndrome, first described by Fisher in 1957, is a clinical disorder characterized by conjugate horizontal palsy in one direction (the “one”) and INO in the other direction (the “one half”). [1,2] When facial nerve is also involved the manifestation is named as “eight and a half syndrome” first described by Eggenberger in 1998 [3]. Here we present a rare manifestation of eight and a half syndrome secondary to basilar artery aneurysm.

#### References:

1. Fisher CM, “Some neuro-ophthalmological observations.” J Neurol Neurosurg Psychiatry. 1967;30(5):383-392
2. Wall M, Wray SH, “The one-and-a-half syndrome—a unilateral disorder of the pontine tegmentum: a study of 20 cases and review of the literature.” Neurology. 1983;33(8):971-980
3. Eggenberger E, “Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy.” J Neuroophthalmol. 1998;18(2):114-116

**Keywords:** Pprf, Ino, Eight And A Half Syndrome, Abducent Nerve, Basilar Artery

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 72

### Where the Lung Meets the Eye

Nathan W Blessing<sup>1</sup>, Audrey C Ko, Angela M Herro

*Bascom Palmer Eye Institute, Miami, FL, USA*

#### Introduction:

The cavernous sinus is an important neurovascular crossing point. Careful examination of patients presenting with multiple cranial neuropathies may suggest a lesion of this region but the differential is wide and often difficult to confirm.<sup>1</sup> Potential infectious and inflammatory etiologies as well as local malignant invasion should be addressed. However, metastases from distant sites are more rarely seen.<sup>2</sup>

#### Methods:

#### Results:

A 63-year-old man presented with unilateral ptosis and denied significant past medical or ocular history. A review of systems revealed headache, periorbital pain, cough, and decreased appetite. Examination showed complete left ptosis and ophthalmoplegia, symmetric 20/50 vision, diminished V<sub>1</sub>/V<sub>2</sub> sensation, oral thrush, and cachexia. He was additionally febrile, hypotensive, tachycardic, confused, and ataxic. Chart review unveiled a history of uncontrolled AIDS with poor compliance. He was urgently admitted with concern for septic cavernous sinus syndrome, pan-cultured, and started on broad-spectrum antibiotics pending neuroimaging. Initial CT was negative but routine chest x-ray revealed a left hilar mass that biopsy confirmed as small cell lung cancer with post-obstructive pneumonia. MRI ultimately disclosed a lesion of the sella with extension into the left cavernous sinus consistent with metastasis for which he received palliative radiation.

#### Conclusions:

A cavernous sinus syndrome is suggested when two or more of cranial nerves 3, 4, 5(V<sub>1</sub>, V<sub>2</sub>) or 6 are impaired. The underlying etiologies are diverse and many are not only sight but life-threatening.<sup>1,2</sup> Imaging should be obtained rapidly and serial imaging considered when appropriate clinical suspicion exists without an initial diagnostic lesion.<sup>3</sup> Immunocompromised patients represent a particular diagnostic dilemma given their increased risk of opportunistic infections and potential lack of a robust inflammatory response. This case was diagnostically challenging because the patient presented alone with confusion, an undiagnosed metastatic malignancy not seen on preliminary imaging, and a distant underlying infection; appropriate clinical assessment allowed proper intervention and treatment.

#### References:

1. Fernández S, et al. Cavernous sinus syndrome: a series of 126 patients. *Medicine (Baltimore)*. 86(5):278-81, 2007.
2. Keane JR. Cavernous sinus syndrome. Analysis of 151 cases. *Arch Neurol*. 53(10):967-71, 1996.
3. Ahn Y et al. Cavernous sinus metastasis of non-small cell lung cancer. *Tuberculosis and Respiratory Diseases*. 69: 381–84, 2010.

**Keywords:** Cavernous Sinus, Metastasis, Immunocompromise, Cranial Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 73

### Metastasis or Metamorphosis?

Krista Kinard<sup>5</sup>, Cecinio Ronquillo<sup>1</sup>, Kajsa Affloter<sup>2</sup>, Cheryl Palmer<sup>2</sup>, Edward Quigley<sup>3</sup>, William Couldwell<sup>4</sup>, Kathleen Digre<sup>5,6</sup>, Judith Warner<sup>5,6</sup>, Bradley Katz<sup>5,6</sup>, Alison Crum<sup>5</sup>

<sup>1</sup>University of Utah, School of Medicine, Salt Lake City, UT, USA, <sup>2</sup>University of Utah, Department of Pathology, Salt Lake City, UT, USA, <sup>3</sup>University of Utah, Department of Radiology, Salt Lake City, UT, USA, <sup>4</sup>University of Utah, Department of Neurosurgery Salt Lake City, UT, USA, <sup>5</sup>University of Utah, Department of Ophthalmology Salt Lake City, UT, USA, <sup>6</sup>University of Utah, Department of Neurology Salt Lake City, UT, USA

#### Introduction:

A 36-year-old woman presented in December 2013 with double vision. Three weeks prior, she had a new headache that was relieved with an occipital nerve block. Three days before, she had acute onset of horizontal binocular diplopia that was worse with left gaze. She denied dizziness, nausea, vomiting or weight loss. Her neuro-ophthalmic exam was remarkable for a left eye abduction deficit. MRI of the brain and orbits, from an outside facility, was read as negative for an intracranial mass/lesion. In April 2014, she returned with continued diplopia and new complaints of severe headache, weight loss, night sweats, vertigo and difficulty with balance. Exam showed a left 6<sup>th</sup> nerve palsy, absent corneal reflexes, and bilateral facial paralysis. A lumbar puncture showed mild leukocytosis, elevation of protein, and normal flow cytometry and cytology. All lab tests were within normal limits. Brain MRI showed enhancement in the left cerebellopontine angle, multiple cranial nerves, skull base dura and leptomeninges. A 5-day course of IV steroids was undertaken without improvement of symptoms. Brain biopsy showed an ill-defined soft mass. Pathology showed a benign epidermoid cyst. A second tissue biopsy was done because of high suspicion for malignancy, which showed a poorly differentiated infiltrating squamous cell carcinoma. There was no evidence for a primary tumor by whole body PET.

#### Results:

We have two hypotheses for the origin of the SCC: metastasis from a primary tumor or malignant transformation of the epidermoid cyst. Although rare, malignant transformation of an epidermoid cyst has been reported. The close proximity of the lesions and lack of other primaries on imaging make transformation possible; however, there were no histologic sections that show direct transition of the malignancy from the epidermoid cyst.

#### Conclusions:

Squamous cell carcinoma of unknown origin.

#### References:

1. Agarwal S, Rishi A, Suri V, Sharma MC, Satyarathi GD, Garg A, Sarkar C. Primary intracranial squamous cell carcinoma arising in an epidermoid cyst--a case report and review of literature. Clin Neurol Neurosurg. Dec;109(10):888-91. 2007
2. Lakhdar F, Hakkou el M, Gana R, Maaqili RM, Bellakhdar F. Malignant transformation six months after removal of intracranial epidermoid cyst: a case report. Case Rep Neurol Med. 2011
3. Nakao Y, Nonaka S, Yamamoto T, Oyama K, Esaki T, Tange Y, Mori K, Wada R. Malignant transformation 20 years after partial removal of intracranial epidermoid cyst--case report. Neurol Med Chir (Tokyo). 50(3):236-9. 2010

**Keywords:** Cranial Nerve, Neuroimaging, Neoplasm

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Research to Prevent Blindness, University of Utah

## Poster 74

### Acute Onset Internuclear Ophthalmoplegia And Migraine Headache

Oana M. Dumitrascu<sup>1</sup>, Nili Steiner, Patrick Lyden

*Cedars-Sinai Medical Center, Department of Neurology, Los Angeles, CA, USA*

#### **Introduction:**

Usually a diagnosis of exclusion in adults, ophthalmoplegic migraine is characterized by recurrent attacks of migraine headaches associated with paresis of one or more ocular cranial nerves, in the absence of any demonstrable intracranial lesion other than MRI changes within the affected nerve. Paresis of the cranial nerves third, fourth and sixth were described. Medial longitudinal fasciculus involvement and subsequent internuclear ophthalmoplegia (INO) in association with migraine headache, as well as the INO resolution with migraine abortive therapy is not reported in the literature.

#### **Methods:**

Case Report: 67 year-old right handed Caucasian female presented with acute onset horizontal binocular diplopia and blurry vision during routine household chores. Right-sided throbbing headache and mild nausea accompanied shortly. Patient reported history of HTN, HLD, ischemic heart disease and migraine headaches - one episode 40 years prior associated with limb paresis, while more recently frequent episodes associated with diplopia and/or blurry vision, relieved with rest and nonsteroidal anti-inflammatories. Examination revealed a right INO. Non-contrast head CT and CT angiogram of head and neck were unremarkable. Given prior history and clinical presentation, decision was made to treat with acute migraine therapy, consisting of intravenous hydration with normal saline and IV valproate sodium. Two hours after the therapy initiation patient reported complete resolution of the headache and nausea and significant improvement of the visual symptoms. Examination revealed improvement in the right eye adduction. 10 hours later, after repeat administration of intravenous valproate, the INO was completely resolved and patient returned to baseline functionality. Brain MRI with thin brainstem cuts was unremarkable.

#### **Conclusions:**

Attacks of migraine headaches and acute INO in a patient with history of complicated migraines could be a subtype of ophthalmoplegic migraines. Although an ischemic event should be considered in the differential, acute therapy should target the migrainous phenomenon.

**References:** None.

**Keywords:** Ocular Motility, Nystagmus, Migraine, Vascular Disorders

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Traumatic Avulsion Of The Oculomotor Nerve: First Definitive Documentation On High Resolution MRI

Lauren C Ditta<sup>1,2,3</sup>, Asim F Choudhri<sup>1,2,3,4,5</sup>, Ari Blitz<sup>6</sup>, James C Fleming<sup>1,2,3</sup>, Natalie C Kerr<sup>1,2,3</sup>, Thomas J O'Donnell<sup>1,3</sup>

<sup>1</sup>University of Tennessee, Hamilton Eye Institute, Memphis, TN, USA, <sup>2</sup>Le Bonheur Children's Hospital, Memphis, TN, USA, <sup>3</sup>St. Jude Children's Research Hospital, Memphis, TN, USA, <sup>4</sup>University of Tennessee, Department of Radiology Memphis, TN, USA, <sup>5</sup>University of Tennessee, Department of Neurosurgery Memphis, TN, USA, <sup>6</sup>John's Hopkins University, Department of Radiology Baltimore, MD, USA

### Introduction:

Direct traumatic oculomotor nerve injury is uncommon. In the setting of severe head trauma, third nerve root avulsion from the brainstem can result from differential mass movements and the relationship between the brainstem, supratentorial structures, and the skull bones. Autopsy studies have described traumatic avulsion of the oculomotor nerve, however detailed characterization of mechanisms of injury and precise locations of nerve damage has yet to be definitively described in vivo. We report a case where high resolution MRI imaging confirmed irreversible injury to the left third cranial nerve allowing for more informed management decisions.

### Methods:

Single case report. Retrospective review of a Health Insurance Portability and Accountability Act-compliant database of a pediatric patient sustaining motor-vehicle trauma.

### Results:

A 13 year-old female found unresponsive following a motor vehicle accident was noted to have a fixed and dilated pupil of 8mm on the left. On prism and alternate cover testing there was a 50-prism diopter left exotropia and a 25-prism diopter left hypotropia. She was unable to elevate, depress, or adduct her left eye. MRI of the brain demonstrated avulsion of the left oculomotor nerve from the ventral midbrain at the apparent origin of the cisternal segment.

### Conclusions:

Oculomotor nerve root avulsion from the brainstem may occur in the setting of severe head trauma. High resolution MRI is the best method to detect injury to the cranial nerves. Neurosurgical techniques for possible nerve re-implantation at the brainstem have yet to be investigated, but could be a new area for clinical and surgical research guided by detailed imaging sequences.

### References:

1. Heinze J. Cranial nerve avulsion and other neural injuries in road accidents. *Med J Aust.* 1969 Dec 20;2(25):1246–9.
2. Balcer LJ, Galetta SL, Bagley LJ, Pakola SJ. Localization of traumatic oculomotor nerve palsy to the midbrain exit site by magnetic resonance imaging. *AJOPHT.* 1996 Sep;122(3):437–9.
3. Blitz AM, Choudhri AF, Chonka ZD, Ilica AT, Macedo LL, Chhabra A, Gallia GL, Aygun N. Anatomic Considerations, Nomenclature, and Advanced Cross-sectional Imaging Techniques for Visualization of the Cranial Nerve Segments by MR Imaging. *Neuroimaging Clinics of North America.* 2014 Feb;24(1):1–15.
4. Blitz AM, Macedo LL, Chonka ZD, Ilica AT, Choudhri AF, Gallia GL, Aygun, N. High-resolution CISS MR imaging with and without contrast for evaluation of the upper cranial nerves: segmental anatomy and selected pathologic conditions of the cisternal through extraforaminal segments. *Neuroimaging Clinics of North America.* 2014 Feb;24(1):17–34.

**Keywords:** Oculomotor Nerve, Pediatric, Trauma, Cranial Nerve, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This study was supported in part by an unrestricted grant to the Department of Ophthalmology, University of Tennessee Health Science Center, Memphis, from Research to Prevent Blindness, Inc., in New York, New York.

## Poster 76

### Ocular Motor Cranial Nerve Palsies In Pituitary Apoplexy

Rabih Hage<sup>1</sup>, Beau B Bruce<sup>1</sup>, Sheila Eshraghi<sup>2</sup>, Nelson M Oyesiku<sup>2</sup>, Adriana G Ioachimescu<sup>2,4</sup>, Nancy J Newman<sup>1,2,3</sup>, Valerie Biousse<sup>1,3</sup>

<sup>1</sup>Department of Ophthalmology, Emory University, Atlanta, GA, USA, <sup>2</sup>Department of Neurological Surgery, Emory University, Atlanta, GA, USA, <sup>3</sup>Department of Neurology, Emory University, Atlanta, GA, USA, <sup>4</sup>Department of Medicine, Emory University Atlanta, GA, USA

#### Introduction:

Pituitary apoplexy (PA) is caused by sudden pituitary hemorrhage/necrosis and often presents with acute neuro-ophthalmic manifestations, including ocular motor cranial nerve palsies (CNP). We describe the epidemiology and outcomes of CNP in a large single-center cohort of PA patients.

#### Methods:

Retrospective review of 235 patients with PA seen in our Pituitary Center [01/1995-12/2012]. Presenting neuro-ophthalmic, endocrine and radiologic data were collected; neuro-ophthalmology followup information was obtained when available.

#### Results:

59/235(25%) PA patients had CNP. 56/59(95%) had surgery. 27/59(50%) had neuro-ophthalmic evaluation. 26/27(96%) neuro-ophthalmic-evaluated CNP patients had headaches and 3/27(11%) altered mental status. Pre-operatively, 23/27 patients had unilateral CNP. 18/23 had single (CNVI [N=10], CNIII [N=8]), and 5/23 multiple CNPs (all involving CNIII plus CNIV [N=1], CNVI [N=1] or both [N=3]). 4/27 patients had bilateral CNPs. Median VA was 0.17 [IQR: 0-0.7; VA<20/200 in 9 eyes (6 patients)]. 24/40(62%) eyes with visual field (VF) testing had at least partial temporal hemianopia (complete in 10/40[25%]), 14 patients had chiasmopathy (including one junctional syndrome) and 3 had unilateral optic neuropathy. Post-operatively, 24/27 CNP patients had followup (median duration of followup: 7 months [IQR: 3-17]). At last post-operative followup, 7/24(29%) patients had CNP (5/7 unilateral (CNVI [N=2]; CNIII [N=1]; CNIII and CNIV [N=1]; CNIII, CNIV, and CNVI [N=1]) and 2/7 bilateral). Median VA was 0.1 [IQR:0-0.2] (VA <20/200 in 2 eyes). 15/47(32%) eyes with VF testing had at least partial temporal hemianopia (complete in 3/47(6%) eyes). 7 patients had a chiasmopathy and 2 had a unilateral optic neuropathy. CNP resolved in 3/24(12%) patients within 1 month, 13/21(62%) patients within 6 months (3 lost to followup) and 17/19(89%) within one year (2 lost to followup).

#### Conclusions:

25% of PA patients had CNPs, associated with headaches in 96%, and visual loss in 52% of patients evaluated. 90% of patients had resolution of CNP by one year, most within 6 months.

#### References:

1. Semple PL, Webb MK, De Villiers JC, Laws ER. Pituitary apoplexy. *Neurosurgery* 2005;56(1):65-72.
2. Kim SH, Lee KC, Kim SH. Cranial nerve palsies accompanying pituitary tumour *J Clin Neurosci* 2007;14(12):1158-1162.
3. Murad-Kejbou S, Eggenberger E. Pituitary apoplexy: evaluation, management, and prognosis. *Curr Opin Ophthalmol* 2009;20(6):456-461.
4. Lau KKW, Joshi SM, Ellamushi H, Afshar F. Isolated bilateral oculomotor nerve palsy in pituitary apoplexy: case report and review. *Br J Neurosurg* 2007;21(4):399-402.

**Keywords:** Pituitary Apoplexy, Ocular Motility, Chiasmopathy, Tumors, Pituitary Tumors

**Financial Disclosures:** This work was supported in part by an unrestricted departmental grant (Department of ophthalmology) from Research to Prevent Blindness, Inc., New York. Dr. Hage received research grants from the Philippe Foundation (NYC, USA), the Club de Neuro-Ophthalmologie Francophone (Paris, France) and the Faculté de Médecine des Antilles et de la Guyane (bourse année-recherche). Dr. Bruce receives research support from the NIH/NEI (K23-EY019341)

**Grant Support:** This work was supported in part by an unrestricted departmental grant (Department of ophthalmology) from Research to Prevent Blindness, Inc., New York. Dr. Hage received research grants from the Philippe Foundation (NYC, USA), the Club de Neuro-Ophthalmologie Francophone (Paris, France) and the Faculté de Médecine des Antilles et de la Guyane (bourse année-recherche). Dr. Bruce receives research support from the NIH/NEI (K23-EY019341)

## Poster 77

### Later Life Decompensation of Congenital Trochlear Palsy due to Agenesis

Ji-Soo Kim<sup>2</sup>, SeungHa Lee<sup>1</sup>, Sung-Hee Kim<sup>2</sup>, Hee K Yang<sup>3</sup>, Jeong-Min Hwang<sup>3</sup>, Jae H Kim<sup>4</sup>

<sup>1</sup>Gangnam Severance Hospital/Neurology, Seoul, Korea, <sup>2</sup>Neurology/Seoul National University Bundang Hospital, Seongnam, Korea, <sup>3</sup>Ophthalmology/Seoul National University Bundang Hospital, Seongnam, Korea, <sup>4</sup>Radiology/Seoul National University Bundang Hospital Seongnam, Korea

#### Introduction:

Congenital trochlear palsy may manifest with sudden vertical diplopia due to decompensation during the later life, which may bring a diagnostic challenge. In those cases, imaging demonstration of atrophic superior oblique (SO) muscle and absent trochlear nerve may aid in determining a congenital origin.

#### Results:

Two patients with vertical diplopia for several years after age 50 were referred due to no improvement or sudden aggravation of the diplopia. Findings were consistent with unilateral SO palsy in both patients with a contraversive head tilt. Facial asymmetry was suggestive of a congenital cause in a patient. High resolution MRIs disclosed atrophic SO and absent trochlear nerve in the side of SO palsy in both patients.

#### Conclusions:

Imaging demonstration of SO atrophy and absent trochlear nerve may help diagnosing congenital trochlear palsy due to agenesis that manifests with sudden vertical diplopia during the later life due to delayed decompensation.

**References:** None.

**Keywords:** Diplopia, Trochlear Nerve, Agenesis

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 78

### **Presentation of Primary Hodgkin's Lymphoma as Multiple Cranial Neuropathies**

Hreem N Patel<sup>1</sup>, Aimee J Szewka<sup>1,2</sup>

<sup>1</sup>Rush University Medical Center, Department of Ophthalmology, Chicago, IL, USA, <sup>2</sup>Rush University Medical Center, Department of Neurology, Chicago, IL, USA

#### **Introduction:**

There is a large differential diagnosis when presented with a patient multiple cranial neuropathies (MCN), including neoplastic, infectious and inflammatory causes. We report a case of probable primary Hodgkin's lymphoma (HL) presenting with multiple cranial neuropathies.

#### **Methods:**

Retrospective review of one patient with MCN found to have a mass lesion in the region of Meckel's cave on neuroimaging. History, physical examination, magnetic resonance imaging (MRI), pathology reports and treatment course were reviewed.

#### **Results:**

61 year old gentleman with history of rheumatoid arthritis on immunosuppression presented with worsening left neck pain and horizontal diplopia for four weeks. Afferent examination, including optic nerve and dilated funduscopic examination, was unremarkable. Motility examination showed 50% restriction of left abduction and decreased sensation in first and second division of the trigeminal nerve. MRI was performed and demonstrated enhancing soft tissue within the left Meckel's cave and the left foramen rotundum as well as thickening and enhancement along the falx cerebri and tentorium cerebelli. CSF analysis revealed lymphocytic pleocytosis and elevated protein with nonspecific cytology. Computed tomography of the chest, abdomen, and pelvis showed diffuse lymphadenopathy. Lymph node biopsy confirmed classic HL. Biopsy of the skull base lesion performed after intravenous steroid therapy was inconclusive for lymphoma. Patient improved with chemotherapy and his MCN improved significantly over the next three months. Neurologic symptoms from HL are a rare occurrence. A review of the literature from 1983 found that only 0.5% of patients with HL developed neurological symptoms or signs (1). Presentation of HL with multiple cranial neuropathies has not been reported, but there was a case of cavernous sinus involvement reported in a patient with known HL in 1996 (2).

#### **Conclusions:**

The diagnosis of Hodgkin's lymphoma should be considered in patients presenting with multiple cranial neuropathies as well as for lesions affecting Meckel's cave.

#### **References:**

1. Sapozink MD, Kaplan HS. Intracranial Hodgkin's disease: A report of 12 cases and review of the literature. *Cancer*. 52. 1301-7. 1983.
2. Kasner SE, Galetta SL, Vaughn DJ. Cavernous sinus syndrome in Hodgkin's disease. *J Neuroophthalmol*. 16. 204-207. 1996.

**Keywords:** Ocular Motility, Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 79

### Acute Angle Closure Glaucoma In A Patient With Miller Fisher Syndrome Without Pupillary Dysfunction

Won Yeol Ryu<sup>1</sup>, Jong Kuk Kim<sup>2</sup>

<sup>1</sup>Department of Ophthalmology, Dong-A University College of Medicine, Busan, South Korea, <sup>2</sup>Department of Neurology, Dong-A University College of Medicine, Busan, South Korea

#### Introduction:

To report a case of an acute angle closure glaucoma (AACG) in a patient with Miller Fisher syndrome (MFS) without pupillary dysfunction.

#### Methods:

We present a case report of a 75-year-old male presenting with total ophthalmoplgia, complete bilateral ptosis, and gait disturbance. Two weeks prior to admission, he experienced an upper respiratory infection. An initial neurologic examination demonstrated bilateral complete ptosis, total ophthalmoplegia, and severe gait ataxia. Deep tendon reflexes completely disappeared. The pupillary reaction of the right eye was nonresponsive to light and mid-dilated. Cerebrospinal fluid analysis was acellular and indicated slightly increased protein (53 mg/dl). A diagnosis of MFS was made, and intravenous immunoglobulins were administered.

#### Results:

One day after his admission, the patient complained of ocular pain and blurred vision in the right eye. His visual acuity was hand motion in the right eye and 6/20 in the left eye. In addition, lid swelling, conjunctival chemosis, corneal edema, and a shallow anterior chamber in the right eye were noted. A mid-dilated pupil (4.5 mm) was detected in the right eye. However, the left eye exhibited good reactivity to light. Intraocular pressure, as assessed by Goldmann applanation tonometry, was 50 mmHg, and gonioscopic findings revealed a closed angle on the right eye. After maximal tolerated medical therapy, laser peripheral iridotomy was performed. Subsequent blood tests revealed that the patient was negative for the anti-GQ1b antibody. The unilateral AACG with MFS resolved without further incident.

#### Conclusions:

This is the first reported case of a patient with MFS without autonomic dysfunction with AACG. We believe that pupillary dysfunction or lid ptosis due to neurological disorders may increase the possibility of AACG.

**References:** None.

**Keywords:** Miller-Fisher Syndrome, Guillain-Barré Syndrome, Acute Angle Closure Glaucoma, Pupil, Immune Mediated Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## **Congenital Oculomotor Nerve Paresis With Isolated Cyclic Pupillary Spasms**

Michael S. Salman<sup>1</sup>, Ian H. Clark<sup>3</sup>, Samantha F. Klassen<sup>2</sup>

<sup>1</sup>University of Manitoba/Department of Pediatrics and Child Health, Winnipeg, MB, Canada, <sup>2</sup>University of Manitoba/ Faculty of Medicine, Winnipeg, MB, Canada, <sup>3</sup>University of Manitoba/Department of Ophthalmology, Winnipeg, MB, Canada

### **Introduction:**

Cyclic oculomotor nerve paresis is a rare and mostly congenital disorder. It is characterized by unilateral oculomotor nerve paresis with periodic spasms causing eyelid elevation, miosis and contraction of one or more of the extraocular muscles innervated by the oculomotor nerve. We discuss a 20-month old girl who presented initially with right partial congenital oculomotor nerve paresis. She subsequently developed permanent partial ptosis and isolated cyclic spasms of the pupil.

### **Methods:**

A four-month old girl presented with right exotropia since birth. Over the ensuing weeks, parents noticed right pupillary size variation several times every day. They also reported right eye photophobia at 8 months of age and intermittent ptosis of the right upper eyelid at 15 months. The latter became permanent at 16 months.

### **Results:**

At 6 months, there was right pupillary cyclical 2mm constriction for 30 seconds followed by 4mm dilatation for 30-60 seconds during which she became photophobic. The right pupil was not reactive to light or accommodation. No cycling spasms of the right extraocular muscles were seen. Right partial ptosis and a complete paresis of right eye elevation, depression and adduction were evident at 16 months. Abduction remained intact. Random brief right upper eyelid twitches were noted intermittently but there was no periodicity. The rest of her examination was unremarkable. Brain MRI was normal. The examination was unchanged on follow up. Strabismus surgery improved her exotropia but the pupillary cyclic spasms persisted.

### **Conclusions:**

Variable manifestations and progression rates of congenital cyclic oculomotor nerve paresis can be seen. Cycling, which may appear later, can be isolated or involve multiple muscles innervated by the oculomotor nerve. The mechanism is unknown but is postulated to be the result of prenatal partial damage to the oculomotor nerve with retrograde degeneration producing changes in the nucleus.

**References:** None.

**Keywords:** Ocular Motility, Pediatric Neuro-Ophthalmology, Pupils

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 81

### Abducent Nerve Palsy In A Patient With Castleman Disease!

Tarek A Shazly<sup>1</sup>, Islam M Zaydan

*University of Pittsburgh, Department of Ophthalmology, Pittsburgh, PA, USA*

#### Introduction:

Castleman disease (angiofollicular lymph node hyperplasia) is a very rare disorder characterized by non-cancerous growths that may develop in the lymph node tissue at a single site or throughout the body.

#### Methods:

Case report and review of literature.

#### Results:

We present a 44-year-old African-American female with history of Castleman's disease with large pelvic mass diagnosed in 2008 status post resection and radiation as well as type II diabetes mellitus and systemic arterial hypertension. She presented in September of 2014 with 3 weeks history of refractory headaches improving on lying down and 4 days of horizontal binocular diplopia more on left gaze following upper respiratory tract infection and persistent cough. Her visual acuity was 20/20 in both eyes with glasses. On examination, she had right longstanding blepharoptosis and left abducent nerve palsy. Her optic discs were pink, with no pallor or swelling and a cup/disc ratio of 0.1. Brain Magnetic Resonance Imaging revealed diffuse meningeal thickening. Blood patch was performed for management of suspected spontaneous intracranial hypotension with transient improvement of headaches. Myelography was performed revealing a dural defect in the region of the right C7-T1.

#### Conclusions:

To the best of the authors' knowledge this is the first report of cranial neuropathy in a patient with Castleman disease.

**References:** None.

**Keywords:** Castleman, Abducent, Nerve, Diplopia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** 1. McAdams, H. Page, et al. "Castleman disease of the thorax: radiologic features with clinical and histopathologic correlation." *Radiology* 209.1 (1998): 221-228. 2. McAdams, H. Page, et al. "Castleman disease of the thorax: radiologic features with clinical and histopathologic correlation." *Radiology* 209.1 (1998): 221-228. 3. Herrada, Juan, et al. "The clinical behavior of localized and multicentric Castleman disease." *Annals of Internal Medicine* 128.8 (1998): 657-662. 4. Meador, Toni L., and John K. McLarney. "CT features of Castleman disease of the abdomen and pelvis." *American Journal of Roentgenology* 175.1 (2000): 115-118.

## Poster 82

### **Nivolumab Plus Ipilimumab Induced Aseptic Meningitis And Bilateral Sixth Nerve Palsy In Metastatic Melanoma Treatment.**

Mitchell Strominger<sup>1</sup>

*Tufts Medical Center / Ophthalmology, Boston, MA, USA*

#### **Introduction:**

Nivolumab and Ipilimumab are recombinant human immunoglobulin monoclonal antibodies that prolong overall survival in metastatic melanoma. Reported adverse effects include orbital inflammation and hypophysitis. Aseptic meningitis is exceedingly rare with one case reported in Ipilimumab monotherapy.

#### **Methods:**

Case report of a 52 year old gentleman who developed progressive bilateral sixth nerve palsy secondary to Nivolumab and Ipilimumab induced aseptic meningitis.

#### **Results:**

The patient was enrolled in a Phase 2 study of Nivolumab given sequentially with Ipilimumab in metastatic melanoma. He received his last dose of Nivolumab when he complained of intermittent horizontal diplopia. An MRI scan of the brain with gadolinium was unremarkable and he received his first dose of Ipilimumab. Three days later he presented with worsening diplopia. On examination his vision was 20/20 with -4.00 OU. He had a relatively comitant 12 diopter intermittent esotropia at distance with -1/4 bilateral abduction deficits. He was treated with a 10 base out press on prism. Two weeks later he had worsening diplopia and demonstrated a 16 diopter constant esotropia and progressive bilateral abduction deficits. Repeat MRI demonstrated diffuse leptomeningeal and cranial nerve enhancement. Lumbar puncture showed a 1 white blood cell, total protein of 80 and negative cytology. He received Solumedrol followed by oral prednisone. Follow up MRI one month later demonstrated resolution of the leptomeningeal enhancement. His ophthalmic examination showed improved abduction and an intermittent esotropia of 10 prism diopters.

#### **Conclusions:**

Progressive 6<sup>th</sup> nerve palsy can occur secondary to aseptic meningitis in patients treated with Nivolumab plus Ipilimumab. This patient is interesting in that he presented with an initial normal MRI and what appeared to be a breakdown of a strabismic myopic divergence insufficiency. Over one month, however the deviation progressed and the leptomeningeal involvement became evident.

#### **References:**

1. Wolchok JD, et al, Nivolumab plus Ipilimumab in advance melanoma, N Engl J Med, 369:122, 2013.

**Keywords:** Sixth Nerve Palsy, Aseptic Meningitis, Nivolumab, Ipilimumab, Melanoma

**Financial Disclosures:** The author had no disclosures.

**Grant Support:** None.

## Combined Third Nerve Palsy And Vertical Gaze Palsy In Midbrain Thalamic Stroke

Kara Fister<sup>1</sup>, Padmaja Sudhakar<sup>1</sup>, Sourabh Lahoti<sup>1</sup>, Flavius Raslau<sup>2</sup>, Sachin Kedar<sup>3</sup>

<sup>1</sup>University Of Kentucky/Dept of Neurology, Lexington, KY, USA, <sup>2</sup>University Of Kentucky, Dept of Neuroradiology, Lexington, KY, USA, <sup>3</sup>University Of Nebraska Medical Center, Dept of Neurology, Omaha, NE, USA

### Introduction:

We present neuro-anatomical substrates for a rare presentation of ipsilateral third nerve palsy and vertical gaze palsy in a patient with rostral midbrain and paramedian thalamic infarct.

### Methods:

Single case report and review of literature

### Results:

A 56 year old male presented with a 3 day history of left hemiparesis and binocular horizontal and vertical diplopia. 3 months ago, a right posterior cerebral artery infarct had resulted in left homonymous hemianopia. At presentation he had dysarthria, left hemiparesis and appendicular ataxia. Neuro-ophthalmic examination revealed a left homonymous superior quadrantanopia (pre-existing), anisocoria (mid-dilated non-reactive right pupil, normal left pupil), right hypertropia and exotropia with limited adduction and depression and decreased bilateral vertical up-gaze saccades without ptosis. CT angiogram revealed a proximal right P1 thrombus. Non-contrast MRI head showed an acute right paramedian thalamic and right rostral midbrain infarct. 5 days later he had skew deviation and torsional nystagmus.

### Conclusions:

Using high resolution MRI sequences we were able to identify the neuroanatomical substrates for this rare clinical presentation.

- Fascicular incomplete pupil involving third nerve palsy: Ksiazek et al had proposed a rostrocaudal organization of the oculomotor nerve fascicles starting with pupillary fibers followed by inferior rectus, inferior oblique, medial rectus, superior rectus, and levator palpebrae superioris. This patient's lesion most likely involved the rostral fascicles (pupillary fibers, inferior rectus, inferior oblique and medial rectus fibers) sparing the caudal fascicles.
- Bilateral vertical upgaze palsy from ipsilateral lesion of the rostral interstitial nucleus of medial longitudinal fasciculus (riMLF), interstitial nucleus of Cajal (INC) and nucleus of posterior commissure (NPC). The paramedian thalamic infarct additionally impaired up-gaze via interruption of supranuclear fibers of the medial thalamus en route to the pretectal and prerubral area.
- Skew deviation and torsional nystagmus from ipsilateral lesion of INC and/or NPC

### References:

1. Khalil M, Malik TG, Farooq K. Weber's syndrome with vertical gaze palsy. J Coll Physicians Surg Pak. 2009 Oct;19(10):668-9.
2. Takami T, Sakaguchi M, Murata K, Nakabayashi H, Nakagawa O, Kawasaki H. [A case report of Weber's syndrome associated with supranuclear vertical gaze palsy caused by the ipsilateral thalamomesencephalic lesion]. No To Shinkei. 1993 May;45(5):461-4. Japanese.
3. Clark JM, Albers GW. Vertical gaze palsies from medial thalamic infarctions without midbrain involvement. Stroke. 1995 Aug;26(8):1467-70.
4. Gentilini M, DeRenzi E, Crisi G. Bilateral paramedian thalamic artery infarcts: report of eight cases. J Neurol Neurosurg Psychiatry. 1987;50:900-909.
5. Saeki N, Murai H, Mine S, Yamaura A. Fascicular arrangement within the oculomotor nerve MRI analysis of a midbrain infarct. J Clin Neurosci. 2000 May;7(3):268-70.
6. Schwartz TH, Lycette CA, Yoon SS, Kargman DE. Clinicoradiographic evidence for oculomotor fascicular anatomy. J Neurol Neurosurg Psychiatry. 1995 Sep;59(3):338.

**Keywords:** Midbrain Infarct, Thalamic Infarct, Vertical Gaze Palsy, Third Nerve Palsy, Hemiparesis

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 84

### A Case Of Abducens Nerve Palsy Followed By Cyclic Esotropia: A Case Report And Review Of The Literature

Takako Sugimoto<sup>1</sup>, Hideki Chuman, Nobuhisa Naoi

*the Department of Ophthalmology, Miyazaki University School of Medicine, Miyazaki, Japan*

#### **Introduction:**

Cyclic esotropia is a rare acquired disorder in which days with esotropia and orthotropia alternate. We encountered a patient with abducens nerve palsy caused by a clival tumor who developed cyclic esotropia during follow-up without treatment.

#### **Methods:**

Case report and review of the literature

#### **Results:**

The right eye of a 6-year-old girl was found to be internally rotated by her mother in February 2009. After 17 days, the girl was referred to our ophthalmology department. On the first examination, her visual acuity was bilaterally 20/20, and cycloplegic refraction revealed no hyperopia. Both pupils measured 7.0 mm in a dark room and 3.5 mm in a bright room, and the reaction to light was rapid and complete. The right eye showed a restriction of abduction, and 25Δ esotropia was noted in the primary position. The left eye moved smoothly and fully in all directions. No abnormality was noted in the eyegrounds. Head MRI demonstrated a lobulated tumorous lesion in the retroclivus, suggesting a chordoma. On examination in the neurosurgery department, the tumor was small, and surgery was expected to be highly invasive; thus, periodic follow-up was indicated. Around August of the same year, her mother noticed alternation of days with esotropia and orthotropia. On follow-up examinations, 25-30Δ esotropia or orthotropia was noted. When there was esotropia, no oculomotor restriction was observed in either eye. A diagnosis of cyclic esotropia was made, and the patient continued to be followed up. Since no growth of the tumor or change in cyclic esotropia was observed, bilateral recession of the medial rectus muscle was performed in 2013. The ocular alignment was normalized after surgery.

#### **Conclusions:**

In our patient, cyclic esotropia occurred secondarily to abducens nerve palsy. According to our review of the literature, one case of traumatic abducens nerve palsy followed by cyclic esotropia has been reported.

**References:** None.

**Keywords:** Abducens Nerve Palsy, Cyclic Esotropia, Clival Tumor, Chiled, Surgery

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Functional MRI And MRI Tractography In Progressive Supranuclear Palsy-Like Syndrome

Michael Vaphiades<sup>1</sup>, Kristina Visscher<sup>3</sup>, Janet Rucker<sup>4</sup>, Surjith Vattoth<sup>2</sup>, Glenn Roberson<sup>2</sup>

<sup>1</sup>University of Alabama at Birmingham/Ophthalmology, Birmingham, AL, USA, <sup>2</sup>University of Alabama at Birmingham/Radiology, Birmingham, AL, USA, <sup>3</sup>University of Alabama at Birmingham/Neurobiology, Birmingham, AL, USA, <sup>4</sup>Icahn School of Medicine at Mount Sinai/Neurology New York, NY, USA

### Introduction:

Progressive supranuclear palsy (PSP)-like syndrome is a rare disorder which occurs most commonly following ascending aortic aneurysm repair, but may occur after other cardiac procedures. The syndrome, which may manifest immediately after surgery or hours or days later, usually includes supranuclear gaze palsy, dysarthria, emotional lability and gait ataxia mimicking PSP.

### Methods:

Contrasted cranial and orbital fat-suppressed magnetic resonance imaging (MRI), MRI tractography and functional MRI (fMRI) were obtained on an 18-year-old woman who underwent an uneventful ascending aortic aneurysm repair and 48 hours later developed a progressive supranuclear palsy (PSP)-like syndrome manifesting with dysarthria, dysphagia (requiring a gastric tube), emotional lability and ophthalmoplegia.

### Results:

Contrasted cranial and orbital fat-suppressed magnetic resonance imaging (MRI), MRI tractography and functional MRI (fMRI) were all normal.

### Conclusions:

Our case attests to the occult nature of this rare and devastating syndrome and perhaps supports the contention that the dysfunction is microscopic in nature and cannot be detected even with advanced neuro-imaging.

### References:

1. Mokri B, Ahlskog E, Fulgham J, Matsumoto J. Syndrome resembling PSP after surgical repair of ascending aorta dissection or aneurysm. *Neurology*. 2004;62:971–973.
2. Kim HT, Shields S, Bhatia K, Quinn N. Progressive supranuclear palsy-like phenotype associated with bilateral hypoxic-ischemic striatopallidal lesions. *Mov Disord*. 2005;20:755–757.
3. Josephs KA, Ishizawa T. A clinicopathological study of vascular progressive supranuclear palsy. *Arch Neurol*. 2002;99:1597–1601.
4. Solomon D, Ramat S, Tomsak R, Reich SG, Shin RK, Zee DS, Leigh RJ. Saccadic palsy after cardiac surgery: characteristics and pathogenesis. *Ann Neurol*. 2008;63:355–365.
5. Antonio-Santos A, Eggenberger E. Asaccadia and ataxia after repair of ascending aortic aneurysm. *Semin Ophthalmol*. 2007;22:33–34.
6. Nandipati S, Rucker JC, Frucht SJ. Progressive supranuclear palsy-like syndrome after aortic aneurysm repair: a case series. *Tremor Other Hyperkinet Mov (N Y)*. 2013 Dec 11;3.
7. Yee RD, Purvin V. Acquired ocular motor apraxia after aortic surgery. *Trans Am Ophthalmol Soc*. 2007;105:152–158.
8. Eggers SDZ, Horn A, Härtig W, Reich DS, Roeber S, Zee DS, Leigh RJ. Saccadic palsy following cardiac surgery: Possible role of perineuronal nets. *Ann NY Acad Sci* 2014 (in press).
9. Hobohm C, Günther A, Grosche J, Rossner S, Schneider D, Brückner G. Decomposition and long-lasting downregulation of extracellular matrix in perineuronal nets induced by focal cerebral ischemia in rats. *J Neurosci Res*. 2005;80:539-548.
10. Petersen SE, Fox PT, Posner MI, Mintun M, Raichle ME. Positron emission tomographic studies of the processing of single words. *J Cogn Neurosci*. 1989;1:153-170.
11. Raichle ME, Fiez JA, Videen TO, MacLeod AM, Pardo JV, Fox PT, Petersen SE. Practice-related changes in human brain functional anatomy during nonmotor learning. *Cereb Cortex*. 1994;4:8-26.
12. Paus T. Location and function of the human frontal eye-field: a selective review. *Neuropsychologia*. 1996;34:475-483.
13. Ruff CC, Blankenburg F, Bjoertomt O, Bestmann S, Freeman E, Haynes JD, Rees G, Josephs O, Deichmann R, Driver J. Concurrent TMS-fMRI and psychophysics reveal frontal influences on human retinotopic visual cortex. *Curr Biol*. 2006;16:1479-1488.

**Keywords:** Progressive Supranuclear Palsy, Aortic Aneurysm, Functional MRI, MRI Tractography, Ophthalmoplegia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 86

### Abducens Palsy In A Patient With Pityriasis Rubra Pilaris

Amanda L Way<sup>1</sup>, Tarek A Shazly, Ellen B Mitchell

*University of Pittsburgh Medical Center/ Department of Ophthalmology, Pittsburgh, PA, USA*

#### **Introduction:**

Pityriasis rubra pilaris (PRP) is rare idiopathic papulosquamous dermatosis of unknown etiology. PRP has been associated with several ocular manifestations including interstitial keratitis, epithelial defects and keratinization of the conjunctiva. However, no previous studies have reported an association with cranial nerve palsy. Here we report a case of abducens palsy in a patient with PRP.

#### **Methods:**

Case report and review of literature.

#### **Results:**

A 61 year old female with a history of PRP and bilateral pseudophakia presented with acute onset horizontal diplopia. She had been diagnosed with biopsy-proven diagnosis of PRP one month prior. She denied a history of recent or remote head trauma. Her medication list included a prednisone steroid taper, cyclosporine, conjugated estrogen and daily multivitamin. Her visual acuity was 20/20 bilaterally with normal pupillary function. Her examination was notable for restriction in abduction on left gaze with normal pursuit and saccadic function. Her anterior slit lamp exam and posterior fundus examination were within normal limits. Contrast enhanced MRI of the brain and orbits did not show evidence of a compressive lesion. Screening tests for hyperlipidemia and hypertension were normal. Observation and a trial of prism glasses were recommended for the patient. On repeat examination 5 months later, the patient had improvement in abduction with a smaller angle esotropia.

#### **Conclusions:**

To our knowledge, cranial neuropathy has not been reported in association with PRP. Here we present a case of abducens nerve palsy in a patient with PRP in the absence of compressive or microangiopathic causes.

**References:** None.

**Keywords:** Ocular Motility, Neuro-Ophth & Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Characterization of Idiopathic Intracranial Hypertension (IIH) in Qatar

Mais N. Alkawaz<sup>1</sup>, Nour B. Barakat<sup>2</sup>, Fatema AlMannaie<sup>2</sup>, Marc Dinkin<sup>3</sup>

<sup>1</sup>Weill Cornell Medical College in Qatar, Doha, Qatar, <sup>2</sup>Hamad Medical Corporation, Doha, Qatar, <sup>3</sup>Weill Cornell Medical College, New York, NY, USA

### Introduction:

IIH is a disease of young obese women that can lead to significant vision loss. Previous studies described the nature of IIH within the Middle East. However, no such data has been published from Qatar. Given the rising epidemic of obesity in Qatar, we hypothesize a more severe IIH presentation and worse visual outcomes.

### Methods:

A retrospective chart review of IIH patients seen at the Neuro-ophthalmology clinic between January 2000 – present was done. All patients satisfied the Modified Dandy criteria.

### Results:

34 patients were included; mean age of presentation was 31.6 ( $\pm$  11.6) years with a 4.5:1 female/male ratio. 90.63% of patients had BMI $>$ 25. Most common presenting symptoms were headache (76.67%) and transient visual obscurations (73.33%). 93.33% had papilledema, 73.1% had visual field defects, 32.1% had reduced visual acuity, and 87.5% had thickened RNFL (average =170.4mm). On MRI, 45.5% of patients had an empty sella, and 30.3% had papilledema. Average CSF opening pressure was 36.6 cmH<sub>2</sub>O. All patients were treated medically, no surgical interventions were performed. At an average of 6 months follow-up time: six patients were lost to follow up, 21.4% had persistent visual loss, 48% had VF defects, 77% had papilledema, and 69.2% had RNFL thickening. 42.3% of patients developed RNFL atrophy. No significant weight changes were observed.

### Conclusions:

Incidence of obesity in our patients was higher than previously reported IIH studies in the gulf region <sup>(1,2,3)</sup>, suggesting a more robust correlation between obesity and IIH in Qatar. Patients presented with larger VF defects, more papilledema reflected by RNFL thickening, and higher CSF opening pressure. Recovery was sub-optimal with persistent field defects in most patients and significant number of RNFL atrophy. These observations indicate a more severe course of IIH in Qatar, which necessitates introducing new therapeutic modalities and emphasizing patient education and disease prevention.

### References:

1. Mezaal, M., and M. Saadah. "Idiopathic Intracranial Hypertension in Dubai: Nature and Prognosis." *Acta Neurologica Scandinavica* 112.5 (2005): 298-302.
2. Alkali, Nurah, Malli Dorasanamma, Abdulkader Daif, and Mansoura Almoallem. "Prognosis of Idiopathic Intracranial Hypertension in Saudi Arabia." *Annals of African Medicine* 10.4 (2011): 314.
3. Idiculla, Thara, George Zachariah, Keshav Br, and Nasir Mohamood. "The Incidence and Prevalance of Idiopathic Intracranial Hypertension in South Sharaqiah Region, Oman." *Oman Journal of Ophthalmology* 6.3 (2013): 189.

**Keywords:** Idiopathic Intracranial Hypertension, Obesity, Qatar

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 88

### Osmometry of Cerebrospinal Fluid from Patients with Idiopathic Intracranial Hypertension (IIH)

Steffen E. Hamann<sup>1</sup>, Elisabeth A. Wibroe<sup>1</sup>, Hanne M. Yri<sup>2</sup>, Rigmor H. Jensen<sup>2</sup>

<sup>1</sup>Department of Ophthalmology, Glostrup Hospital, University of Copenhagen, Glostrup, Denmark, <sup>2</sup>Danish Headache Center, Department of Neurology, Glostrup Hospital, University of Copenhagen, Glostrup, Denmark

#### Introduction:

This study aims to investigate osmolality of cerebrospinal fluid (CSF) from patients with idiopathic intracranial hypertension (IIH). IIH is a disorder of increased intracranial fluid pressure (ICP) of unknown etiology. Investigating CSF osmolality could provide knowledge of underlying pathophysiological mechanisms.

#### Methods:

CSF samples were collected from individuals referred on suspicion of IIH from 2011-2013. Subjects included as patients fulfilled the Modified Dandy Criteria; individuals in whom IIH was refuted were included as controls. ICP was measured by lumbar puncture manometry at inclusion and in patients after three months treatment with weight loss and acetazolamide or topiramate. Osmolality was measured with a Vapor Pressure Osmometer.

#### Results:

We collected 90 CSF samples from 38 patients and 28 controls. We found no significant differences in osmolality between groups: Neither when newly diagnosed patients were compared with the control group ( $p = 0,86$ ), when newly diagnosed patients were compared to the osmolality after 3 months treatment ( $p = 0,97$ ) nor when patients with normalized pressure after 3 months had their CSF osmolality compared to their baseline values ( $p = 0,79$ ). We found no significant differences when comparing osmolality in individuals with normal ICP from 6-25 cmH<sub>2</sub>O ( $n = 47$ ) with: patients with moderately elevated ICP from 26-45 cmH<sub>2</sub>O ( $n = 37$ ) ( $p = 0,20$ ) and patients with high ICP from 46-65 cmH<sub>2</sub>O ( $n = 6$ ) ( $p = 0,88$ ) respectively. There was no correlation between osmolality and ICP, BMI, age and body height respectively. Mean CSF osmolality was 270 mmol/kg ( $\pm 1$  SE, 95% confidence interval 267-272) for both patients and controls.

#### Conclusions:

CSF osmolality was normal in patients with IIH and there was no relation to treatment outcome, ICP, BMI, age and body height. Mean CSF osmolality was 270 mmol/kg for both patients and controls. Other pathophysiological mechanisms behind IIH must be searched.

**References:** None.

**Keywords:** Idiopathic Intracranial Hypertension, Pseudotumor Cerebri, High Intracranial Pressure/Headache, Cerebrospinal Fluid/CSF, Osmolality

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 89

### Corneal Biomechanics : A Journey Through Uncharted Territories

Ashwin Mohan, Chaithra D Aroor<sup>1</sup>, Rohit Shetty, Chaitra Jayadev, Tejal SJ, Bhujang Shetty

*Narayana Nethralaya, Bangalore, India*

#### Introduction:

To ascertain the relationship of corneal biomechanics with the severity of visual field loss in patients with Papilloedema due to Idiopathic Intracranial Hypertension (IIH).

#### Methods:

15 patients referred to the Neuro-ophthalmology department, diagnosed to have IIH were included in the study. Best corrected visual acuity, anterior segment and pupil evaluation, fundus evaluation, optic nerve photography, corneal biomechanics measured using CORVIS ST(Oculus Germany), B scan ultrasonography to detect fluid around the optic nerve and visual field analysis were performed. Corneal biomechanics were compared to the severity of visual field loss.

#### Results:

Patients with deformation amplitude (DA) <1.15 were considered normal, DA >1.15 subnormal and more than 1.30 is considered to have poor biomechanics. The mean DA of 8 eyes out of the total cohort was 1.10, whereas 7 eyes had a mean DA of 1.30. 6 eyes which showed progression and severe visual field defect (mean >5db) had poor biomechanics (1.30) compared to those with stable fields and non progressive IIH, suggesting that biomechanical parameters may be one of the possible factors in progression.

#### Conclusions:

Corneal biomechanics may be an indirect indicator of scleral biomechanics. Weaker ocular biomechanics might be associated with greater visual field loss in patients with Papilloedema due to IIH. Thus corneal biomechanics may be an indirect parameter in prognosticating IIH patients. However our cohort size is small and a study on larger cohort would be needed to bring a better insight into this domain.

**References:** None.

**Keywords:** Corneal Biomechanics, Idiopathic Intracranial Hypertension, Progression, Fluid Around Optic Nerve, CORVIS

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Clinical and Prognostic Significance of CSF Closing Pressure in Pediatric Pseudotumor Cerebri Syndrome

Shannon J Beres<sup>1,2</sup>, Claire A Sheldon<sup>1,2</sup>, Chantal Boisvert<sup>5</sup>, Christina L Szperka<sup>7</sup>, Grace L Paley<sup>1,4</sup>, Evanette K Burrows<sup>6</sup>, Marianne R Chilutti<sup>6</sup>, Geraldine W Liu<sup>7</sup>, Shana E McCormack<sup>3,4</sup>, Grant T Liu<sup>1,2,4</sup>

<sup>1</sup>Hospital of the University of Pennsylvania / Departments of Neurology and Ophthalmology, Philadelphia, PA, USA, <sup>2</sup>The Children's Hospital of Philadelphia / Division of Ophthalmology, Neuro-ophthalmology Service, Philadelphia, PA, USA, <sup>3</sup>The Children's Hospital of Philadelphia / Division of Endocrinology and Diabetes, Philadelphia, PA, USA, <sup>4</sup>University of Pennsylvania / Perelman School of Medicine Philadelphia, PA, USA, <sup>5</sup>Rady Children's Hospital San Diego / Division of Ophthalmology San Diego, CA, USA, <sup>6</sup>The Children's Hospital of Philadelphia / Center for Biomedical Informatics Philadelphia, PA, USA, <sup>7</sup>The Children's Hospital of Philadelphia / Division of Neurology Philadelphia, PA, USA

### Introduction:

The significance of CSF opening pressure (OP) in diagnostic lumbar puncture in pediatric pseudotumor cerebri syndrome (PTCS) has been well-studied.<sup>[1]</sup> Little consensus exists as to whether the closing pressure (CP) or removing large volumes of CSF has any prognostic utility.

### Methods:

This is a retrospective observational series of 93 patients with definite pseudotumor cerebri syndrome (by 2013 revised criteria). Age, gender and body mass index Z-scores (BMI-Z) at diagnosis, as well as OP, CP and volume of CSF removed were abstracted. The primary outcome measure was time to resolution of papilledema by ophthalmoscopy from initial diagnosis.

### Results:

No significant differences in gender, age or BMI-Z were observed between subjects with (N=35) and without (N=58) documented CP. The 35 subjects were 74% female, average 12.9yrs (4.9 SD), and had a mean BMI-Z of 1.46 (1.10 SD). Mean OP was 413 mmH<sub>2</sub>O (98 SD). Mean CP was 173 mmH<sub>2</sub>O (60 SD) and removed volume documented in 17 subjects ranged 7-35ml (median=16). Mean change in pressure ( $\Delta P = OP - CP$ ) was 240mmH<sub>2</sub>O (median = 230). BMI-Z ( $r=0.38$ ,  $p=0.02$ ) and age ( $r=0.32$ ,  $p=0.06$ ) were both positively associated with  $\Delta P$ , likely driven by higher OP related to both BMI-Z ( $r=0.51$ ,  $p=0.002$ ) and age ( $r=0.38$ ,  $p=0.02$ ). Median time to resolution of papilledema was similar (119 days vs 113 days,  $p=0.17$ ) in children with  $\Delta P$  below versus above the median. Median time to resolution of papilledema in children with CSF volume removed above vs. below the median was insignificantly shorter (80 vs 113 days,  $p=0.30$ ).

### Conclusions:

No significant effect of change in pressure or amount of CSF removed on time to resolution of papilledema in pediatric PTCS was detected.

### References:

1. Avery RA, Licht DJ, Shah SS, et al. CSF opening pressure in children with optic nerve head edema, Neurology, 76, 1658-1661, 2012..

**Keywords:** Pseudotumor, Closing Pressure, Papilledema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 91

### Subretinal Fluid in Idiopathic Intracranial Hypertension: Clinical Course and Outcome

John J. Chen<sup>1</sup>, Matthew J. Thurtell<sup>2, 4, 5</sup>, Reid A. Longmuir<sup>2, 5</sup>, Mona K. Garvin<sup>3, 5</sup>, Jui-Kai Wang<sup>3, 5</sup>, Michael Wall<sup>1, 4, 5</sup>, Randy H. Kardon<sup>2, 5</sup>

<sup>1</sup>Mayo Clinic, Department of Ophthalmology, Rochester, MN, USA, <sup>2</sup>University of Iowa Hospitals and Clinics, Department of Ophthalmology and Visual Sciences, Iowa City, IA, USA, <sup>3</sup>University of Iowa, Department of Electrical and Computer Engineering, Iowa City, IA, USA, <sup>4</sup>University of Iowa Hospitals and Clinics, Department of Neurology Iowa City, IA, USA, <sup>5</sup>Department of Veterans Affairs Iowa City, IA, USA

#### Introduction:

Decreased visual acuity at presentation in idiopathic intracranial hypertension (IIH) can be produced by several mechanisms.<sup>1-3</sup> When resulting from subfoveal subretinal fluid (SRF) and/or photoreceptor disruption, it is usually reversible, but visual recovery may be delayed following resorption of SRF. We examined the incidence and prognosis of SRF in IIH.

#### Methods:

A retrospective review of 660 patients with IIH (2009-2013) identified 10 eyes from 8 patients (1.2%) with SRF on OCT and 8 eyes from 5 patients (0.75%) with photoreceptor disruption (disruption of the IS/OS junction) without SRF on OCT, producing best-corrected visual acuity (BCVA) of 20/25 or worse on presentation. Fundus photography, spectral-domain OCT of the disc and macula, and perimetry were used to evaluate SRF, photoreceptor disruption, and visual prognosis. The volume of SRF was determined by segmentation of the macula OCT with the Iowa Reference Algorithm.

#### Results:

The volume of SRF correlated with BCVA at presentation ( $r^2 = 0.7156$ ). On average, SRF resolved in 2.2 weeks (STD 0.4, range 2-3 weeks) after starting treatment, but there was often residual photoreceptor disruption and persistently decreased BCVA. Photoreceptor disruption subsequently resolved with complete visual recovery over an average of 1.8 weeks (STD 1.6, range 0-3 weeks) following resorption of SRF. In patients with photoreceptor disruption at presentation, visual and photoreceptor recovery occurred an average of 3.75 weeks (STD 1.9, range 1-6 weeks) after initiation of treatment. There was no significant difference in the initial BCVA of patients with SRF vs. photoreceptor disruption only (average LogMAR VA of 0.375 and 0.25 respectively,  $p=0.41$ ). All recovered visual acuity to 20/20, although there was often residual photoreceptor disruption nasal to the fovea, even on extended follow-up.

#### Conclusions:

Vision loss from SRF is reversible in IIH, but maximum recovery after fluid resorption occurs only after subsequent photoreceptor reorganization has taken place.

#### References:

1. Corbett JJ, Savino PJ, Thompson HS, et al. Visual loss in pseudotumor cerebri. Follow-up of 57 patients from five to 41 years and a profile of 14 patients with permanent severe visual loss. Arch Neurol. 1982;39:461-474.
2. Mitchell DJ, Steahly LP. Pseudotumor cerebri and macular disease. Retina. 1989;9:115-117.
3. Hoyer VJ, 3rd, Berrocal AM, Hedges TR, 3rd, Amaro-Quireza ML. Optical coherence tomography demonstrates subretinal macular edema from papilledema. Arch Ophthalmol. Sep 2001;119(9):1287-1290.

**Keywords:** Idiopathic Intracranial Hypertension, Subretinal Fluid, Optical Coherence Tomography, Photoreceptor Disruption

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 92

### Overdiagnosis Of Idiopathic Intracranial Hypertension (IIH).

Adeniyi Fisayo<sup>1</sup>, Valerie Biousse<sup>1,2</sup>, Nancy J. Newman<sup>1,2,3</sup>, Beau B. Bruce<sup>1,2,4</sup>

<sup>1</sup>Emory University, Department of Ophthalmology, Atlanta, GA, USA, <sup>2</sup>Emory University, Department of Neurology, Atlanta, GA, USA, <sup>3</sup>Emory University, Department of Neurological Surgery, Atlanta, GA, USA, <sup>4</sup>Emory University, Department of Epidemiology Atlanta, GA, USA

#### Introduction:

Primary benign headaches are common, occurring in up to 24% of young women. This, combined with the increasing prevalence of obesity and awareness of IIH, has led to an increase in costly and invasive evaluations for headache, often in the absence of documented papilledema. Our objective was to quantify overdiagnosis of IIH among patients seen on our Neuro-Ophthalmology service.

#### Methods:

Review of new patients seen between 11/01/2013 and 03/21/2014. Patients referred for a working diagnosis of IIH, abnormal optic nerve appearance, or suspicion of IIH were included. We applied the Diagnosis Error Evaluation and Research (DEER) taxonomy tool to classify cases according to the location and type of error in the diagnostic process.<sup>1,2</sup>

#### Results:

84/664 new patients (13%) were seen to either rule out IIH (n=43; 34 abnormal optic nerve appearance, 9 headache) or for management of IIH (n=41; 98% women, median age 31 (IQR 22–41), mean BMI 35 kg/m<sup>2</sup>). 20/41 (48.7%) patients referred for IIH did not have IIH. Final diagnoses were pseudopapilledema (9[45%]), primary headache (4[20%]), optic atrophy (3[15%]) and one each of optic disc drusen, optic neuritis, sequential NAION, dominant optic atrophy, and physiologic blind spot. 17/20 had a lumbar puncture, 2/20 had >1 lumbar puncture, 18/20 had a brain-MRI, 3/20 had an MR-venogram/CT-venogram, 1/20 had a lumbar drain, 2/20 were referred for surgery. Diagnostic errors resulted primarily from failure to appreciate or misinterpretation of a physical examination finding (13[65%]), failure to order or misinterpretation of an appropriate test (3[15%]), errors in assessment (2[10%]), and poor history (1[5%]).

#### Conclusions:

Diagnostic errors resulted in the incorrect diagnosis of IIH in 48.7% of patients and prompted unnecessary tests in 44%, invasive procedures in 41% and missed diagnoses in 10%. The most common error was inaccurate funduscopic examination in headache patients, reinforcing the need for rapid and easy access to neuro-ophthalmologists.<sup>3</sup>

#### References:

1. Schiff GD, Hasan O, Kim S, et al. Diagnostic error in medicine. Arch Intern Med, 169(20), 1881-1887, 2009.
2. Schiff GD, Kim S, Abrams R, et al. Diagnosing diagnosis errors: Lessons from a multi-institutional collaborative project. Advances in Patient Safety: From Research to Implementation Vol 2. Agency for Healthcare Research and Quality. Website: [www.ahrq.gov/qual/advances](http://www.ahrq.gov/qual/advances). Accessed July 22, 2014
3. Sadun AA, Chu ER, Boisvert CJ. Neuro-ophthalmology safer than MRI? Ophthalmology, 120(4), 879, 2013

**Keywords:** Pseudotumor Cerebri, Idiopathic Intracranial Hypertension, Papilledema, Headache, Diagnostic Error

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York, and by NIH/NEI core grant P30-EY06360 (Department of Ophthalmology). Dr. Bruce receives research support from the NIH/NEI (K23-EY019341).

## An Update On Terson Syndrome: Prevalence And Prognosis

Philip S. Garza<sup>1</sup>, Beau B. Bruce<sup>1,2,3</sup>, Nancy J. Newman<sup>1,2,4</sup>, Valérie Biousse<sup>1,2</sup>

<sup>1</sup>Department of Ophthalmology, Emory University, Atlanta, GA, USA, <sup>2</sup>Department of Neurology, Emory University, Atlanta, GA, USA, <sup>3</sup>Department of Epidemiology, Emory University, Atlanta, GA, USA, <sup>4</sup>Department of Neurological Surgery, Emory University Atlanta, GA, USA

### Introduction:

Terson syndrome (TS), originally defined as vitreous hemorrhage (VH) in the setting of subarachnoid hemorrhage (SAH), now includes any intraocular hemorrhage (subretinal, intraretinal, subhyaloid, or vitreous) secondary to acute intracranial hemorrhage (ICH) and elevated intracranial pressure. Bedside ocular fundus imaging technology may allow for improved diagnosis of TS in patients acutely ill with ICH. We reviewed the literature for publications on the prevalence and prognostic significance of intraocular hemorrhage in patients with acute ICH.

### Methods:

Medline and Embase databases (English, French, and German) were searched using the keywords "TS" and the combination of "intraocular hemorrhage" (and its subtypes, e.g., "VH," "subhyaloid hemorrhage," "intraretinal hemorrhage") and "ICH" (and its subtypes, e.g., "SAH," "intraparenchymal hemorrhage," "hemorrhagic stroke"). We excluded studies enrolling fewer than 10 patients and cohort studies enrolling nonconsecutive ICH patients. The prevalence of TS and its subtypes was calculated, and mortality rates were compared for those with and without TS.

### Results:

112 publications were included: 16 cohort studies (11 prospective/5 retrospective) and 8 case series (1 prospective/7 retrospective). 165/978 patients (16.9%) assessed prospectively had TS; 222/786 patient records (28.2%) reviewed retrospectively documented TS. The overall prevalence of TS was 21.9%. In studies reporting subtypes of intraocular hemorrhage, retinal hemorrhages were most common (63.5%), followed by subhyaloid hemorrhages (20.5%) and VH (19.6%). Mortality was higher among those with TS than those without TS (24% vs. 10%,  $p=0.01$ ).

### Conclusions:

TS is associated with higher mortality in patients with acute ICH. Among studies reporting subtypes of intraocular hemorrhage, VH ("classic" TS) was the least common type of intraocular hemorrhage observed, whereas retinal hemorrhages were far more common. Systematic examination of the ocular fundus for all types of intraocular hemorrhage, perhaps using new bedside fundus imaging technologies, is important for identifying ICH patients most at risk for poor outcomes and in need of ophthalmic follow-up.

**References:** None.

**Keywords:** High Intracranial Pressure, Subarachnoid Hemorrhage, Retina, Fundus Imaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This work was supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York. Mr. Garza receives research support from the Emory Eye Center/Research to Prevent Blindness Pilot Grant and from the NIH/NCATS (TL1-TR000456-08) via the Atlanta Clinical and Translational Science Institute (ACTSI). Dr. Bruce receives research support from the NIH/NEI (K23-EY019341).

## Poster 94

### Aberrant Presentation of Pseudotumor Cerebri

Sawyer B. Hall<sup>1, 2</sup>, Marc H. Levy<sup>2</sup>, Jody G. Abrams<sup>2</sup>

<sup>1</sup>Lake Erie College of Osteopathic Medicine, Bradenton, FL, USA, <sup>2</sup>Sarasota Retina Institute, Sarasota, FL, USA

#### Introduction:

The purpose of this abstract is to illustrate a case of Pseudotumor Cerebri that presented with a third nerve palsy instead of the typical sixth nerve palsy. A 24-year-old white woman presented with a two day history of headache and blurred vision. Bilateral papilledema and a complete third nerve palsy were noted during her hospital admission.

#### Methods:

Neuro-Ophthalmic evaluation, Lumbar Puncture, Computerized Tomography (CT) of the brain with and without contrast, CT Angiography, and Magnetic Resonance Venography were also performed while she was admitted at a local hospital. She was examined by Ocular Coherence Tomography (OCT) and Funduscopy exams on follow up visits. Orthoptic examination was done regularly.

#### Results:

Neuro-ophthalmic evaluation showed bilateral papilledema and complete third nerve palsy on the right side. Non-traumatic Lumbar Puncture showed an elevated opening pressure at 33cm H<sub>2</sub>O, but normal cytology, culture, protein, and glucose. The CT with and without contrast, CTA, and MRV showed normal cerebral and vascular anatomy. OCT showed NFL thickness of 339 microns OD and 264 microns OS. Funduscopy exam yielded optic disc edema bilaterally. Orthoptic evaluation showed a right complete third nerve palsy. Her symptoms and findings resolved when the patient was treated with Prednisone 10mg QD and Acetazolamide 1000mg QD over the course of 10 weeks.

#### Conclusions:

In light of the series of negative imaging, findings on Lumbar Puncture, clinical presentation, and the patients' response to the standard of care in the treatment of Pseudotumor Cerebri, it can be concluded that her third cranial nerve palsy was directly associated with this condition. This is an isolated and unreported presentation of an otherwise common disease. To conclude, any patient with sudden onset third nerve palsy should be secondarily worked up for Pseudotumor Cerebri if vascular and cerebral causes have been appropriately ruled out.

**References:** None.

**Keywords:** Pseudotumor Cerebri, Ocular Motility, High Intracranial Pressure/ Headache

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 95

### Jugular Vein Thrombosis as a cause of Intracranial Hypertension

Josepha Horowitz<sup>1</sup>, Keren Zissman

*Carmel Medical Center, Haifa, Israel*

#### Introduction:

Cerebral venous sinus thrombosis is one of the well-known causes of increased intracranial pressure (ICP)<sup>1</sup>. Jugular vein thrombosis as a sole cause of increased ICP was rarely described before <sup>2,3</sup>.

#### Methods:

Retrospective case series

#### Results:

Two patients with increased ICP secondary to jugular vein thrombosis are presented. Case 1: a 38-y-old healthy cardiologist who presented with complaints of localized pain at the right mastoid process and along the sternocleidomastoid muscle. A week later he noticed a blurred vision in the lateral field of his left eye. Ocular examination was unremarkable except bilateral papilledema. Brain CT and MRI were normal. LP showed an opening pressure of 430 mm H<sub>2</sub>O and normal content. Thrombosis of the right jugular vein was demonstrated with Doppler of the cervical veins, and confirmed by CTV of the brain and neck. Case 2: 37 y-old female presented to the emergency room with complains on headache, vomiting and chest pain. MRI/MRV of the brain was normal. Chest CT/CTA revealed massive left pulmonary emboli. She presented to the ophthalmology clinic two months later with diplopia consistent with divergence insufficiency and right jugular vein thrombosis was demonstrated by Doppler. LP was not done because of anticoagulation until 6 months later, when she developed papilledema. Opening pressure was 380 mm H<sub>2</sub>O.

#### Conclusions:

Unilateral jugular vein thrombosis may cause increased ICP and should be considered in the differential diagnosis of patients with increased ICP-related signs and symptoms.

#### References:

1. Crassand I, Bousser MG. Cerebral Venous Thrombosis. J Neurol. Ophthalmol. Vol 24: 156-163, 2004.
2. Masood I, While A. Bilateral jugular vein thrombosis: a rare cause of papilloedema. Eye Vol 20: 249-250, 2006
3. Duke BJ, Ryu RK, Brega KE, Coldwell DM. Traumatic bilateral jugular vein thrombosis: case report and review of the literature. Neurosurgery 41: 680-3, 1997.

**Keywords:** Jugular Vein, Thrombosis, Intracranial Hypertension, Papilledema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 96

### Autism Spectrum Disorder in Pediatric Pseudotumor Cerebri Syndrome

Anne K. Jensen<sup>1,2</sup>, Claire A. Sheldon<sup>1,2</sup>, Grace L. Paley<sup>1,3</sup>, Grant T. Liu<sup>1,2,3</sup>, Shana E. McCormack<sup>3,4</sup>

<sup>1</sup>Departments of Neurology and Ophthalmology, Hospital of the University of Pennsylvania, Philadelphia, PA, USA, <sup>2</sup>Division of Ophthalmology, Neuro-ophthalmology Service, The Children's Hospital of Philadelphia, Philadelphia, PA, USA, <sup>3</sup>Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA, <sup>4</sup>Division of Endocrinology and Diabetes, The Children's Hospital of Philadelphia Philadelphia, PA, USA

#### Introduction:

Having recently cared for pediatric patients with suspected pseudotumor cerebri syndrome (PTCS) and concurrent autism spectrum disorder (ASD), we examined our cohort for additional evidence of this association.

#### Methods:

Design was a retrospective observational case series. Possible PTCS subjects ages 2-18 years were identified by ICD-9 search and review of a pediatric neuro-ophthalmologist's database (July 1993-April 2013). Recently revised diagnostic criteria<sup>1</sup> assigned "definite" (n=78) and "probable" (n=10) cases. "Suspected" cases met all criteria but were missing either LP (n=47), MRI/MRV (n=36), or documented papilledema (n=15). Remaining cases were considered "not PTCS" and were excluded. ASD diagnoses within this PTCS cohort were identified by ICD-9 search and database review. Patient-specific factors extracted from the medical record included demographics, anthropometrics, and where relevant, ASD diagnostic criteria and common co-morbidities.

#### Results:

In this cohort (n=186), 3 subjects had concurrent ASD per sub-specialist assessment. All 3 were non-obese males; 2 had suspected PTCS missing only an MRI/MRV, and 1 had definite PTCS. The most common ASD co-morbidities included intellectual disability (3/3), language delay (3/3), sleep difficulty (3/3), aggression (3/3), self-injury (2/3), motor abnormality (2/3), and ADHD (2/3). The rate of ASD among all subjects with definite, probable, or suspected PTCS was 1.6% (3/186). The rate of ASD among non-obese males was between 6.8% (3/44, conservatively assuming all boys missing anthropometric measurements were non-obese), and 9.1% (3/33, assuming all boys missing anthropometric measurements were obese). These rates seem higher than would be expected from ASD population estimates (0.62-2%).<sup>2</sup> Since April 2013, two subsequent PTCS cases in boys with ASD have been identified: one obese with probable PTCS, and one non-obese with definite PTCS.

#### Conclusions:

This experience suggests a possible association between ASD and pediatric PTCS among non-obese boys. Further studies with larger sample sizes are needed to confirm these findings and to elucidate potential underlying mechanism(s).

#### References:

1. Friedman, Liu, Digre, Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children, *Neurology*, 81(13), 1159-65, 2013.
2. Meng-Chuan, Lombardo, Baron-Cohen, Autism, *Lancet*, 383, 896-910, 2014.

**Keywords:** Pseudotumor Cerebri, Pediatric Neuro-Ophthalmology

**Financial Disclosures:** G.T.L. acknowledges consultancy for Ipsen (Cambridge, MA).

**Grant Support:** S.E.M. acknowledges grant support from K12: 5K12DK094723-03, PI: S. Willi.

## Poster 97

### Lumbar Drain in Fulminant Intracranial Hypertension

Mays A El-dairi<sup>1</sup>, Kim Jiramongkolchai<sup>1</sup>, Edward G Buckley<sup>1</sup>, Mohammed T Bhatti<sup>1, 2</sup>, Robert E Wiggins<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, Duke Eye Center, Durham, NC, USA, <sup>2</sup>Department of Neurology, Duke University Medical Center, Durham, NC, USA, <sup>3</sup>Asheville Eye Associates, Asheville, NC,

#### Introduction:

Fulminant intracranial hypertension (FIH) is a subtype of intracranial hypertension characterized by rapid severe progressive vision loss. The current practice recommends ventricular peritoneal shunt (VPS), lumbar peritoneal shunt (LPS), or optic nerve sheath (ONSF) be performed immediately to halt or reverse vision loss [1]. These interventions carry significant risk of morbidity with up to 75% of shunts failing at 2 years [2]. Here we describe the successful management of 2 teenagers with FIH with a temporary lumbar drain.

#### Methods:

A chart review of the clinical course of 2 female teenagers with FIH treated immediately with lumbar drain followed by medical therapy.

#### Results:

Two obese pubertal girls with acute severe bilateral visual loss (case 1: light perception OD and 20/800 OS; case 2: 20/400 OD and counting fingers OS) of less than 2 weeks duration had bilateral papilledema (grade 5) and an abducens palsy. Both patients had normal neuroimaging, intracranial pressure >35 cm H<sub>2</sub>O, and initially treated with acetazolamide (1500 mg daily or less than 12 mg/Kg). Each received 1g/day of intravenous methylprednisolone, 2g/day acetazolamide and placement of a lumbar drain (11 days for case 1 and 3 days for case 2). They responded with marked decrease in the papilledema, resolved abducens palsy and improvement in vision (Case 1 at 18 months visual acuity of 20/200 OD and 20/50 OS; case 2 at 3 months visual acuity of 20/70 OD and 20/60 OS).

#### Conclusions:

A temporary lumbar drain may be considered in cases of FIH and can circumvent the need for a cerebrospinal fluid diversion procedure or optic nerve sheath fenestration.

#### References:

1. Thambisetty M, Lavin PJ, Newman NJ, Biousse V, Fulminant idiopathic intracranial hypertension, *Neurology*, 68, 229-32, 2007.
2. McGirt MJ, Woodworth G, Thomas G, Miller N, Williams M, Rigamonti D, Cerebrospinal fluid shunt placement for pseudotumor cerebri-associated intractable headache: predictors of treatment response and an analysis of long-term outcomes, *Journal of Neurosurgery*, 101, 627-32, 2004.

**Keywords:** Fulminant Intracranial Hypertension, Lumbar Drain, Papilledema, Pediatric Patient, Severe Vision Loss

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 98

### Papilledema With Unilateral Occlusion Of Internal Jugular Vein

Abhishek Thandra<sup>1</sup>, Bokkwan Jun<sup>2</sup>, Miguel Chuquilin Arista<sup>1</sup>

<sup>1</sup>University of Missouri Health System/Department of Neurology, Columbia, MO, USA, <sup>2</sup>University of Missouri Health System/Department of Ophthalmology, Columbia, MO, USA

#### Introduction:

To report and review a case of papilledema and increased intracranial pressure in setting of unilateral complete occlusion of internal jugular vein

#### Methods:

This is a descriptive case report based on clinical records, patient observation and analysis of diagnostic studies.

#### Results:

A 45-year-old man, who had history of squamous cell laryngeal carcinoma, was found to have papilledema on his ophthalmic evaluation 6 months after the completion of chemotherapy and radiation. MRI brain and MRV brain were unremarkable and lumbar puncture showed opening pressure was 35cmH<sub>2</sub>O at lateral decubitus position. The patient started Diamox but could not tolerate and had to stop. CTA neck was done for the evaluation of tightening and feeling hard in the right side of neck and it showed complete occlusion of the right jugular vein and patent left jugular vein. The patient started Topamax and it was titrated up to 100mg twice a day. Without improvement of headache and blurry vision with the medical treatment, ventriculoperitoneal shunt placement was performed. After the shunt placement, the patient noticed his blurry vision and also headache had improved.

#### Conclusions:

This is a case of papilledema and increased intracranial pressure in setting of complete occlusion of unilateral jugular vein. Bilateral occlusion or stenosis of jugular veins have been known to cause the increased intracranial pressure. It is uncommon but complete occlusion of unilateral jugular vein also could be related to increased intracranial pressure. MRV or CTV of neck may be considered as a work-up for the medically refractory idiopathic intracranial hypertension.

**References:** None.

**Keywords:** High Intracranial Pressure/Headache, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 99

### Papilledema Secondary to Internal Jugular Vein Thrombosis in a Dialysis Patient

Shauna W. Berry<sup>1,2</sup>, Clint W. Kellogg<sup>1,2</sup>, Kathryn E. Ireland<sup>1,2</sup>, Matthew D. Kay<sup>2</sup>

<sup>1</sup>Larkin Community Hospital, South Miami, FL, USA, <sup>2</sup>Nova Southeastern University, Fort Lauderdale, FL, USA

#### Introduction:

We report a case of papilledema due to unilateral jugular venous thrombosis resulting from temporary hemodialysis using a jugular venous approach.

#### Methods:

A 64-year old thin woman presented for neuro-ophthalmology consultation complaining of intermittent graying of vision lasting seconds at a time, which was not exacerbated by positional changes. The patient denied headache, diplopia, vision loss, or pulsatile tinnitus.

#### Results:

Bilateral disc edema was found on physical examination. Humphrey visual field demonstrated mild central depression in the right eye, while the left eye showed moderate enlargement of the physiologic blind spot, an inferonasal step, and a superior arcuate scotoma. Brain MRI was unremarkable, while the brain MRV demonstrated a congenitally hypoplastic left vertebral artery. A venous Doppler of the neck was ordered given her history of hemodialysis via the right internal jugular vein (IJV) demonstrating absence of Doppler flow consistent with thrombosis. Subsequently, a lumbar puncture performed in the lateral decubitus position revealed opening pressure of 40 cm H<sub>2</sub>O and normal CSF constituents.

#### Conclusions:

In this case, the thrombosed jugular vein would have gone undetected based upon the findings on initial imaging. A thorough history and high level of clinical suspicion led to further testing which elucidated the etiology of the patient's bilateral disc edema. The elevated ICP appeared to be a consequence of right IJV thrombosis superimposed upon a congenitally atretic left vertebral artery resulting in cerebral venous hypertension. At presentation, there was already evidence for visual loss secondary to papilledema. It is important for physicians to be aware of possible consequences of IJV access thereby enabling rapid evaluation to prevent progressive visual loss in this unusual clinical scenario. This case brings into question whether thrombosis of the IJV found incidentally in dialysis patients should prompt fundus examination looking for evidence of papilledema.

**References:** None.

**Keywords:** Papilledema, Venous Thrombosis, Hemodialysis, Cerebral Venous Hypertension

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Ultramicroscopic study of the Optic Nerve Sheath in Patients with Severe Vision Loss from Idiopathic Intracranial Hypertension- Methodology**

Joshua W Evans<sup>1</sup>, Marla Davis<sup>1</sup>, Sachin Kedar<sup>2, 3</sup>, Deepta Ghate<sup>3</sup>, Peter J Timoney<sup>1</sup>, Richard Kielar<sup>1</sup>, Bruce E Maley<sup>4</sup>, William O'Connor<sup>5</sup>

<sup>1</sup>University of Kentucky/Ophthalmology, Lexington, KY, USA, <sup>2</sup>University of Nebraska Medical Center/Neurology, Omaha, NE, USA, <sup>3</sup>University of Nebraska Medical Center and Truhlsen Eye Institute/Ophthalmology, Omaha, NE, USA, <sup>4</sup>University of Kentucky/Anatomy and Neurobiology Lexington, KY, USA, <sup>5</sup>University of Kentucky/Pathology and Laboratory Medicine Lexington, KY, USA

**Introduction:**

Very little is known about the anatomy and pathology of the optic nerve sheath (ONS) in normal and diseased states. The objective of our study is to explore microscopic changes in the ONS in patients with severe and/or progressive vision loss from IIH.

**Methods:**

ONS specimens were obtained from patients with IIH after ONS fenestration (cases) and painful blind eye after enucleation (controls). Both procedures were performed by a single surgeon. A 2 x 1 mm ONS window was obtained, divided into 2 parts, fixed and stained per following protocol. Light Microscopy (LM): All specimens were formalin fixed, stained using Hematoxylin-Eosin and examined at 10x and 40x magnification. Transmission electron microscopy (TEM): All specimens were immediately fixed (in Operating room) using 4% paraformaldehyde and 3.5% glutaraldehyde on ice; washed in Cacodylate buffer; postfixed using Osmium tetroxide/buffer mixture; dehydrated through graded concentrations of ethanol washes; and resin embedded. Ultrathin sections were obtained, stained and examined in Philips Tecnai Biotwin 12 TEM at 2900X magnification. Polarization microscopy (PM): All specimens were stained using Picrosirius red and examined under polarization microscope using previously published methods. (Junqueira et al., 1979) Masked experts in LM, TEM and PM performed evaluations for specimen quality (desiccation, trauma and staining) and tissue anatomy (cellularity and collagen arrangement).

**Results:**

Of 13 subjects and 16 eyes enrolled, 7 specimens (6 cases and 1 control) met inclusion and quality control criteria for analysis. Tissue was not obtained in 4 eyes at surgery. 2 specimens were excluded due to alternative diagnosis (meningioma, panophthalmitis). 3 specimens were excluded due to tissue desiccation resulting from increased time interval from resection to tissue fixation.

**Conclusions:**

Adequate quantity and quality ONS specimens were obtained in patients undergoing ONS fenestration and enucleation. Since ONS is susceptible to desiccation, tissue should be fixed immediately in the operating room.

**References:**

1. Junqueira LC, Bignolas G, Brentani RR. Picrosirius staining plus polarization microscopy, a specific method for collagen detection in tissue sections. *Histochem J* 1979; 11: 447-55.

**Keywords:** Optic Nerve Sheath, Pseudotumor Cerebri, High Intracranial Pressure/Headache, Orbit/Ocular Pathology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Research to prevent blindness.

**Ultramicroscopic study of the Optic Nerve Sheath in Patients with Severe Vision Loss from Idiopathic Intracranial Hypertension- Results**

Marla Davis<sup>1</sup>, Joshua W Evans<sup>1</sup>, Sachin Kedar<sup>2,3</sup>, Deepta Ghate<sup>3</sup>, Peter J Timoney<sup>1</sup>, Richard Kieler<sup>1</sup>, Bruce E Maley<sup>4</sup>, William O'Connor<sup>5</sup>

<sup>1</sup>University of Kentucky/Ophthalmology, Lexington, KY, USA, <sup>2</sup>University of Nebraska Medical Center/Neurology, Omaha, NE, USA, <sup>3</sup>University of Nebraska Medical Center and Truhlsen Eye Institute/Ophthalmology, Omaha, NE, USA, <sup>4</sup>University of Kentucky/Anatomy and Neurobiology Lexington, KY, USA, <sup>5</sup>University of Kentucky/Pathology and Laboratory Medicine Lexington, KY, USA

**Introduction:**

The objective of our study is to describe microscopic changes in the optic nerve sheath (ONS) of patients with severe and/or progressive vision loss from IIH.

**Methods:**

ONS specimens were obtained at ONS fenestration in IIH (cases) and enucleation for painful blind eye (controls). Both procedures were performed by a single surgeon. After fixation and staining, specimens were examined by masked experts using light (LM), transmission electron (TEM) and polarization (PM) microscopy for specimen quality and tissue anatomy (cellularity and collagen structure). (Details in accompanying abstract).

**Results:**

Of 12 specimens, 7 (6 cases, 1 control) met diagnostic and quality criteria. For cases: Mean age was 31.25y; mean disease duration 57 days; Mean CSF OP 40.25cm water. Papilledema grade s were II (3 eyes); III (1 eye); IV (2 eyes). 5/6 patients showed severe visual field loss (MD> 15 dB); 3/6 had acuity worse than 20/40 and 2/6 had RAPD. All had normal CSF contents. Control ONS was obtained from an enucleated eye of a 18y with childhood ocular trauma. Collagen abnormalities on TEM: irregular arrangement (all cases), collagen disruption and increased extracellular fluid (5/6 cases) while control ONS showed regular, compact, normal collagen. PM showed marked decrease in green yellow birefringence (<20% in all cases with <5% in 4 cases) compared to control ONS (75% birefringence) which confirmed abnormal collagen content and arrangement in cases. All 6 cases showed increased cellularity on LM and/or TEM with active fibroblasts (3/6 cases) and chronic inflammatory cells (lymphocytes) in all 6 cases.

**Conclusions:**

Disruption and disorganization of dural collagen in the ONS from IIH points to significant shear forces on distal ONS from raised ICP while increased cellularity indicates tissue repair. These mechanical and inflammatory components could contribute to visual loss in IIH. Early aggressive medical and/or surgical treatment may be beneficial in IIH.

**References:** None.

**Keywords:** Optic Nerve Sheath, Pseudotumor Cerebri, High Intracranial Pressure/Headache, Orbit/Ocular Pathology, Dura

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Research to prevent blindness

## Exploratory Study of the Relationship between the Levonorgestrel-Releasing Intrauterine System and Idiopathic Intracranial Hypertension

Reuben M Valenzuela<sup>1</sup>, Ruju Rai<sup>1</sup>, Brian H Kirk<sup>1</sup>, Subhashree Sundar<sup>1</sup>, Jessica N Sanders<sup>2</sup>, Judith E A Warner<sup>1</sup>, Kathleen B Digre<sup>1</sup>, Alison V Crum<sup>1</sup>, Bradley J Katz<sup>1</sup>

<sup>1</sup>University of Utah/John A Moran Eye Center, Salt Lake City, UT, USA, <sup>2</sup>University of Utah/Obstetrics and Gynecology, Salt Lake City, UT, USA

### Introduction:

Unconfirmed reports have linked the levonorgestrel-releasing intrauterine system (LNG-IUS aka Mirena), a long-acting contraceptive, to idiopathic intracranial hypertension (IIH). In this pilot study, we assessed signs and symptoms of IIH among a cohort of patients using LNG-IUS.

### Methods:

A series of 473 IIH patients seen between 2008 and 2013 was screened for female gender, age  $\geq 18$ , and absence of causative etiologies. Of 176 eligible participants, 59 completed birth control histories by telephone.

### Results:

Eight of these 59 patients (14%) were using an LNG-IUS within 3 months of onset of IIH symptoms. Nine patients (15%) were on another contraceptive, and 42 (71%) were not using any contraceptives. All LNG-IUS users developed symptoms while the device was *in situ*. There were no significant differences between LNG-IUS users and non-users in terms of age ( $30 \pm 6$  vs  $32 \pm 9$ ,  $p=0.62$ ), body mass index ( $35 \pm 9$  vs  $35 \pm 7$ ,  $p=0.92$ ), history of recent weight gain (50% vs 25%,  $p=0.16$ ) or presenting symptoms. Signs were also similar between the two groups: opening pressure ( $357 \pm 101$  vs  $404 \pm 13$ ,  $p=0.25$ ), Frisén papilledema grade (stage 2 most frequent, worse and fellow eye,  $p=0.09$ ), perimetric mean deviation ( $-5.6 \pm 5.5$  vs  $-6.2 \pm 7.5$  worse eye,  $p=0.83$ ;  $-2.9 \pm 3.1$  vs  $-4.1 \pm 5.4$  fellow eye,  $p=0.54$ ;  $-2.7 \pm 2.7$  vs  $-1.8 \pm 3.0$  difference between eyes,  $p=0.38$ ), and LogMAR best corrected visual acuity ( $0.06 \pm 0.11$  vs  $0.09 \pm 0.30$  worse eye,  $p=0.81$ ;  $-0.01 \pm 0.10$  vs  $0.05 \pm 0.29$  fellow eye,  $p=0.56$ ;  $0.08 \pm 0.09$  vs  $0.04 \pm 0.11$  difference between eyes,  $p=0.42$ ).

### Conclusions:

Our preliminary findings suggest that IIH patients using an LNG-IUS have clinical features comparable to those not using an LNG-IUS. We are currently investigating the incidence of IIH among users of LNG-IUS and will present these data at the Annual Meeting. Although a causative role for LNG-IUS has not been established, until further data are collected, we recommend augmenting the routine evaluation for IIH with a birth control history.

**References:** None.

**Keywords:** Pseudotumor Cerebri, High Intracranial Pressure/Headache, Levonogestrel, Contraception, IUD

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Supported in part by grants from the Center for Clinical and Translational Sciences (8UL1TR000105) and the Research to Prevent Blindness, Inc., New York, NY.



## Coning Following Lumbar Puncture In A Patient With Idiopathic Intracranial Hypertension

Christian J Lueck<sup>1,2</sup>, Adeniyi Borire<sup>1,2</sup>, Andrew Hughes<sup>1,2</sup>

<sup>1</sup>Department of Neurology, The Canberra Hospital, Canberra, Australia, <sup>2</sup>Australian National University Medical School, Canberra, Australia

### Introduction:

Lumbar puncture is performed frequently in patients with idiopathic intracranial hypertension. Cerebellar tonsillar descent is not uncommon, but frank herniation (coning) is extremely rare.<sup>1</sup> We are aware of only one previous case reported in the literature.<sup>2</sup> We describe a case in which the patient coned following a lumbar puncture, necessitating acute neurosurgical intervention.

### Methods:

Case Report

### Results:

A 30 year old obese Caucasian woman with well-controlled idiopathic intracranial hypertension (IIH) presented to the emergency department with a week's history of severe bifrontal headache, associated with nausea, photophobia and neck pain. Neurologic examination was unremarkable apart from bilateral papilloedema. She had had three therapeutic lumbar punctures (LP) previously, all of which showed high opening pressures and non-inflammatory CSF. Her brain MRI at initial diagnosis was in keeping with IIH. She had also had a CT venogram which showed hypoplasia of the entire left venous system and was waiting for insertion of a venous stent at another hospital. A therapeutic lumbar puncture was performed under fluoroscopic guidance. Her CSF opening pressure was 47 cmH<sub>2</sub>O and 20ml of CSF was removed. 12 hours later, she developed severe (grade 10/10) global headache with excruciating neck pain. She was unable to lie flat and held her neck in extreme extension. This was followed 12 hours later by hypotension, reduced level of consciousness, and anisocoria. A CT brain showed tonsillar herniation. She was treated by external ventricular drain and then bifrontal craniectomy.

### Conclusions:

This patient also had renal tubular acidosis and she was quite acidotic at the time of presentation. It is speculated that this may have been related to the pathophysiology of the coning, explaining why she suffered this unusual complication of lumbar puncture.

### References:

1. Paruchuri SRA, Lawlor M, Kleinhomer K, Mason L, Johnson C. Risk of cerebellar tonsillar herniation after diagnostic lumbar puncture pseudotumor cerebri. *Anesth Analg* 77, 398-410, 1993
2. Sullivan HC. Fatal tonsillar herniation in pseudotumor cerebri. *Neurology* 41, 1142-4, 1991

**Keywords:** Idiopathic Intracranial Hypertension, Cerebellar Tonsillar Herniation, Coning, Renal Tubular Acidosis

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 104

### Long term follow-up of PTC: pre-pubertal children Vs adolescents and adults

Assaf Hilely<sup>1</sup>, Nitza Goldenberg Cohen<sup>2</sup>, Hana Leiba<sup>1,3</sup>

<sup>1</sup>Kaplan Medical Center, Rehovot, Israel, <sup>2</sup>Shneider Medical Center, Rama Gan, Israel, <sup>3</sup>The Hebrew University, Jerusalem, Israel

#### Introduction:

Pseudotumor-cerebri syndrome (PTCS) can produce a prolonged or recurrent course. Data in the literature regarding the long term results of the disease is limited. The aim of this study was to evaluate the long term results of the disease course, recurrence rates, and final visual outcome of PTCS in pre pubertal children and adolescents and adults strictly diagnosed for PTCS

#### Methods:

A retrospective observational study.

Included were patients with at least 5 years of follow-up. Excluded were patients with any systemic disease that can cause elevated intracranial pressure. Patients were divided into pre-pubertal children (group A) and all the others (group B).

Main outcome measures were final visual outcome and recurrences

#### Results:

included were 61 patients (46 females, 15 males). 16 were pre-pubertal children (9 girls 7 boys). Mean age at diagnosis was 9.9 years (5-13) for group A and 28 for group B. Mean follow-up time was 8.9 years for both groups (A: 5-11; B: 5-32) Mean duration of treatment was 2.5 years for both groups. Overall 40% had recurrent event; 9/16 (56 %) in group A and 14/45 (31 %) in group B. Recurrent attack occurred within 0.8 years from treatment cessation in group A and 2.8 years in group B. 9 patients needed surgical intervention (2 group A, 7 group B), mean in 3 years (3 months - 7 years) after initial diagnosis. Optic neuropathy in at least one eye was seen in 28/61 (60%) [10/16 (62%) group A, 18/45 (40%) group B]

#### Conclusions:

in spite of the limitation of a small retrospective cohort, we confirmed recurrence of PTC events to occurs years after remission, thus, revealing the importance of longer follow up. Prepubertal children tend to experience recurrences earlier than adults; but develop optic neuropathy similar to adolescents and adults

**References:** None.

**Keywords:** PTC, Long Term, Prepuberty, Children, Outcome

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Hemodialysis Graft-Induced Intracranial Hypertension

Devin D. Mackay<sup>1</sup>, Valerie Biousse<sup>2</sup>

<sup>1</sup>Indiana University, Departments of Neurology, Ophthalmology, and Neurosurgery, Indianapolis, IN, USA, <sup>2</sup>Emory University, Departments of Ophthalmology and Neurology, Atlanta, GA, USA

### Introduction:

Intracranial hypertension (IH) is rarely associated with peripheral hemodialysis shunts, presumably in association with central venous stenosis.<sup>1,2</sup> Hemodialysis Reliable Outflow (HeRO™) grafts are designed to bypass pre-existing central venous stenosis by connecting the brachial artery with the venous circulation through the ipsilateral internal jugular (IJ) vein.<sup>3</sup> We report a case of IH immediately after placement of a HeRO graft and discuss possible pathophysiology.

### Methods:

Case report with review of the English literature.

### Results:

A 60-year-old woman with a nonfunctioning right arm hemodialysis arteriovenous (AV) shunt developed blurred vision and optic disc edema three days following placement of a left-sided HeRO graft. Brain MRI/MRV showed signs of IH and lumbar puncture CSF opening pressure was 38 cm H<sub>2</sub>O. There was 60-70% focal stenosis of a right subclavian vein stent overlying the ostium of the right internal jugular vein and a HeRO graft entering the left IJ vein, resulting in superior vena cava syndrome with IH and papilledema. A ventriculoperitoneal shunt was placed, followed by papilledema resolution and vision improvement. At least 12 cases of presumed IH from central venous stenosis and hemodialysis AV shunts have been published: none involving a HeRO graft. All cases involved brachiocephalic vein stenosis/occlusion (10/12) or IJ vein occlusion (2/12). All were treated with venoplasty +/- AV shunt ligation, and 1/12 was additionally treated with a lumboperitoneal shunt.

### Conclusions:

Despite increasing venous flow in patients with central venous stenosis, IH has not been reported in association with HeRO grafts, suggesting the contribution of additional factors to the pathogenesis of hemodialysis graft-induced IH. Although usually treated by graft ligation or venoplasty +/- stent, CSF shunting without graft ligation is also a treatment option in patients with isolated IH whose venous anatomy is seriously compromised. Imaging of chest/neck veins in hemodialysis patients with unexplained IH should be considered in cases with previous AV shunts.

### References:

1. Taban M, Taban M, Lee MS, Smith SD, Heyka R, et al. Prevalence of optic nerve edema in patients on peripheral hemodialysis. *Ophthalmology* 114:1580-1583, 2007.
2. Simon MA, Duffis EJ, Curi MA, Turbin RE, Prestigiacomo CJ, et al. Papilledema Due to a Permanent Catheter for Renal Dialysis and an Arteriovenous Fistula: A "Two Hit" Hypothesis. *J Neuro-Ophthalmol* 34:29-33, 2014.
3. Katzman HE, McLafferty RB, Ross JR, Glickman MH, Peden EK, et al. Initial experience and outcome of a new hemodialysis access device for catheter-dependent patients. *J Vasc Surg* 50:600-607, 2009.

**Keywords:** High Intracranial Pressure, Pseudotumor Cerebri, Vascular Disorders, Neuroimaging

**Financial Disclosures:** This work was supported in part by an unrestricted departmental grant (Department of Ophthalmology, Emory University) from Research to Prevent Blindness, Inc., New York.

**Grant Support:** This work was supported in part by an unrestricted departmental grant (Department of Ophthalmology, Emory University) from Research to Prevent Blindness, Inc., New York.

## Incidence of Idiopathic Intracranial Hypertension (IIH) Among Users of Tetracycline Antibiotics

Samuel F Passi<sup>1</sup>, Ryan Butcher<sup>2</sup>, Judith Warner<sup>1</sup>, Bradley Katz<sup>1</sup>, Alison Crum<sup>1</sup>, Ramkiran Gouripeddi<sup>2</sup>, Kathleen Digre<sup>1</sup>

<sup>1</sup>John A. Moran Eye Center, University of Utah, Salt Lake City, UT, USA, <sup>2</sup>Biomedical Informatics Core, Center for Clinical and Translation Sciences, Salt Lake City, UT, USA

### Introduction:

Previous studies have shown an association between the use of tetracycline antibiotics and the development of IIH. We sought to calculate the incidence of tetracycline-associated IIH in the University of Utah patient population.

### Methods:

Using the newly completed REDCap IIH database at the University of Utah Moran Eye Center we identified 45 patients diagnosed with IIH secondary to tetracycline use during the period 2007-2013. Utilizing the FURTHeR database, we then queried the University of Utah Enterprise Data Warehouse (a database of all individuals seen at University of Utah hospitals and clinics) for both men and women between the ages of 12 and 50 who were ever prescribed a tetracycline antibiotic (doxycycline, minocycline, tetracycline, tigecycline, demeclocycline) over the same time period.

### Results:

We found that 960 patients between the ages of 12 and 50 were prescribed a tetracycline antibiotic from 2007-2013 out of a total of 876,358 patients in the database with the same age and year restrictions. Dividing our 45 patients with IIH secondary to tetracycline use by the population at risk (960) and seven patient-years of patient-study and multiplying our result by 100,000 years, we calculated the incidence of tetracycline associated IIH to be 670 per 100,000 patient-years of study.

### Conclusions:

This investigation provides the first calculation of the incidence of IIH among patients taking this class of antibiotics. The incidence of IIH among users of tetracycline antibiotics is greater than the incidence of IIH in the general population. Although these data do not provide a causal link between IIH and antibiotics, this observation further substantiates the hypothesis that tetracycline antibiotics may cause IIH in susceptible individuals.

**References:** None.

**Keywords:** Idiopathic Intracranial Hypertension, Pseudotumor Cerebri, Tetracycline, Antibiotics, Incidence

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## CSF Characteristics in Patients with High Frisén Papilledema Grades from the IIHTT

John H. Pula<sup>1</sup>, Jorge C. Kattah<sup>2</sup>, Luis J. Mejico<sup>3</sup>, Michael P. McDermott<sup>4</sup>, Michael Wall<sup>5</sup>

<sup>1</sup>Northshore University Healthsystem, Evanston, IL, USA, <sup>2</sup>University of Illinois College of Medicine in Peoria, Peoria, IL, USA, <sup>3</sup>SUNY Upstate Medical University, Syracuse, NY, USA, <sup>4</sup>University of Rochester Rochester, NY, USA, <sup>5</sup>University of Iowa Iowa City, IA, USA

### Introduction:

Papilledema is optic disc edema due to increased intracranial pressure. The severity of papilledema grade may be related to the density of trabeculations in the subarachnoid space, magnitude of CSF pressure, or another mechanism. We investigated the relationship of CSF pressure to high Frisén papilledema grades (4 and 5), and the response to randomized treatment with acetazolamide plus diet versus placebo plus diet in the Idiopathic Intracranial Hypertension Treatment Trial (IIHTT).

### Methods:

Papilledema was present and graded in all IIHTT subjects, each of whom had mild to moderate visual field loss. In the IIHTT, 165 patients met inclusion criteria with lumbar puncture (LP) CSFp > 250 mm H<sub>2</sub>O, or > 200 mm H<sub>2</sub>O with other compelling IIH clinical/imaging criteria. All participants were asked to repeat the LP 6 months later. We correlated CSF pressure (CSFp) with Frisén grade and treatment category.

### Results:

Fifty patients had high Frisén grade (HFG). Of these, 21 had CSFp <350 and 29 had CSFp > 350. HFG was found in 22% of patients having CSFp <350, and 39% of patients having CSFp >350 ( $p = 0.0262$ ). 6 months LP was performed in 13/29 patients having both CSFp > 350 and HFG. In this group, eleven patients were on acetazolamide. All of these improved: Seven by Frisén grade of 0-1, one by 2, and three by 3. Mean CSFp decreased from 446.5 to 233.5. In contrast, the two patients in this category on diet alone sustained persistently high CSFp and unchanged HFG. The one patient who required optic nerve sheath fenestration was on diet alone.

### Conclusions:

Higher CSFp was associated with significantly higher (worse) Frisén grade. Treatment with acetazolamide improved patients with HFG.

**References:** None.

**Keywords:** High Intracranial Pressure/Headache, Pseudotumor Cerebri

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Visual Outcomes After Treatment of Venous Sinus Stenosis with Dural Venous Sinus Stenting

Ahmara G Ross<sup>1</sup>, Andrew Carlson<sup>2</sup>, Timothy Winter<sup>3</sup>

<sup>1</sup>UPMC/ University of Pittsburgh, Pittsburgh, PA, USA, <sup>2</sup>University of New Mexico Department of Surgery, Division of Ophthalmology, Albuquerque, NM, USA, <sup>3</sup>University of New Mexico Department of Neurosurgery, Albuquerque, NM, USA

### Introduction:

Dural venous sinus stenting (DVSS) has been utilized for years to treat Idiopathic Intracranial Hypertension (IIH), but the literature is scant illustrating its utility in venous sinus stenosis. In most clinical studies that involve this technique, there is an overwhelming group of patients with IIH with few that have known venous sinus abnormalities causing visual symptoms at the time of diagnosis. Although DVSS has been found to be safe and effective at lowering intracranial pressure, this study reviews the effect on the visual system after DVSS with or without addition of acetazolamide.

### Methods:

Two patients, ages 19 and 35, with an initial diagnosis of venous sinus stenosis will be reviewed. Visual acuity, color vision, visual fields, alignment, and optic nerve abnormalities were noted preoperatively, immediately postoperatively and during extended follow-up.

### Results:

Initial visual acuity ranged from 20/20 to LP and MD were (x) on visual field testing prior to surgery. At long-term follow-up, average visual acuities ranged from 20/20 to 20/50 and one patient improved by visual field testing. Optic nerve edema recorded pre- and post-operatively were found to have improved immediately after DVSS. One patient needed angioplasty for the thrombosis before DVSS, as well as evaluation for coagulopathy; this second patient required additional treatment with acetazolamide to improve visual acuity and reduce optic disc edema noted on examination and on subsequent optical coherence tomography.

### Conclusions:

There are few studies evaluating the use of dural venous sinus stenting to treat venous sinus stenosis. While medical management and close targeted monitoring remain a cornerstone of management, this technique may become another surgical option to treat venous sinus stenosis. Additional medical and hematologic management may be required to prevent additional vision threatening disease.

**References:** None.

**Keywords:** Venous Sinus Thrombosis, Dural Venous Sinus Stenting

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 109

### Relationship between High Opening Pressure on Lumbar Puncture and Failure of Optic Nerve Sheath Decompression to Prevent Progressive Visual Loss in Patients with Idiopathic Intracranial Hypertension.

Mark E. Robinson<sup>1</sup>, John Pagteilan<sup>2</sup>, Ryan O'Myeilia<sup>2</sup>, Annie Moreau<sup>1</sup>, Bradley K. Farris<sup>1</sup>

<sup>1</sup>Dean McGee Eye Institute, Oklahoma City, OK, USA, <sup>2</sup>College of Medicine, Oklahoma University Health Sciences Center, Oklahoma City, OK, USA

#### Introduction:

To our knowledge, there have been no prior published studies that address the relationship between high opening pressures and progression of visual loss despite optic nerve sheath decompression (ONSD) in patients with idiopathic intracranial hypertension (IIH). Past experience suggests that very high opening pressures (50cm water) place patients at higher risk for ONSD failure. If this hypothesis is confirmed, this knowledge will aid in decision making regarding when it is appropriate to proceed with an ONSD versus a lumbo-peritoneal (LP) or ventriculo-peritoneal (VP) shunt as the initial procedure in patients with IIH who have opening pressures of 50 or greater.

#### Methods:

Retrospective chart review of patients with IIH who underwent ONSD between January, 1992 and January, 2014. The primary question will compare the failure of ONSD to prevent progressive visual loss in patients with opening pressures less than 50, to those with opening pressures of 50 or greater. Also, if the data allow, we will construct a graph based on categories of opening pressure (e.g. 35-40, 41-45, 46-50, etc.) and their relationship to the failure rate. The definition of failure of the ONSD to protect vision will be the progression of visual loss or persistence of papilledema, leading to a VP or LP shunt within 3 months of the ONSD. Inclusion criteria will be diagnosis of idiopathic intracranial hypertension, recorded opening pressure on lumbar puncture, and papilledema on fundus examination. Exclusion criteria will be a diagnosis other than IIH, prior VP or LP shunt procedure, lack of a recorded opening pressure on lumbar puncture, or subsequent VP or LP shunt to treat uncontrolled headache rather than vision loss.

#### Results:

Pending.

#### Conclusions:

Pending.

**References:** None.

**Keywords:** Pseudotumor Cerebri, Optic Neuropathy, High Intracranial Pressure/Headache

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Venous Sinus Stenting For Treatment Of Increased Intracranial Pressure Secondary To Venous Sinus Stenosis

Tarek A Shazly<sup>1</sup>, Nikisha Richards<sup>1</sup>, Ashutosh Jadhav<sup>2</sup>, Tudor Jovin<sup>2</sup>, Gabrielle R Bonhomme<sup>1</sup>

<sup>1</sup>University of Pittsburgh, Department of Ophthalmology, Pittsburgh, PA, USA, <sup>2</sup>University of Pittsburgh, Department of neurology, Pittsburgh, PA, USA

### Introduction:

Medical treatment, CSF shunting and optic nerve sheath fenestration are the standard treatments for increased Intracranial Pressure (ICP). Venous sinus obstruction is a well-known cause of secondary elevation of ICP. Venous sinus stenting provides a novel, alternative surgical treatment in cases of venous sinus stenosis.

### Methods:

Twelve consecutive patients with papilledema, increased ICP and MRV signs of dural sinus narrowing underwent direct retrograde cerebral venography and manometry. All patients demonstrated radiological signs of obstruction of the venous lateral sinuses. The CSF opening pressure was measured via lumbar puncture in all patients prior to venography. Funduscopy examination demonstrated papilledema for all patients. Venous stenting was offered to all patients.

### Results:

Six patients opted to undergo venous stenting (Group-A) and 6 were managed conservatively with acetazolamide and monitoring (Group-B). Pressure gradients were measured across the venous stenosis, and was 20 mm Hg in Group-A and 13 mm HG in Group-B ( $p=0.11$ ). The CSF pressure on lumbar puncture ranged from 26.5 to 55 CmH<sub>2</sub>O (mean 40.6 +/- 11.2 CmH<sub>2</sub>O) with bland composition. Mean ICP for Group-A was 48.4mmHg, while it was 35.3mmHg in Group-B. Average binocular retinal nerve fiber layer thickness (RNFL) at presentation was 310 microns in Group-A and 210 in Group-B ( $p=0.002$ ). However, binocular RNFL at 3 months was 80.6 microns in Group-A and 124.5 in Group-B ( $p=0.08$ ). Average acetazolamide dosage at 2-4 weeks was 1200mg/day in Group-A and 1083mg/day in Group-B ( $p=0.9$ ). However, at 6 months, the mean acetazolamide dosage was 900mg/day in Group-A and 1666mg/day in Group-B ( $p=0.19$ ). Papilledema resolved in all patients at 3-6 months.

### Conclusions:

Patients with increased ICP and evidence of dural sinus narrowing on MRV should be evaluated with direct cerebral venography and manometry. In patients with venous sinus stenosis endovascular stenting seems to be a successful treatment with a trend towards quicker resolution of papilledema and less need for acetazolamide at 2-4 weeks when compared to conservative treatment.

### References:

1. Higgins, J. N. P., et al. "Idiopathic intracranial hypertension: 12 cases treated by venous sinus stenting." *Journal of Neurology, Neurosurgery & Psychiatry* 74.12 (2003): 1662-1666.
2. Higgins, J. Nicholas P., et al. "Venous sinus stenting for refractory benign intracranial hypertension." *The Lancet* 359.9302 (2002): 228-230.
3. Friedman, Deborah I. "Cerebral venous pressure, intra-abdominal pressure, and dural venous sinus stenting in idiopathic intracranial hypertension." *Journal of neuro-ophthalmology* 26.1 (2006): 61-64.
4. Rajpal, Sharad, David B. Niemann, and Aquilla S. Turk. "Transverse venous sinus stent placement as treatment for benign intracranial hypertension in a young male: case report and review of the literature." *Journal of Neurosurgery: Pediatrics* 102.3 (2005): 342-346.
5. Arac, Ahmet, et al. "Efficacy of endovascular stenting in dural venous sinus stenosis for the treatment of idiopathic intracranial hypertension." *Neurosurgical focus* 27.5 (2009): E14.

**Keywords:** Intracranial Pressure, Dural Sinus, Stenting, Papilledema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



**Pediatric Primary Pseudotumor Cerebri Syndrome (PTCS): A Detailed, Retrospective, Multicenter Analysis Of Anthropometrics**

Claire A. Sheldon<sup>1,2</sup>, Grace L. Paley<sup>1,9</sup>, Anat Kesler<sup>3</sup>, Ori Eyal<sup>4</sup>, Chantal Boisvert<sup>5</sup>, Robert A. Avery<sup>6</sup>, Gena Heidary<sup>7</sup>, Melissa W. Ko<sup>8</sup>, Shana E. McCormack<sup>9,10</sup>, Grant T. Liu<sup>1,2,9</sup>

<sup>1</sup>Departments of Neurology and Ophthalmology, Hospital of the University of Pennsylvania, Philadelphia, PA, USA, <sup>2</sup>Division of Ophthalmology, Children's Hospital of Philadelphia, Philadelphia, PA, USA, <sup>3</sup>Department of Ophthalmology, Tel- Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel, <sup>4</sup>Pediatric Endocrinology and Diabetes Unit, Dana-Dwek Children's Hospital, Tel- Aviv Sourasky Medical Center Sackler Faculty of Medicine, Tel Aviv University Tel Aviv, Israel, <sup>5</sup>Department of Ophthalmology, Rady Children's Hospital San Diego, CA, USA, <sup>6</sup>Departments of Neurology, Ophthalmology and Pediatrics, Children's National Medical Center Washington, DC, USA, <sup>7</sup>Department of Ophthalmology, Boston Children's Hospital Boston, MA, USA, <sup>8</sup>Perelman School of Medicine at the University of Pennsylvania Philadelphia, PA, USA, <sup>9</sup>Division of Endocrinology and Diabetes, Children's Hospital of Philadelphia Philadelphia, PA, USA

**Introduction:**

Obesity, gender, and pubertal status influence the risk for pediatric PTCS. Preliminary studies have identified three subtypes of pediatric PTCS, based on anthropometrics. In an international, collaborative effort, this study was designed to refine this observation further.

**Methods:**

This multi-site study included 5 tertiary pediatric centers. Cases of PTCS were identified retrospectively, based on ICD9 code 348.2 and/or pediatric neuro-ophthalmologist database. Diagnosis of definite and probable PTCS was defined by applying updated PTCS diagnostic criteria (2013). Anthropometric data were abstracted and standardized growth charts determined age-/sex-specific height, weight, and BMI Z-scores. Where available, Tanner staging documented development of secondary sexual characteristics.

**Results:**

162 definite (n=155) or probable (n=7) cases of primary PTCS were identified (64% female, mean age 11.9±4.4yrs). There was a positive association between BMI Z-score and age ( $r=0.48$ ,  $p<0.001$ ;  $n=162$ ), that, using linear regression analysis, crossed the 'overweight threshold' (BMI Z-score $\geq 1.04$ ) at 9.4yrs and 'obese threshold' (BMI Z-score $\geq 1.64$ ) at 14yrs. In children younger than 9.4yrs, no significant association was observed between height Z-score and age ( $r=0.18$ ,  $p=0.21$ ,  $n=48$ ). There was a negative relationship between height Z-score and age in children  $\geq 9.4$ yrs ( $r=-0.31$ ,  $p=0.0008$ ,  $n=113$ ), crossing the x-intercept at 15.6yrs. Data on Tanner staging or menarchal status was available in 38 cases. Before 9.4yrs, 4/5 girls were pre-pubertal. After 14yrs, 17/17 girls were post-menarchal. Limited data were available for intermediate ages and boys.

**Conclusions:**

Before age 9.4 years, pre-pubertal children with PTCS tend to be normal weight. After age 9.4 years, early adolescents with PTCS are typically overweight, and taller for age and sex than peers, suggesting linear growth acceleration. Older, obese, late- or post-pubertal adolescents with PTCS are normal height. Defining key pubertal events that influence the development of pediatric PTCS may guide an understanding of endocrine 'triggers' and, in turn, the pathogenesis of this complex condition.

**References:** None.

**Keywords:** Pediatric Pseudotumor Cerebri Syndrome, Anthropometrics, Obesity, Pubertal status

**Financial Disclosures:** Grant T Liu, M.D. - acknowledges previous consultancy for Ipsen Shana E McCormack, M.D.- K12 grant support from NIH, 5K12DK094723-03 (S.E.M., PI: S. Willi)

**Grant Support:** Shana E McCormack, M.D.- K12 grant support from NIH, 5K12DK094723-03 (S.E.M., PI: S. Willi)

## Poster 112

### Endovascular Venous Stenting In Treatment Of Primary Idiopathic Intracranial Hypertension

Rajeev Sivasankar<sup>1</sup>, Hemant Trehan<sup>2</sup>, Rochan Pant<sup>1</sup>

<sup>1</sup>Department of Imaging & Interventional Radiology, INHS Asvini, Colaba, Mumbai, India, <sup>2</sup>Department of Ophthalmology, INHS Asvini, Colaba, Mumbai, India

#### Introduction:

Idiopathic Intracranial Hypertension (IIH) is a syndrome of elevated intracranial pressure that can be primary or secondary. In its primary form, it is usually a diagnosis of exclusion. Current treatment options both medical and surgical have varying success rates. We wish to present a small series of patients wherein a select group of patients presenting with primary IIH were treated with endovascular venous stenting.

#### Methods:

Patients with clinical features of headaches and visual blurring and/or acuity loss underwent detailed ophthalmological evaluation between Jan 2013 and Aug 2014. Those with local causes were excluded and the rest underwent contrast MRI and MR venograms. Those without secondary cause of raised ICP were evaluated for established imaging markers of primary IIH. These patients were managed medically with Acetazolamide and targeted weight loss. Patients not responding to medical therapy underwent cerebral angiography & direct retrograde cerebral venous pressure manometry (DRCVM). Patients with pressure gradients greater than 8 mm Hg underwent venous stenting. Detailed clinical and ophthalmological evaluation was performed 1,3 and 6 months post procedure.

#### Results:

2121 patients were evaluated prospectively for headaches and visual symptoms. 74 patients had no local cause and underwent contrast enhanced MRI and MRV. 61 patients had secondary causes of raised ICP. 13 patients were medically treated. 3 of these patients showed good clinical response. 2 patients refused endovascular treatment. 8 patients underwent angiography, DRCVM and venous stenting. In the endovascular group there was 100% technical success. 7 of the 8 patients (87.5%) were stented unilaterally and 1 patient (12.5%) bilaterally. Follow up showed resolution of headaches in 7 out of 8 patients (87.5%). Papilloedema resolved in all 8 patients. Visual acuity improvement was in 6 out of 8 (75%) patients.

#### Conclusions:

Endovenous stenting is a safe, easy and effective method of treating patients with primary IIH.

**References:** None.

**Keywords:** Pseudotumor Cerebri, Interventional Neuroradiology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 113

### Idiopathic Intracranial Hypertension Mimicking Foster Kennedy Syndrome

Chuan-bin Sun<sup>1</sup>, Shi-hui Wei<sup>2</sup>

<sup>1</sup>Eye Center, Second Affiliated Hospital of Zhejiang University School of Medicine, Hangzhou, China, <sup>2</sup>General PLA Hospital, Beijing, China

#### Introduction:

To report a case of pseudo- Foster Kennedy Syndrome due to idiopathic intracranial hypertension (IIH).

#### Methods:

A case report.

#### Results:

A 56-year-old man complained of transiently blurred vision in the right eye for 1 month. The patient denied diplopia, headache, or vomiting. His past medical history was unremarkable, neither previous trauma, nor any medication including vitamin A was recorded. Ophthalmic examination revealed a best corrected visual acuity of 20/20 and 20/400 in the right and left eye, respectively. His anterior segment was normal except a relative afferent pupillary defect in the left eye, fundus examination revealed grade II optic disc edema in the right eye and a pale disc in the left eye, the macula and peripheral retina was normal in both eyes. The intraocular pressures was normal in both eyes. Visual field test revealed normal in the right eye and diffuse depression in the left eye. Fundus fluorescein angiography showed late hyper fluorescence of optic disc in the right eye and hypofluorescence of optic disc in the left eye. Cranial MRI and MR venogram was both normal. A lumbar puncture revealed cerebrospinal fluid (CSF) pressure of 40 cm water. CSF analysis including cell counts, protein, and glucose were both within normal limits.

#### Conclusions:

pseudo- Foster Kennedy Syndrome is a rare appearance of IIH.

**References:** None.

**Keywords:** Idiopathic Intracranial Hypertension, Pseudo- Foster Kennedy Syndrome, MRI, Cerebrospinal Fluid, Visual Field

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 114

### Going For The Jugular! Paragangliomas, Papilledema, and Vision Loss

Jonathan Trobe<sup>1</sup>, Paweena Lertakyamanee, Ashok Srinivasan, Lindsey Delott

*University of Michigan/Departments of Ophthalmology, Neurology, and Radiology,, Ann Arbor, MI, USA*

#### **Introduction:**

Paragangliomas are relatively benign neuroendocrine tumors that often arise from the jugular bulb. They are known to present as masses in the neck or with hearing loss, pulsatile tinnitus, and manifestations of lower cranial nerve palsies. Much less recognized is their tendency to cause increased intracranial pressure and papilledema by obstructing jugular venous outflow. Only 7 such cases have been reported, and with minimal ophthalmic documentation.

#### **Methods:**

We describe 3 cases examined at a single academic medical center.

#### **Results:**

Case 1 had papilledema detected on routine optometric examination in a visually asymptomatic patient who had mild visual field loss. Imaging of the brain was normal, but an astute radiologist detected a left neck mass. No intervention took place and findings remained unchanged after 6 months. Case 2 presented with transient obscurations of vision (TOVs) and had papilledema that led to a head CT that disclosed bilateral neck masses. She received external beam radiation therapy that slightly reduced the size of the masses, but she developed optic disc pallor and persistent visual field loss. Case 3 with bilateral paragangliomas developed TOVs after surgical extirpation of a right neck paraganglioma and obliteration of the right jugular vein. Acetazolamide treatment stemmed the TOVs but mild optic neuropathy persisted.

#### **Conclusions:**

These 3 cases provide more ample evidence that jugular paragangliomas may be an overlooked cause of the pseudotumor cerebri syndrome, and that papilledema may not be recognized when a jugular paraganglioma is diagnosed or after it has been treated. Such lapses have led to disabling vision loss from damage to the optic nerves in longstanding papilledema.

**References:** None.

**Keywords:** Paraganglioma, Papilledema, Jugular

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 115

### Visual Outcomes Following Optic Nerve Sheath Fenestration

Neel S Vaidya<sup>1,2</sup>, Steven A Katz<sup>1</sup>

<sup>1</sup>Havener Eye Institute at The Ohio State University Wexner Medical Center, Columbus, OH, USA, <sup>2</sup>The Ohio State University College of Medicine, Columbus, OH, USA

#### Introduction:

Idiopathic intracranial hypertension (or pseudotumor cerebri) is a condition of increased intracranial pressure in the absence of vascular lesion or intracranial mass<sup>1</sup>. Symptoms of IIH include headache, pulsatile tinnitus, transient visual obscurations, and diplopia. Untreated IIH can present with the long-term sequela of visual loss<sup>2</sup>. In approximately 10% of patients with untreated IIH, visual loss will progress to the point of meeting the legal criteria for blindness<sup>3</sup>. The objective of this study was to determine the safety of optic nerve sheath fenestration (ONSF) for the treatment of patients with intracranial hypertension in the immediate 6 month post-operative period and its efficacy in reducing optic disk edema.

#### Methods:

Design: Retrospective, non-comparative interventional case series  
Participants: Two hundred and seven eyes in one hundred and four patients undergoing ONSF between the years 2005 and 2014  
Outcome Measures: Papilledema grade based on modified Frisén scale and mean deviation of Humphrey visual field

#### Results:

207 eyes of 104 patients (101 IIH, 2 IH due to dural sinus thrombosis) were included in the study. The patients were 96.1% female (N = 100) and 3.9% male (N = 4). The average patient age was 28.8 years (SD +/- 9.5 years) and had a mean opening pressure of 39.85 cmH<sub>2</sub>O (SD +/- 8.4 cmH<sub>2</sub>O). Mean follow-up period was 6.0 months (SD +/- 5.9 months). Papilledema resolved in 76.1% of eyes at 1 week (N = 102), 75% of eyes at 1 month (N = 90), and 71% of eyes at 6 months (N = 94). Visual field comparison had a mean of the paired differences at 1 week, 1 month, and 6 months of 1.59dB (p <0.01), 2.53dB (p <0.01), and 1.30dB (p = 0.016), respectively.

#### Conclusions:

ONSF is effective in reducing optic disk edema and does not cause vision loss in the 6-month post-operative period regardless of severity of IIH (as judged by elevation of opening pressure measured at pre-operative assessment).

#### References:

1. Dandy WE. Intracranial pressure without brain tumor: diagnosis and treatment, *Ann Surg*, 106, 492-513, 1937
2. Radhakrishnan K, Ahlsoeg JE, Cross SA, et al, Idiopathic intracranial hypertension (pseudotumor cerebri): Descriptive epidemiology in Rochester, Minn 1976-1990, *Arch Neurol*, 50, 78-80, 1993
3. Corbett JJ, Nerad JA, Tse DT, Anderson RL, Results of Optic Nerve Sheath Fenestration for Pseudotumor Cerebri: The Lateral Orbitotomy Approach. *Arch Ophthalmol*, 106, 1391-1397, 1988

**Keywords:** Pseudotumor Cerebri, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 116

### New Side Effect of Acetazolamide: Palinopsia

Peggy H. Vogt<sup>1</sup>, Charles G. Maitland<sup>1, 2</sup>

<sup>1</sup>FSU College of Medicine, Tallahassee, FL, USA, <sup>2</sup>TMH Foundation HealthCare, Tallahassee, FL, USA

#### Introduction:

Palinopsia is a rare phenomenon in which viewed images persist after the original visual stimulus is removed, and can last from minutes to months<sup>1</sup>. The pathophysiology behind palinopsia is still unclear but hypothesized include an exaggeration of normal after imaging, seizure disorders, psychogenicity, or drug induction<sup>2</sup>. Drugs that have serotonin receptor activity such as LSD, trazodone, nefazodone, and topiramate are known to produce palinopsia<sup>3</sup>.

#### Methods:

Single case and literature review: a 30 year old woman, college professor, developed symptoms typical of idiopathic intracranial hypertension. Risk factors included only obesity. There was no history of psychiatric illness. Visual exam demonstrated papilledema with normal acuity, color vision, visual fields, and motility. Treatment was begun with acetazolamide with titration to 2000 mg daily. At high dose patient noticed ghost images trailing behind her moving hand. In addition, she began to see an image first viewed on Facebook of a lotus pod flower. The image appeared on walls, faces, and behind closed eyelids for the next month. The patient understood the images were not real but they invoked anxiety and insomnia. Acetazolamide was discontinued with almost immediate resolution. The patient refused a rechallenge.

#### Results:

The mechanisms of action of topiramate are unclear but include direct or indirect action on serotonin receptors, and as a weak inhibitor of type II and type IV carbonic anhydrase<sup>4</sup>. The present case suggests that perhaps carbonic anhydrase inhibition is its mechanism of action.

#### Conclusions:

Acetazolamide should be added to the list of drugs producing palinopsia.

#### References:

1. Meadows, Munro. Palinopsia. J Neurol Neurosurg Psychiatry. 40(1):5-8; 1977.
2. Hoyt, Walsh, Miller, Newman. Ovid Technologies Inc. Walsh and Hoyt's clinical neuro-ophthalmology. 6th ed. Philadelphia: Lippincott Williams & Wilkins. 620-622; 2005.
3. Fontenelle. Topiramate-induced palinopsia. J Neuropsychiatry Clin Neurosci. 20(2):249-250. 2008.
4. Mula. Topiramate and cognitive impairment: Evidence and Clinical Implications. Ther Adv Drug Saf. 3(6):279-289; 2012.

**Keywords:** Palinopsia, Idiopathic Intracranial Hypertension, Acetazolamide, Topiramate

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Endovascular Intervention In A Chronic Case Of Idiopathic Intracranial Hypertension (IIH)

Muhammad-Atif Zubairi<sup>1</sup>, Andrew Carlson<sup>2</sup>

<sup>1</sup>University of New Mexico/Department of Neurology, Albuquerque, NM, USA, <sup>2</sup>University of New Mexico/Department of Neurosurgery, Albuquerque, NM, USA

### Introduction:

Idiopathic intracranial hypertension (IIH) is characterized by increased intracranial pressure (ICP) in the absence of any known causative factor. Severe headache and visual field disturbance are usually the primary symptoms. If high ICP is left untreated, there is a high risk of continued visual deterioration with risk of blindness. IIH is primarily managed medically. Surgery (CSF diversion procedures with lumboperitoneal shunting or ventriculoperitoneal shunting and optic nerve fenestration) is reserved for patients who have failed medical therapy (20%). Endovascular stenting of the dural venous sinus stenosis in cases of documented venous pressure gradient has shown significant promise in a select subgroup of patients, both in terms of visual and headache outcomes.

### Methods:

Correlation of clinical presentation, imaging with procedural technique and preliminary response by means of a case report.

### Results:

A 34-year old female presented with severe headache and worsening vision. On exam, she was found to have bilateral papilledema. MRI brain evidenced empty sella. MR venography showed focal area of stenosis in the left dominant transverse-sigmoid (TS) sinus. Lumbar puncture (LP) was performed, showed elevated opening pressure (OP) of 47 mm Hg. Cerebral angiography with venous sinus manometry showed a pressure gradient of greater than 20 mm Hg across the left TS sinus. Stenting of the left TS sinus was performed with pressure normalization across the stented segment. She went home on aspirin and clopidogrel. She later reported significant improvement in both vision and headaches. Post-stenting LP showed an OP of 18 mm Hg. Follow-up cerebral angiogram in 3 months showed patent left TS sinus stent. Clopidogrel was subsequently discontinued.

### Conclusions:

Medical management is usually the first line treatment of IIH. Surgery has shown benefit in the resistant cases of IIH with no randomized controlled trial for comparison. Stenting of dural venous sinuses is the newer and safe alternative for the refractory cases of IIH, although limited clinical data is available about the long term procedural benefit.

**References:** None.

**Keywords:** Idiopathic Intracranial Hypertension (IIH), Papilledema, ICP, Angiogram, Stent

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 118

### Optical Aberrations- A Trigger Factor For Migraine?

Rohit Shetty, Chaithra D Aroor<sup>1</sup>, Chaitra Jayadev, Kareeshma Wadia, Bhujang Shetty

*Narayana Nethralaya, Bangalore, India*

#### Introduction:

Migraine is a neurovascular disorder encompassing a wide clinical spectrum of signs and symptoms. The objective of this study was to evaluate whether aberrations of the eye can be trigger factors for migraine.

#### Methods:

This was an observational, cross-sectional study. 30 patients in the age group of 18-35 years with migraine and a spherical equivalent  $\leq -0.75$  were included. The control group consisted of subjects with the same inclusion criteria as the patient group but without a history of headache. All subjects underwent Schirmer's test to exclude dry eyes. A masked investigator measured the aberrations of the eye on the Optical Path Difference III (OPD-III Nidek, Japan). Only mesopic wavefront sizes were considered for the study.

#### Results:

The mean age group of patients with migraine was  $26.21 \pm 6.23$ , and that of the controls was  $27.31 \pm 6.19$ . The root mean square (RMS) of total aberrations in the right eye of patients with migraine was  $1.63 \pm 0.86$  and left eye was  $1.54 \pm 0.88$ . The RMS of total aberrations in the right eye of the control group was  $0.93 \pm 0.33$  and the same in the left eye was  $0.93 \pm 0.42$ . 63% of patients with a history of 'light' as a trigger factor for migraine had a statistically significant ( $p < 0.001$ ) increase in total, higher order aberrations and coma RMS values.

#### Conclusions:

Ocular aberrations may be a trigger factor for migraine. Wavefront analyzers have enabled visual scientists and clinicians to broaden the perspective on the refractive phenomenon in the eye and their influence in health and disease. Studying ocular aberrations during the acute ictal phase and comparing them to normal would give us a better insight and allow a multidisciplinary approach to patients suffering from headache.

**References:** None.

**Keywords:** Migraine And Trigger Factors, Ocular Aberrations, Wavefront Analyzer, OPD III, Non-Organic Visual Disorders

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None



## Poster 119

### An Interesting Field of Study

Andrew S Camp<sup>1</sup>, Angela M Herro, Norman J Schatz

*Bascom Palmer Eye Institute, Miami, FL, USA*

#### Introduction:

Patients with vague visual complaints may have unrealized visual field defects. Detecting defects with confrontational fields is an important diagnostic skill. Homonymous defects are particularly important as they are associated with stroke, tumor, or trauma in approximately 90% of cases. However, other rare causes of homonymous defects must be considered.

#### Methods:

The following 4 unusual cases presented in the last 6 months.

#### Results:

In January, 2014 a 70 year-old man presented with visual distortion. Exam showed a left homonymous hemianopsia, right gaze deviations, and dysmetria. Continuous electroencephalogram (EEG) monitoring demonstrated status epilepticus and MRI showed a 1 centimeter hemosiderin lined lesion in the right occipital lobe suggestive of cavernoma. The hemianopsia resolved with anti-epileptic medication and the cavernoma is being observed. In March, 2014 a 47 year-old man presented with complaints of swirling colors and shadows in his left eye. Exam revealed a left homonymous hemianopsia and episodes of right gaze deviation. Two bilateral occipital seizures occurred during continuous EEG and MRI showed restricted diffusion of the right occipital lobe. The hemianopsia resolved with anti-epileptic medication and the MRI findings were presumed secondary to seizure. In April, 2014 a 41 year-old woman presented with complaints of dizziness and right sided blurriness. She had a right homonymous hemianopsia and MRI revealed enhancement posterior to the left lateral geniculate consistent with a multiple sclerotic plaque. The deficits resolved with intravenous solomedrol. In May, 2014 a 28 year-old woman presented with headache and shadows in her right periphery. Exam showed a right superior homonymous quadrantanopsia and MRI demonstrated enhancement of the left occipital lobe. The MRI findings were consistent with reversible vasoconstriction syndrome related to recent excessive energy drink consumption. The symptoms resolved after stopping the drinks.

#### Conclusions:

Vague complaints can be misleading. However, a careful history and exam can alert clinicians to rare diagnoses.

#### References:

1. Calabrese LH, Dodick DW, Schwedt TJ, Singhal AB. Narrative review: Reversible cerebral vasoconstriction syndromes. *Annals of Internal Medicine*. 2007;146(1).
2. Rosenow F, Alonso-Vanegas MA, Baumgartner C, Blumcke I, Carreno M, et al. Cavernoma-related epilepsy: review and recommendations for management. *Epilepsia*. 2013; 54(12).
3. Shaw S, Kim P, Millett D. Status epilepticus amauroticus revisited: ictal and peri-ictal homonymous hemianopsia. *Archives of Neurology*. 2012;69(11).
4. Zhang X, Kedar S, Lynn MJ, Newman NJ, Biousse V. Homonymous hemianopias: clinical-anatomic correlations in 904 cases. *Neurology*. 2006;66(6).

**Keywords:** Visual Fields, Neuroimaging, Diagnostic Tests (ERG, VER, OCT, HRT, Mferg, Etc)

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 120

### Visual Field Assessment In The “Split-Brain” Patient: Effect Of The Method/Laterality Of Obtaining Patient Responses On Apparent Visual Field Defect Size

J. Alexander Fraser<sup>1,2</sup>, Jorge G. Burneo<sup>1</sup>

<sup>1</sup>Western University, Dept. of Clinical Neurological Sciences, London, ON, Canada, <sup>2</sup>Western University, Dept. of Ophthalmology, London, ON, Canada

#### Introduction:

Corpus callosotomy is an important tool in the management of certain refractory epilepsies, but may cause an interhemispheric disconnection syndrome.

#### Methods:

A 23 year old right-handed man underwent complete corpus callosotomy for intractable epilepsy. Post-operatively, he had features of the callosal disconnection (“split-brain”) syndrome, including anomia for objects presented in the left visual field or left hand, left-sided ideomotor apraxia, and “alien limb phenomenon” in the left hand. When asked to respond verbally to confrontation finger counting, he appeared to have a dense left homonymous hemianopia. An MRI showed post-surgical changes in the corpus callosum, but no lesions within either hemisphere. Visual fields were carefully mapped via several different methods: confrontation finger counting with verbal responses and with saccades, and Goldmann perimetry using the right hand to press the buzzer, using the left hand to press the buzzer, and with saccades to the stimulus light.

#### Results:

Some degree of left homonymous hemianopia was found with all methods, but there was a strong dissociation between the size of the visual field defect and the method by which the patient was asked to respond to left visual field stimuli. He appeared to have a complete left homonymous hemianopia when asked to respond verbally or by pressing the buzzer with his right hand, but had only a partial left inferior homonymous quadrantanopia when allowed to respond with saccades or by pressing the buzzer with his left hand. Of all protocols, saccadic responses produced the smallest visual field defect. Interestingly, the right homonymous hemifield was much less dependent on the method of response.

#### Conclusions:

The extent of a split-brain patient’s visual field defect depends on the method of assessment. By varying the protocol for visual field testing in such a patient, one can expose the intra- and inter-hemispheric connections underlying the normally unified visual experience.

**References:** None.

**Keywords:** Visual Fields, Higher Visual Functions, Perimetry, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 121

### Binasal Hemianopsia

Erica Ballard<sup>1</sup>, Bokkwan Jun

*Mason Eye Institute, Columbia, MO, USA*

#### Introduction:

To report and review a case of binasal heteronymous hemianopic visual field defect and its clinical course and review historical cases of binasal hemianopsia.

#### Methods:

This is a descriptive case report based on clinical records, patient observation and analysis of diagnostic studies.

#### Results:

A 25-year-old woman presented with 4-week history of new onset of headache and visual disturbance. The headache is described as sharp, 10/10, frontal, constant and the visual disturbance is described as difficulty reading, photophobia with no eye pain and no eye movement pain. Her optometrist found that she had binasal visual field defect and referred her for Neuro-Ophthalmic evaluation. On examination, the patient had dense binasal heteronymous hemifield defect without macular sparing on confrontation and also Humphrey automated perimetry. The patient endorsed more increased photosensitivity consistently when the temporal hemianopic retinas were stimulated by slit beam of light. The rest of Neuro-Ophthalmic examination was unremarkable. MRI brain with and without contrast and MRA head were performed and they were unremarkable. Detailed and repeated ophthalmic examination showed no change and further studies including OCT, ERG and VEP were performed and they were unremarkable as well. For her symptomatic management, a week course of oral corticosteroid was initiated. After completion, repeat visual field test showed near complete resolution of the right nasal hemifield defect and mild improvement of the left nasal hemifield defect. The patient started topiramate for headache prevention and her visual field defect continued to improve along with improvement of her headache. .

#### Conclusions:

This is a case of complete binasal heteronymous hemianopsia in setting of new onset migraine. The binasal hemianopsia is uncommon but can occur due to more commonly ocular factors and also intracranial pathologies. The increased photosensitivity of the hemianopic retina could be useful clinical information to judge organic vs nonorganic hemianopsia.

**References:** None.

**Keywords:** Visual Fields Defect, Perimetry, Non-Organic Visual Disorders

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 122

### Voice Processing In Developmental Prosopagnosia

Ran R Liu<sup>1</sup>, Sherryse Corrow<sup>1</sup>, Raika Pancaroglu<sup>1</sup>, Brad Duchaine<sup>2</sup>, Jason JS Barton<sup>1</sup>

<sup>1</sup>University of British Columbia, Departments of Ophthalmology and Visual Sciences, Medicine (Neurology), Vancouver, BC, Canada, <sup>2</sup>Dartmouth University, Department of Psychology, Hanover, NH, USA

#### Introduction:

Prosopagnosia is a selective impairment of face recognition, but voice recognition, another function involving the anterior temporal lobe, has rarely been evaluated to confirm that it is modality-specific. Our previous work has shown that acquired prosopagnosic patients with right anterior temporal lesions have intact voice recognition, while bilateral anterior temporal lesions can impair both face and voice recognition. The anatomic basis of the developmental form of prosopagnosia remains uncertain, and the status of voice recognition is also not known.

#### Methods:

We studied 73 control subjects and 10 subjects with developmental prosopagnosia, whose diagnosis was confirmed on tests of famous face familiarity and short-term familiarization, the Warrington Recognition Memory Test and the Cambridge Face Memory Test. We developed two novel tests: a match-to-sample test of voice discrimination and a test of short-term familiarity for voice recognition, and administered a questionnaire about face and voice identification in daily life.

#### Results:

Nine subjects had intact voice discrimination and voice recognition, with scores superior to those for face recognition. One subject was impaired on voice discrimination and borderline for voice recognition, with equivalent degrees of impairment for both face and voice recognition. The questionnaire results show that the prosopagnosic subject that showed mild impairment in voice processing was unaware he had any voice processing impairments.

#### Conclusions:

Most subjects with developmental prosopagnosia have a modality-specific disorder of face recognition. However, there may be heterogeneity—in some subjects, prosopagnosia may be one component of a multi-modal impairment of person recognition. Tests of voice recognition may help increase the accuracy and specificity of the process of diagnosing prosopagnosia.

**References:** None.

**Keywords:** Developmental, Prosopagnosia, Recognition, Voice, Face

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Fight for Sight Summer Student Fellowship American Academy of Neurology Medical Student Summer Research Scholarship NSERC grant RGPIN 355879-08 Canada Research Chair program

## Poster 123

### Building a Repository For Posterior Cortical Atrophy

Jennifer J. Olds<sup>1</sup>, William Hills<sup>2</sup>

<sup>1</sup>Naval Medical Center San Diego, San Diego, CA, USA, <sup>2</sup>Oregon Health and Science University, Portland, OR, USA

#### Introduction:

Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative disease with prominent cortical visual dysfunction, also known as the visual variant of Alzheimer's disease. Since initial description by Benson in 1988, PCA has been described in small case series and case reports, but only over the last decade has attention been increased. Clinical presentation is heralded by subjective vision loss, yet a normal ophthalmic examination, although there is not yet definitive diagnostic criteria.

#### Methods:

First we conducted a literature review to better understand the signs and symptoms that have been reported. Then, we established a repository to collect patient data with the goal of better understanding the most common signs and symptoms. We hope this will aid in creating diagnostic criteria for the disease.

#### Results:

Literature review demonstrates that PCA has both a large economic burden on a population scale and a significant emotional burden on a personal/family scale. Common signs and symptoms, as well as radiologic and lab abnormalities were investigated and used as the basis for the questions in the repository. The process for and hurdles encountered in developing a repository are discussed.

#### Conclusions:

Earlier diagnosis is key to developing new treatments, hopefully aimed at impacting PCA in a disease modifying state. The only way to ensure earlier diagnosis is by developing more concrete diagnostic criteria and increasing awareness. We developed the only PCA repository to our knowledge and hope this will lead to firm diagnostic criteria in the future.

#### References:

1. Borruat, Posterior Cortical Atrophy: Review of the Recent Literature, Current Neurology and Neuroscience Reports, 13, 406(1-8), 2013.
2. Alzheimer's Association, Alzheimer's disease facts and figures, Alzheimer's and Dementia, 10, e47-e92, 2014.
3. Alves et al, Posterior cortical atrophy and Alzheimer's disease: a meta-analytic review of neuropsychological and brain morphometry studies, Brain Imaging and Behavior, 7, 353-361, 2013.
4. Fiandaca et al, The critical need for defining preclinical biomarkers in Alzheimer's disease, Alzheimer's and Dementia, 10, 196-212, 2014.
5. Woods et al, Dementia: issues in early recognition and intervention in primary care, Journal of the Royal Society of Medicine, 96, 320-324, 2003.

**Keywords:** Posterior Cortical Atrophy, Atypical Alzheimer's Disease, Higher Visual Functions, Repository, Higher Visual Cortical Functions

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Subretinal Choroidal Neovascularization Associated with Unilateral Papilledema and IIH: Case Report**

Jalpa M Patel<sup>1</sup>, Gerry G Maitland<sup>1,2</sup>

<sup>1</sup>Florida State University College Of Medicine, Tallahassee, FL, USA, <sup>2</sup>TMH Foundation Healthcare, Tallahassee, FL, USA

**Introduction:**

Choroidal Neovascularization (CNV) secondary to Idiopathic Intracranial Hypertension (IIH) is rare. CNV produces visual obscurations, decreased visual acuity (VA), and permanent retinal damage if the CNV moves toward and into the fovea. The biomechanics of the inception of CNV with IIH are not fully understood, hypothetically pressure exerted by a swollen optic disk may create a discontinuity in the Bruch's Membrane resulting in growth of vessels under the RPE and sensory retina. Hypoxia due to axonal swelling may create necessary conditions for angiogenesis and vasculogenesis via expression of VEGF, HSCs, and other angiogenic cytokines in subretinal space. CNV associated with IIH may successfully regress following intervention, but there is no consensus on a treatment choice.

**Methods:**

A 51 year old obese woman with history of untreated IIH and weight gain complained of worsening vision in right eye over two months. Visual exam showed VA 20/200 OD with Afferent Pupillary Defect, and 20/20 OS. Fundoscopic exam showed diffuse disk swelling right eye only. Visual fields were normal OS and showed enlarged blind spot OD with arcuate defects and cecocentral depression. Lumbar puncture opening pressure of 300cm with acellular fluid, normal chemistry, and negative cytology. Subsequently, Orbital CT is normal and OCT revealed RNFL thickness at 173µ OD with substantial subretinal fluid consistent with CNV and 93µ OS. In addition there was a subretinal CNV with hemorrhaging.

**Results:**

The immediate cause of rapid visual failure was uncertain until OCT identified the CNV. Once, identified therapy was initiated with Bevacizumab with resultant resorption of subretinal fluid, persistence of CNV, and without change in VA.

**Conclusions:**

Optical Coherence Tomography can be useful in the identification of Choroidal Neovascular Membrane, a condition that should be considered in patients with IIH with rapid visual decline. Failure to respond to Bevacizumab may be due to the presence of hemorrhaging, a finding that has been reported previously in other therapeutic attempts.

**References:**

1. Campochiaro PA. Retinal and choroidal neovascularization. J Cell Physiology, vol.184(3), pg.301-310, 2000.
2. Jamerson SC, Arunagiri G, Ellis BD, Leys MJ. Intravitreal bevacizumab for the treatment of choroidal neovascularization secondary to pseudotumor cerebri. Int Ophthalmology, vol.29(3), pg.183-185, 2009.
3. Lee IJ, Maccheron LJ, Kwan AS. Intravitreal bevacizumab in the treatment of peripapillary choroidal neovascular membrane secondary to idiopathic intracranial hypertension. J Neuroophthalmology, vol.33(2), pg.155-157, 2013.
4. Wendel L, Lee AG, Boldt HC, Kardon RH, Wall M. Subretinal neovascular membrane in idiopathic intracranial hypertension. Am J Ophthalmology, vol.141(3), pg.573-574, 2006.
5. Sathornsumetee B, Webb A, Hill DL, Newman NJ, Biousse V. Subretinal hemorrhage from a peripapillary choroidal neovascular membrane in papilledema caused by idiopathic intracranial hypertension. J Neuroophthalmology, vol.26(3), pg.197-199, 2006.

**Keywords:** Idiopathic Intracranial Hypertension, Choroidal Neovascular Membrane, OCT Scans, Papilledema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## The Initiation Of Smooth Pursuit Eye Movements In Anisometropic Amblyopia

Rana A. Raashid<sup>1</sup>, Alan Blakeman<sup>2</sup>, Herbert C. Goltz<sup>1,2</sup>, Agnes M.F. Wong<sup>1,2,3</sup>

<sup>1</sup>Neuroscience and Mental Health, Hospital for Sick Children, Toronto, ON, Canada, <sup>2</sup>Department of Ophthalmology and Vision Science, University of Toronto, Toronto, ON, Canada, <sup>3</sup>Department of Ophthalmology and Vision Science, Hospital for Sick Children, Toronto, ON, Canada

### Introduction:

Amblyopia is a spatiotemporal visual impairment caused by abnormal visual experience early in development. Recent studies have shown that amblyopia impacts visuomotor behaviors in addition to impairing sensory functions. Here we investigate the effects of anisometropic amblyopia on smooth pursuit eye movements responsible for accurately maintaining moving objects on the fovea. We hypothesize that given the visual processing delays in amblyopia there will be a significant delay in the initiation of pursuit in patients during amblyopic eye viewing.

### Methods:

Fourteen visually normal controls and 9 people with anisometropic amblyopia participated. The participants were presented with a red laser moving at  $\pm 15^\circ/\text{s}$  horizontally for approximately one second for 40 trials. The experiment was repeated for each participant under three viewing conditions in the following order: amblyopic/nondominant eye, binocular, and fellow/dominant eye viewing. Outcome measures were pursuit initiation latency, open-loop gain (eye/target velocity ratio 100 ms post-onset), and steady state gain (eye/target velocity ratio throughout movement).

### Results:

When viewing monocularly with the amblyopic eye, patients took longer to initiate pursuit movements ( $203 \pm 20$  ms) compared to controls viewing with their nondominant eye ( $183 \pm 17$  ms,  $p=0.004$ ). However, the latency of pursuits in patients during binocular ( $168 \pm 17$  ms) and monocular fellow eye ( $176 \pm 22$  ms) viewings was comparable to controls (binocular:  $172 \pm 19$  ms and dominant eye:  $169 \pm 15$  ms respectively). Mean open-loop gains and steady state gains did not differ significantly between the two groups.

### Conclusions:

This study provides novel evidence of delayed initiation of smooth pursuit eye movements in people with anisometropic amblyopia with the amblyopic eye viewing. A similar observation was documented previously for saccadic eye movements in anisometropic amblyopia, and we suggest that it might be due to a delay in visual processing of target motion in amblyopia. Once initiated, however, smooth pursuit movement accuracy in amblyopic participants is equivalent to that seen in visually normal observers.

**References:** None.

**Keywords:** Ocular Motility, Smooth Pursuit Eye Movement, Amblyopia, Visual Processing

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## The Pulfrich Phenomenon: An Objective Signature-Biomarker of MS Pathophysiology

Millad J Sobhanian<sup>1</sup>, Rohit Agarwal<sup>1</sup>, Darrel Conger<sup>1</sup>, Amy Conger<sup>1</sup>, Laura J Balcer<sup>2,3</sup>, Robert Rennaker<sup>4</sup>, Shin C Beh<sup>1</sup>, Owen White<sup>5,6</sup>, Randy Kardon<sup>7</sup>, Teresa Frohman<sup>1</sup>, Elliot M Frohman<sup>1,8</sup>

<sup>1</sup>Department of Neurology, University of Texas Southwestern Medical Center, Dallas, TX, USA, <sup>2</sup>Department of Neurology, New York University, New York, NY, USA, <sup>3</sup>Department of Ophthalmology, New York University, New York, NY, USA, <sup>4</sup>School of Brain and Behavioral Sciences University of Texas at Dallas Richardson, TX, USA, <sup>5</sup>Department of Neurology, Royal Melbourne Hospital Parkville, Australia, <sup>6</sup>Department of Medicine, University of Melbourne Parkville, Australia, <sup>7</sup>Department of Ophthalmology and Visual Science, University of Iowa Hospitals and Clinics Iowa City, IA, USA, <sup>8</sup>Department of Ophthalmology, University of Texas Southwestern Medical Center Dallas, TX, USA

### Introduction:

The Pulfrich Phenomenon (PF) is the illusory perception that an object moving linearly along a 2-D plane, appears to instead move in an elliptical 3-D trajectory. The PF is thought to reflect inter-eye differences within the visual processing network. We have developed an innovative method by which we can ascertain both the presence and magnitude of the PF.

### Methods:

MS patients with a history of acute optic neuritis (AON) and a control group were asked to indicate whether movement of a pendulum bob followed a 2D linear versus a 3D elliptical object-motion trajectory. In the case of the latter, participants were asked to ascertain the magnitude and direction of the 3D motion, by virtue of the bob's approximation with respect to one of five colored wires oriented horizontally in close proximity to the bob. Subjects were asked to make the same approximations following monocular application of graduated neutral density filtering (G-NDF).

### Results:

Monocular application of G-NDF to controls resulted in the stereotypic illusion of PF, the magnitude of which intensified and corresponded to increasing levels of filter tinting. Alternately, the application of the G-NDF to AON eyes produced a PF of greater prominence whereas filtering the less affected eyes resulted in attenuation of the PF through consequent neutralization of the interocular brightness disparity brought on by disease.

### Conclusions:

We have designed a novel, objective method by which to characterize the presence and magnitude of PF in normal subjects (via G-NDF), and MS patients with AON history. Application of G-NDF to the less affected eye in MS patients experiencing PF can be employed to identify the magnitude of filtering necessary to abolish the effect. Furthermore, PF can be employed as a pathophysiologic signature-biomarker for the investigation of changes in visual system architecture and corresponding functional consequences.

**References:** None.

**Keywords:** Pulfrich, Psychophysics, Multiple Sclerosis, Acute Optic Neuritis

**Financial Disclosures:** Laura Balcer received personal compensation from Biogen Idec and consulting for Biogen Idec, Vaccinex and Genzyme. She is on a clinical trial advisory board for Biogen-Idec. Randy Kardon: Novartis steering committee OCTiMS multicenter international study to determine the value of optical coherence tomography in monitoring MS Acorda steering committee to plan research studies of a new medication to improve visual symptoms in MS Zeiss Meditec consultant for perimetry Robert Rennaker is the owner of the company Vulintus LLC Shin C Beh is the recipient of the 2012 Biogen Idec Clinical MS Fellowship award Teresa Frohman has received speaker fees for Biogen Idec, Novartis and Acorda and received compensation for participation on Scientific Advisory Board for Biogen Idec, Novartis and Questcor in the past year Elliot Frohman has participated in the speakers' bureau for Teva Neurosciences, Acorda, and Novartis and has received consulting fees from TEVA Neurosciences, and Acorda.

**Grant Support:** None.



## Distractibility In Multiple Sclerosis: A Potential Cause Of Morbidity

Derek Sears<sup>1,2</sup>, Ashley Frohman<sup>1,2</sup>, Teresa C Frohman<sup>1,2</sup>, Elliot M Frohman<sup>1,2</sup>, Lynette Millist<sup>4,5</sup>, Meaghan Clough<sup>3</sup>, Joanne Fielding<sup>3,5</sup>, Owen B White<sup>3,4,5</sup>

<sup>1</sup>Department of Neurology & Neurotherapeutics University of Texas Southwestern School of Medicine, Dallas, TX, USA, <sup>2</sup>Texas Tech Health Sciences Center Paul L. Foster School of Medicine, Dallas, TX, USA, <sup>3</sup>School of Psychological Sciences, Monash University, Clayton, Australia, Melbourne, Australia, <sup>4</sup>Department of Neurology, Royal Melbourne Hospital, Parkville, Australia Melbourne, Australia, <sup>5</sup>Department of Medicine, University of Melbourne, Parkville, Australia Melbourne, Australia

### Introduction:

Objective: To undertake a pilot study to elucidate objective and reproducible ocular motor abnormalities reflecting cognitive dysfunction in multiple sclerosis (MS). MS patients near universally suffer fatigue and impaired concentration, often prior to clinically evident "eloquent" lesions. Pathology in MS is diffuse, affecting widespread interconnecting networks subserving cognition. Dysfunction in these networks represents the commonest manifestations of MS impacting gainful employment, provoking demoralization, and compromising quality of life. An ominous consequence of cognitive impairment involves the predilection for dysphoria, reduced socialization and abulia, compromising adherence to treatment and healthy living.

### Methods:

Methods: We employed paradigms of cognitive network-mediated activation, inhibition and executive function in the integrated ocular motor system. Patients were instructed to perform saccadic eye movements toward or away from a visual stimulus. We also investigated the capacity of subjects to persist with a task and suppress distraction, the paradigms being: 1. Appearance of a differentially colored cross, directing the subject to make a prosaccade to a visible target or an antisaccade away from the target. 2. Subjects were instructed to pursue a target and not look at distractor targets occurring unpredictably. Error rate represented the primary outcome. Further, we assessed eye movement onset latency as a measure of central nervous system (CNS) information processing speed.

### Results:

Results: Saccadic latency was significantly prolonged in MS patients compared control subjects. MS patients exhibited a significantly higher error rate suggestive of compromise in 'top-down' inhibitory mechanisms (emanating from the dorsolateral prefrontal cortex and other cognitively eloquent regions). Patients showed greater susceptibility to distraction in smooth pursuit paradigms. Characterising and quantifying these deficits may permit the development of focused rehabilitation and allow evaluation of response to new neurotherapeutic agents.

### Conclusions:

Conclusions: Results in this pilot study corroborate the hypothesis that MS patients exhibit slowed information processing speed and have difficulty suppressing distraction. Our patients demonstrated significant prolongation of onset latency for saccadic eye movements.

**References:** None.

**Keywords:** Demyelinating Disease, Higher Visual Cortical Functions, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Disconjugacy Of Eye Alignment Is Greater With Near Fixation During Binocular Viewing In Amblyopia

Vivian Xu<sup>1</sup>, Robert Geary, Boris Gramatikov, David L. Guyton, Kristins Irsch, Howard S. Ying

*Wilmer Eye Institute at The Johns Hopkins University, Baltimore, MD, USA*

### Introduction:

To determine whether ocular disconjugacy in amblyopic subjects is dependent on viewing condition.

### Methods:

Binocular eye movements were recorded at 500 Hz using the EyeLink 1000 eye tracker (SR Research Ltd., Ontario, Canada). Twenty subjects (nine normal and eleven amblyopic; age: 8-45 years) were asked to fixate on a blue cross subtending 0.5° at a near distance of 57 cm or at a far distance of 4 m for 20-second epochs of binocular or monocular viewing. Disconjugacy of eye alignment was estimated by the area of the 68% bivariate contour ellipse (BCEA) for the difference between right and left eye positions, by the percentage of fixation time within a 0.1°x 0.1° range, and microsaccade characteristics. Mean ± standard error of the mean were shown, log(BCEA) was used to normalize the distribution, and significance testing was performed with the Student's t-test.

### Results:

Normal subjects during binocular viewing showed a 68% BCEA of  $0.75 \text{ deg}^2 \pm 0.09 \text{ deg}^2$  with near fixation and  $0.81 \text{ deg}^2 \pm 0.14 \text{ deg}^2$  with far fixation ( $P=0.69$ ) while subjects with amblyopia during binocular viewing showed a 68% BCEA of  $5.60 \text{ deg}^2 \pm 0.78 \text{ deg}^2$  with near fixation and  $3.08 \text{ deg}^2 \pm 0.31 \text{ deg}^2$  with far fixation ( $P=0.01$ ). Mean percentage of conjugate fixation time for normal subjects was  $99.5\% \pm 0.3\%$  for near fixation and  $99.6\% \pm 0.1$  for far fixation ( $P=0.66$ ), and for amblyopic subjects was  $98.4\% \pm 0.2\%$  for near fixation and  $99.2\% \pm 0.1\%$  for far fixation ( $P=0.02$ ). Disconjugacy was associated with more rapid slow drift, more microsaccades, and more square wave jerks. Monocular viewing trials showed more disconjugacy than binocular trials for normal subjects but not for amblyopic subjects.

### Conclusions:

Disconjugacy increases with near fixation for amblyopic subjects during binocular viewing due to increased slow drift and microsaccades. Further research is required to determine which metrics have greater diagnostic utility in amblyopia.

**References:** None.

**Keywords:** Higher Visual Functions, Nystagmus, Ocular Motility, Pediatric Neuro-Ophthalmology, Higher Visual Cortical Functions

**Financial Disclosures:** RG, KI, BG, DLG, and HSY have a pending patent application for the above technology with The Johns Hopkins University

**Grant Support:** NIH R01-EY19347, Research to Prevent Blindness Disney Award, Research to Prevent Blindness core grant, William Cross Foundation.

## Poster 129

### An Adjustable Magnetic Prism Carrier for Strabismus Evaluation

David S Bardenstein<sup>1</sup>

*University Hospitals of Cleveland Eye Institute, Cleveland, OH, USA*

#### **Introduction:**

The analysis and quantitation of strabismus can be challenging to examiner and patient, especially when the deviation is large or complex. This can be due to the difficulty of holding multiple prisms in proper alignment and also the difficulty of conducting trials of prisms during real life use for prolonged periods. Most trial frame sets have limits on prism size. We have designed a continuously adjustable, head mounted magnetic prism carrier (MPC) which allows for placement of any strength prism in proper alignment to facilitate strabismus evaluation.

#### **Methods:**

We analyzed the ideal goals of a prism carrier device and based on those designed a simple carrier mechanism available in 2 forms. The features sought included the ability to: easily and incrementally adjust the location of the prisms in 3 dimensional space, allow the use of multiple or thick prisms with extreme stability, manipulate the prisms independent of wearing glasses or trial frames, hold the prisms held in place without the participation of the examiner to allow patients to perform trial wearings prior to prescribing, achieve all these goals in children as well as adults. Devices were used on an unselected series of child and adult patients. Patients underwent strabismus analysis in primary gaze with handheld prisms and the MPC. Examiners were queried as to the comparative ease of use. Patients were queried as to comfort of the exam.

#### **Results:**

The devices were used successfully in all patients and tolerated well without inducing discomfort or fear, when used with children. Examiners reported the MPC was equal or easier to use in all cases. Prism exchange was easier with the MPC for moderate to larger deviations.

#### **Conclusions:**

The continuously adjustable magnetic prism carrier, is a simple useful instrument which can facilitate an often challenging diagnostic process with limited cost, shorter exam time and equal accuracy.

**References:** None.

**Keywords:** Adult Strabismus, Diagnostic Tests, Ocular Motility

**Financial Disclosures:** David Bardenstein MD InventorDevice Licensed through University Hospitals of Cleveland to Gulden Ophthalmic

**Grant Support:** Rainbow Pediatric Innovation Fund Jeanne P Schroeder Fund

## Poster 130

### Patient Satisfaction With Prismatic Correction Of Diplopia

Shauna E Berry<sup>1</sup>, Matthew D Kay<sup>2</sup>, Clint W Kellogg<sup>1</sup>, Kathryn E Ireland <sup>1</sup>

<sup>1</sup>Larkin Community Hospital, Miami, FL, USA, <sup>2</sup>Nova Southeastern, Ft. Lauderdale, FL, USA

#### Introduction:

We retrospectively evaluated the percentage of patients with an ocular motility disorder that opted for strabismus surgery versus prismatic correction.

#### Methods:

All new patients billed between March and June 2013 with a 378.xx ICD9 code were selected for chart review. We assessed whether the diplopia was managed with observation, occlusion, prism glasses, or strabismus surgery. Patients diagnosed with ocular myasthenia gravis were excluded, while diplopic events from a cranial nerve palsy were analyzed separately. Other factors taken into consideration were patient satisfaction with prism glasses, previous strabismus surgery, if surgery was discussed as an option, and if pathology was evident on radiologic studies.

#### Results:

122 patients with a total of 140 independent diplopic events were reviewed in regards to management. Of these diplopic events, 34% resulted from a cranial nerve palsy. In the remaining diplopic events, surgery was discussed with 20 % of patients of which 39% selected surgical management. Prism glasses were prescribed to 58% of patients, of which 82% admitted to improvement. In the patients diagnosed with a cranial nerve palsy as the cause of their diplopia, prism glasses were prescribed to 38%, while only 20% selected surgical management

#### Conclusions:

A large volume of patients present to neuro-ophthalmology practices with ocular motility disorders. In this setting it is important that the neuro-ophthalmologist and strabismologist work together to manage the diplopia. Strabismus surgery was discussed as an option with the patient when deemed appropriate. Diplopic events resulting from a cranial nerve palsy spontaneously resolved in 62% of cases, and therefore did not require management with either prism glasses, surgery, or occlusion. Other factors such as positive imaging studies and failure to follow-up may explain the trend towards non-surgical management. Therefore, it can be concluded that the majority of patients presenting with diplopia were satisfied with conservative non-surgical management.

**References:** None.

**Keywords:** Adult Strabismus, Diplopia, Ocular Motility, Cranial Nerve Palsy, Strabismus Surgery

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Duane Retraction Syndrome in Duchenne Muscular Dystrophy

Thomas M Bosley<sup>1</sup>, Mustafa A.M. Salih<sup>2</sup>, Darren T Oystreck<sup>3</sup>, Khaled K Abu-Amero<sup>1</sup>

<sup>1</sup>King Saud University/Ophthalmology, Riyadh, Saudi Arabia, <sup>2</sup>King Saud University/Pediatrics, Riyadh, Saudi Arabia, <sup>3</sup>University of Stellenbosch/Ophthalmology, Tygerberg, South Africa

### Introduction:

Patients with Duchenne muscular dystrophy (DMD) almost always have clinically normal eye movements, although two DMD patients have been described previously with Duane retraction syndrome (DRS), the most common congenital cranial dysinnervation disorder (CCDD). We describe the clinical features of a boy with Duchenne muscular dystrophy, bilateral Duane retraction syndrome, and other neurologic problems.

### Methods:

This child was followed for more than a decade by Pediatric Neurology and Neuro-ophthalmology. His chart was reviewed, including the results of a muscle biopsy. Multiplex ligation-dependent probe amplification (MLPA) was used to interrogate the 79 exons of the *dystrophin* gene.

### Results:

The clinical diagnosis of Duchenne muscular dystrophy was based on the proband's clinical course of progressive proximal weakness, very elevated creatine kinase levels, and a muscle biopsy showing significant dystrophic changes including contracted, degenerative, and regenerative fibers. He had bilateral DRS with otherwise normal ocular motility. He also had developmental delay, mild mental retardation, and seizures. MLPA documented duplication of exons 3 and 4 of the *dystrophin* gene.

### Conclusions:

This boy had syndromic DRS in which bilateral type 3 DRS occurred in association with genetically and clinically proven DMD, developmental delay, mental retardation, and seizures. He is the third patient to be reported with DRS and DMD, the second with bilateral DRS and the only one with other neurologic features. DRS is the most common CCDD, and DMD is the most common muscular dystrophy in males, leaving open the possibility that these two disorders have occurred together at least three times by chance. Mutated *dystrophin* is present in extraocular muscles and in the central nervous system (CNS), and the co-occurrence of DRS with DMD may be the developmental result of the genetic muscle abnormality, the CNS effects of mutated *dystrophin*, other co-existent genetic abnormalities, or some combination of these factors.

**References:** None.

**Keywords:** Duchenne Muscular Dystrophy, Duane Retraction Syndrome, Congenital Cranial Dysinnervation Disorders, Dystrophin, Seizures

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** KSA National Program for Science and Technology Grant # 12-MED2621-02

## Peribulbar Botulinum Toxin As A Treatment For Symptomatic Opsoclonus In Oculopalatal Myoclonus

Katherine E. Duncan<sup>1</sup>, Moran Levin, Janet L Alexander, Erica L Archer

*University of Maryland Department of Ophthalmology, Baltimore, MD, USA*

### Introduction:

Oculopalatal myoclonus is the result of a lesion of the dento-rubro-olivary tract in the brainstem and is characterized by rhythmic movements of the soft palate and pendular vertical nystagmus.<sup>1</sup> The acquired nystagmus results in debilitating oscillopsia. Various pharmacologic agents and surgical techniques have been employed to alleviate these symptoms with limited success. There have been very few reports of treatment with retrobulbar botulinum injection in the literature.<sup>1,2</sup> We describe a case of oculopalatal myoclonus in which peribulbar botulinum toxin injection was extremely successful in alleviating nystagmus and improving visual acuity.

### Methods:

A 62 year old male with a history of left midbrain and pontine hypertensive hemorrhage 2 years prior presented to the ophthalmology clinic with complaint of "unsteady images." His vision was 20/80 OU. Motility exam was significant for left gaze palsy and upbeating nystagmus OU. Nystagmus decreased with convergence however attempts to induce convergence with prism did not result in any symptomatic relief. The decision was made to try a peribulbar injection of 25 units of botulinum toxin to the left orbit.

### Results:

The patient returned 2 weeks later with dramatic reduction in nystagmus OS and visual acuity of 20/30 in that eye. The patient noted significant improvement in his quality of life and is now pursuing peribulbar botulinum toxin injection in the right eye.

### Conclusions:

Patients with acquired nystagmus suffer from incapacitating oscillopsia, a condition that has historically had few successful treatment options. Peribulbar botulinum toxin injection represents a safe and efficacious treatment that results in significant improvement in visual acuity as well as quality of life for these patients.

### References:

1. Talks SJ and Elston JS. Oculopalatal myoclonus: eye movement studies, MRI findings and the difficulty of treatment. *Eye* 1997;11:19-24
2. Repka MX, Savino PJ, Reinecke RD. Treatment of acquired nystagmus with botulinum neurotoxin A. *Arch Ophthalmol* 1994;112:1320-1324

**Keywords:** Nystagmus, Oscillopsia, Opsoclonus, Botulinum Toxin, Oculopalatal Myoclonus

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 133

### Thalamic Stroke In A Young Patient Presenting With Sudden Onset Large Skew Deviation

Oana M. Dumitrascu<sup>1</sup>, Patrick Lyden<sup>1</sup>, Mohamad Shafie<sup>2</sup>

<sup>1</sup>*Cedars-Sinai Medical Center, Department of Neurology, Los Angeles, CA, USA*, <sup>2</sup>*Loma Linda University, Department of Neurology, Loma Linda, CA, USA*

#### Introduction:

Introduction: Skew deviation usually localizes to the utriculo-ocular and brainstem pathways. Rare literature reports based largely on imaging findings suggested medial thalamic implication in the supranuclear vertical gaze control.

#### Methods:

Case Report: A 26-year-old male without any past medical history presented 1.5 hours after abrupt onset of blurry and tilted vision, vertical binocular diplopia, as well as postural and gait instability during defecation. On exam, his right eye was hypertropic and slightly incyclotorted, with no change in the large prismatic deviation in different gaze positions or head tilt. The upright-supine test was positive. Direction changing eccentric nystagmus and truncal ataxia accompanied. Noncontrast head CT and CT angiogram of head and neck were unremarkable. Clinical presentation was concerning for an acute ischemic stroke localizing to the brainstem. Given his severely disabling symptoms, a decision was made to treat with intravenous tissue plasminogen activator per acute stroke protocol. One hour later visual symptoms and imbalance were resolved, and normal ocular alignment was noted on the examination. Brain magnetic resonance imaging revealed an isolated area of restricted diffusion in involving the mid portion of the right thalamus consistent with an acute ischemic stroke. There was no evidence of any other previous fluid attenuated inversion recovery (FLAIR) sequence hyperintensities suggestive of older lesions. Both transcranial Doppler ultrasound and transthoracic echocardiogram verified the presence of a large patent foramen ovale (PFO). Further complete stroke work up including full hypercoagulable panel was negative. Patient was discharged home asymptomatic, on daily aspirin and enrolled in a clinical trial for the PFO closure.

#### Conclusions:

Conclusions: Isolated thalamic infarct can present with sudden onset vertical diplopia and skew deviation. The acute clinical presentation must raise the suspicion for the appropriate and emergent management of stroke. More clinico-pathological correlation and functional imaging studies are needed to characterize the involved pathways.

**References:** None.

**Keywords:** Skew Deviation, Vascular Disorders, Neuroimaging, Stroke

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

### Ophthalmological Spectrum of Locked-In Syndrome

Martin Graber<sup>1</sup>, Georges Challe<sup>1</sup>, Marie-Francoise Alexandre<sup>1</sup>, Bertrand Gaymard<sup>2</sup>, Bahram Bodaghi<sup>1</sup>, Phuc LeHoang<sup>1</sup>, Valérie Tuitou<sup>1</sup>

<sup>1</sup>1. Ophthalmology Department, DHU Vision and Handicaps, Pitie-Salpetriere Hospital, PARIS, France, <sup>2</sup>2. Neurology Department, Pitie-Salpetriere Hospital, PARIS, France

#### Introduction:

Although a few authors have reported abnormal eye movements in Locked in syndrome (LIS), there is no study focusing on the large spectrum of ophthalmological manifestations of LIS and their consequences on the visual function. The aim of this study is to describe the range of ophthalmological manifestations observed in patients with LIS, and to evaluate their visual impairment.

#### Methods:

Thirteen cases of LIS seen in a single tertiary center between 1997 and 2013 were retrospectively reviewed. For each patient, a neuro-ophthalmological evaluation was performed, including whenever possible, refraction, visual acuity, eye movements, slit lamp examination (portable device), pupils, visual field (confrontational) and fundus examination. For each evaluation, the environmental parameters (luminosity, contrasts, head position, distance of examination) were modified in order to achieve the best evaluation of the visual function.

#### Results:

Mean visual acuity was 20/80. Diplopia was reported by 46 % of patients. Visual field was impaired in 23% of patients. Ocular movements were impaired in 77% of patients: 69% presented unilateral or bilateral VIth nerve palsy, 8% had a complete ophthalmoplegia, 61% had a nystagmus with oscillopsia, and 38% had oculopalatal tremor. Bilateral mydriasis was observed in 22% of patients. Lagophthalmia, keratitis and dry eye manifestations were observed in most patients and constituted the main complaint of the patients.

#### Conclusions:

Eventhough it is usually admitted that the visual function is preserved in locked-in syndrome, we demonstrated that most of the patients suffer from visual impairment from several origin. Visual acuity is poor, nearly half of the patients suffer from disabling diplopia or oscillopsia and most of them suffer from dry eye syndrome. However, visual field seems to be more preserved than expected. A specialised ophtalmological work-up should be performed for every patient with LIS in order to optimize their visual function, to improve their quality of life and to evaluate their ability to communicate.

**References:** None.

**Keywords:** Locked-In Syndrome, Opsoclonus, VI Nerve Palsy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Cigarette Smoking and Activities of Daily Living in Ocular Myasthenia Gravis

Sean M Gratton<sup>1,2,3</sup>, Angela Herro<sup>1</sup>, William Feuer<sup>1</sup>, Byron L Lam<sup>1</sup>

<sup>1</sup>Bascom Palmer Eye Institute, Miami, FL, USA, <sup>2</sup>University of Missouri--Kansas City Department of Neurology and Cognitive Neuroscience, Kansas City, MO, USA, <sup>3</sup>University of Missouri--Kansas City Department of Ophthalmology, Kansas City, MO, USA

### Introduction:

Myasthenia Gravis (MG) is an autoimmune disorder of the neuromuscular junction most often caused by autoantibodies targeting the acetylcholine receptor (AChR). Cigarette smoking is known to affect neuronal AChRs and it has been shown to influence other autoimmune diseases, but the effect of cigarette smoking on MG has not been thoroughly investigated.

### Methods:

Cigarette smoking status and MG disease activity were assessed by a prospective telephone survey administered to patients with MG encountered from 2006-2014 in a single neuro-ophthalmology practice. MG disease activity was evaluated by calculating the Myasthenia Gravis-specific Activities of Daily Living (MG-ADL) score, and cigarette smoking status was determined by administering questions adapted from the National Health Interview Survey.

### Results:

84 patients with MG were identified during the study period; 40 were excluded due to an uncertain diagnosis or an inability to obtain correct contact information. In all, surveys were administered to 44 patients. Comparison of MG-ADL scores between current smokers ( $5.6 \pm 4.5$ ), former smokers ( $2.9 \pm 3.1$ ), and never smokers ( $1.4 \pm 2.5$ ) revealed a statistically significant relationship ( $p=0.003$ , one way analysis of variance) where current smokers had the highest MG-ADL scores and never smokers the lowest. Comparison of the MG-ADL ocular subscore revealed the same relationship (current  $3.4 \pm 2.6$ , former  $1.8 \pm 2.1$ , never  $1.1 \pm 1.5$ ,  $p=0.031$ , table 2 and figure 4). There were borderline significant correlations of pack-years with MG-ADL score ( $r=0.30$ ,  $p=0.051$ ) and MG-ADL ocular subscore ( $r=0.27$ ,  $p=0.074$ ).

### Conclusions:

Our study reveals a significant association between cigarette smoking and disease activity in MG. In light of these results, we urge clinicians to strongly recommend smoking cessation to myasthenic patients in the hopes that successful smoking cessation may lead to improved MG disease activity. Even though this study does not specifically address whether MG disease activity may improve after smoking cessation, the overwhelming health benefits from smoking cessation make this recommendation fairly uncontroversial.

**References:** None.

**Keywords:** Myasthenia Gravis, Neuroophthalmology & Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Supported by NIH Center Core Grant P30EY014801 and Research to Prevent Blindness Unrestricted Grant.

## An Evaluation Of Educational Neurological Eye Movement Disorder Videos Posted On Internet Video Sharing Sites

Simon J Hickman<sup>1</sup>

*Royal Hallamshire Hospital / Department of Neurology, Sheffield, United Kingdom*

### Introduction:

Internet video sharing sites allow the free dissemination of educational material. This study investigated the quality and educational content of videos of eye movement disorders posted on such sites.

### Methods:

Videos were identified by searching the title of the eye movement disorder of interest on the video sharing sites YouTube and Vimeo, and also by following the links suggested. The number of hits and the hits per day since the video was uploaded were recorded. The videos were then rated for the picture quality and also sound quality if there was a verbal commentary. Any errors in the title and content were noted. Lastly, the educational content of the videos were rated as to the presence of a description of the abnormality, whether the anatomical location of the lesion was identified and an appropriate diagnosis discussed.

### Results:

130 videos were viewed. The mean number of hits was 14532 (range 5-249480) with a mean hit rate of 8.45/day (range 0.025-91.45). The mean picture quality was 1.93/3 and the mean sound quality was 2.67/3. 20 (15.4%) had errors and it was not possible in 3 (2.3%) to confirm the diagnosis from what was shown on the video. 48 (36.9%) had no commentary apart from the title. 26 (20%) had excellent educational value in that they included a commentary on the eye movement abnormality, the anatomical location of the lesion, if appropriate, and the pathological diagnosis, of which 5 (3.8% of total) combined these with excellent picture and sound quality.

### Conclusions:

It is possible to view a wide range of eye movement abnormalities on these sites, however the lack of peer review means that there is a significant error rate. The educational value of a large proportion of the videos is limited by a lack of commentary on the abnormalities shown.

**References:** None.

**Keywords:** Nystagmus, Ocular Motility

**Financial Disclosures:** The author had no disclosures.

**Grant Support:** None.

**Poster 137**

**How Do Patients with Strabismus Locate Visual Targets?**

Jonathan C. Horton<sup>1</sup>, Daniel L. Adams, John R Economides

*UCSF/Ophthalmology, San Francisco, CA, USA*

**Introduction:**

In alternating strabismus, it is unknown which eye provides information about the location of the next visual target so that an accurate saccade can be made.

**Methods:**

Subjects with alternating exotropia and no amblyopia wore red/blue filter glasses for dichoptic stimulation while viewing stimuli rear-projected onto a tangent screen. Each trial began with a fixation cross, visible to either the right, left, or both eyes. After the subject fixated the cross, a peripheral stimulus appeared for 200 msec, visible to either the right, left, or both eyes. The subject's task was to look at it.

**Results:**

There were 3 main findings: 1) the eye to which the target was presented usually was the eye used to fixate the target; 2) when stimuli were presented in the far nasal field of the perceiving eye, subjects occasionally performed a "cross-over" saccade by placing the other eye on the target, to avoid having to make a large adducting saccade. In such cases, information about target location was obtained in one eye and used to program a saccade for the other eye. Cross-over saccades had a longer latency and were less accurate; 3) In a separate experiment, binocular sensory maps were compiled to delineate the portions of the visual field perceived with each eye. For each subject, the layout of suppression scotomas closely matched the map of target acquisition by eye. In other words, targets were fixated by the eye used to perceive them.

**Conclusions:**

By comparing maps of sensory perception and oculomotor behavior, these studies have shown how patients with strabismus are able to fixate accurately objects of interest in their visual environment, despite misalignment of the eyes. The deviated eye, rather than being completely suppressed, is engaged in locating targets that precede a fixation swap.

**References:** None.

**Keywords:** Diplopia, Exotropia, Saccade, Suppression, Strabismus

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** National Eye Institute

## Superior Cerebellar Peduncle Demyelination Causing Geotropic Central Positional Nystagmus

Cristina Duque<sup>1</sup>, Daniela Pereira<sup>2</sup>, Margarida Amorim<sup>3</sup>, Sonia Batista<sup>1</sup>, Joao Lemos<sup>1</sup>

<sup>1</sup>Department of Neurology, Coimbra University Hospital Center, Coimbra, Portugal, <sup>2</sup>Department of Neuradiology, Coimbra University Hospital Center, Coimbra, Portugal, <sup>3</sup>Department of Otolaryngology, Coimbra University Hospital Center, Coimbra, Portugal

### Introduction:

While the presence of spontaneous nystagmus in multiple sclerosis (MS) patients frequently constitutes a sign of disease relapse, the occurrence of positional nystagmus is more often due to a non-MS cause (e.g., benign paroxysmal positioning vertigo (BPPV)). Rarely, a strategic infratentorial lesion can cause central positional/positioning nystagmus (CPPN) and mimic BPPV. Apogeotropic and downbeat CPPN constitute the commonest forms. We describe a MS patient with geotropic CPPN caused by a superior cerebellar peduncle (SCP) demyelinating plaque.

### Methods:

A 46-year old woman with a previous diagnosis of relapsing-remitting MS presented with disabling positional vertigo and vomiting exacerbated by head movements while lying down. Eye movements were filmed and subsequently recorded using Video-nystagmography (VNG). Brain magnetic resonance imaging (MRI) was performed.

### Results:

Exam in the upright position demonstrated mild spontaneous right horizontal nystagmus with a vertical downbeating component. In the supine position with the head straight, nystagmus became more intense and accompanied by vertigo and vomiting; head rotations caused persistent and non-fatigable direction-changing geotropic horizontal nystagmus. Log-roll maneuver did not abate nystagmus. The remainder of the neurologic exam was normal, except for mild gait ataxia. VNG performed on the second day revealed right horizontal and downbeat nystagmus (3.1°/s; 0.3°/s) in the upright position that changed to right horizontal and upbeat nystagmus (5.2°/s; 4.4°/s) in the supine position; geotropic nystagmus was no longer evident. T2/FLAIR brain MRI showed a new demyelinating non-enhancing lesion in the inner part of the right SCP. Patient was treated with a 5-day course of 1 g/day intravenous methylprednisolone. After 4 days, nystagmus and vertigo had completely subsided.

### Conclusions:

This is the first report of central positional geotropic nystagmus associated with focal superior cerebellar peduncle demyelination. Disruption of the central otolithic connections is hypothesized to be the mechanism underlying the presence of central positional nystagmus in this case.

### References:

- 1.) Frohman EM, Zhang H, Dewey RB, Hawker KS, Racke MK, et al. Vertigo in MS: utility of positional and particle repositioning maneuvers. *Neurology*. 2000 Nov 28;55(10):1566-9.
- 2.) Brandt T. Positional and positioning vertigo and nystagmus. *J Neurol Sci*. 1990 Jan;95(1):3-28.
- 3.) Anagnostou E, Varaki K, Anastasopoulos D. A minute demyelinating lesion causing acute positional vertigo. *J Neurol Sci*. 2008 Mar 15;266(1-2):187-9.

**Keywords:** Nystagmus, Demyelinating Disease, Ocular Manifestations of Vestibular Disorders

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Normative Database for the King-Devick Test in Adults and Adolescents

Danielle Leong<sup>1,2,3</sup>, Yi Pang<sup>4</sup>, Leonard V. Messner<sup>4</sup>, Sherry Audycki<sup>5</sup>, James Fanelli<sup>6</sup>, Dan McGehee<sup>7</sup>, Bob Steinmetz<sup>3</sup>, Wendy Stone<sup>4</sup>, Katherine Lynch<sup>4</sup>, Heather E. Moss<sup>8</sup>, Laura J Balcer<sup>9</sup>

<sup>1</sup>King-Devick Test, LLC, Oakbrook Terrace, IL, USA, <sup>2</sup>Salus University Graduate Department of Biomedicine, Philadelphia, PA, USA, <sup>3</sup>SoLo Eye Care, Chicago, IL, USA, <sup>4</sup>Illinois Eye Institute Chicago, IL, USA, <sup>5</sup>Advanced Eye Center Bedford, MA, USA, <sup>6</sup>Cape Fear Eye Institute Wilmington, NC, USA, <sup>7</sup>Swagel Wootton Hiatt Eye Center Mesa, AZ, USA, <sup>8</sup>University of Illinois at Chicago School of Medicine, Department of Ophthalmology and Visual Sciences Chicago, IL, USA, <sup>9</sup>New York University School of Medicine, Department of Neurology New York, NY, USA

### Introduction:

The King-Devick(K-D) test, a <2-minute timed assessment of rapid number naming, has been studied as a rapid, quantitative screening tool for neurological dysfunction associated with concussion, hypoxia, Parkinson's disease, multiple sclerosis and extreme sleep deprivation. All studies to date have either compared within subjects against individual baselines or between affected subjects and age-matched neurologically-normal control subjects. A large normative database has not been previously established. The purpose of this study was to determine the distribution of K-D test performance in normal adults and adolescents and evaluate the effect of potential confounding variables.

### Methods:

In this cross-sectional, multi-center study, subjects  $\geq 15$  yrs old with binocular best-corrected near visual acuity better than 20/30 completed two trials of the K-D test protocol. Exclusion criteria included concussion within 3-months, post-concussion syndrome, dyslexia or neuro-degenerative disorders. History of concussion, amblyopia, strabismus as well as demographic variables of education, race/ethnicity, gender and age were assessed by subject interview. Analysis of Covariance was used to determine if age, education, or race affected performance while controlling other cofounders.

### Results:

Subjects (n=691, age:39.8 $\pm$ 17.7yrs,58%female) were enrolled in 5 sites. Best K-D times were 41.2 $\pm$ 8.2s. Difference between K-D trials was 3.1 $\pm$ 3.3s. Performance did not vary by history of concussion, gender, amblyopia, or strabismus. Younger age, higher education and Caucasian or Hispanic race/ethnicity had better K-D times( $p \leq 0.001$ ). K-D times were stable in the subjects aged  $\leq 39$  yrs and demonstrated worsening  $\geq 40$  yrs.

### Conclusions:

The K-D test requires visual processing, saccades, language, attention, and has been proposed as a marker of integrated neurological function. We report normative data in a large adolescent/adult cohort and report associations with age, education and race. Knowledge of these confounding variables is important for design and interpretation of future K-D studies. Our database will have application to future studies of K-D test performance in neurologically diseased populations and potential future application to clinical settings.

**References:** None.

**Keywords:** Diagnostic Tests, Ocular Motility

**Financial Disclosures:** Danielle Leong is employed by King-Devick Test, LLC as the Director of Research.

**Grant Support:** Illinois Society for the Prevention of Blindness Research Grant

## Poster 140

### Marcus Gunn Jaw Winking with Electronegative Cone-Rod Dystrophy: Case Report

Su Ann Lim<sup>1</sup>

*Tan Tock Seng Hospital, Singapore, Singapore*

#### **Introduction:**

Marcus Gunn jaw winking syndrome (MGJW) is due to aberrant innervation of the levator palpebrae superioris muscle by the trigeminal nerve. This has been reported with other ocular abnormalities like monocular elevation deficiency and Duane's syndrome<sup>1</sup>. We report a case associated with electronegative cone-rod dystrophy. To the best of my knowledge, this is the first report of MGJW with a retinal dystrophy. This report expands our knowledge of ocular disorders associated with MGJW and may assist in the understanding of the pathophysiology of this condition.

#### **Methods:**

Case Report

#### **Results:**

An Iranian boy was born with poor vision, nystagmus, colorblindness and MGJW. He was the product of consanguineous parents. His younger brother was normal and there was no family history of visual problems. Visual acuity was 6/120 colour vision was absent. He had fine vertical nystagmus and his retina appeared normal. Electroretinogram (ERG) revealed electronegative cone-rod dystrophy.

#### **Conclusions:**

The exact mechanism of MGJW and electronegative cone-rod dystrophy have yet to be elucidated. Electronegative ERG has been attributed to dysfunction of the ON-bipolar pathway<sup>2</sup>. Recently this has been reported in Phosphomannomutase Deficiency, a congenital disorder of glycosylation. MGJW has been associated with the cranial dysinnervation syndromes. Genetic studies in to these disorders have enabled us to understand some aspects of the developing brain<sup>4</sup>. These two conditions coexisting in the same individual may further expand our understanding of these conditions.

#### **References:**

1. Shah AD, Kumar AB, Kothari K. Bilateral Marcus Gunn Jaw winking synkineses with monocular elevation deficiency: a case report and literature review. *Int Ophthalmol* 2012;32:199-201.
2. Renner AB, Kellner U, Cropp Elke et al. Dysfunction of transmission in the inner retina: incidence and clinical causes of negative electroretinogram. *Graefes Arch Clin Exp Ophthalmol* 2006;244:1467-1473.
3. Thompson DA, Lyons RJ, Liasis A, et al. Retinal On-Pathway Defect in Congenital Disorder of Glycosylation Due to Phosphomannomutase Deficiency. *Arch Ophthalmol* 2012 ;130(6) :712-19.
4. Assaf AA. Congenital innervation dysgenesis syndrome (CID)/ congenital cranial dysinnervation disorders (CCSSs). *Eye* 2011 ;25 :1251-1261.

**Keywords:** Nystagmus, Cone-Rod Dystrophy, Marcus Gunn Jaw Winking, Ptosis, Genetics

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 141

### Levodopa-Induced Ocular Dyskinesias

Bryan V Pham<sup>1</sup>, John G Nutt<sup>1</sup>, William L Hills<sup>1,2</sup>

<sup>1</sup>Department of Neurology, Oregon Health & Science University, Portland, OR, USA, <sup>2</sup>Casey Eye Institute, Department of Ophthalmology, Oregon Health & Science University, Portland, OR, USA

#### Introduction:

Levodopa-induced dyskinesias (LID) are commonly encountered in advanced Parkinson's disease (PD), and classically affect the limbs, trunk, and head. We present a patient with advanced PD who developed severe, disabling LID, with involvement of his oculomotor system.

#### Methods:

Case report.

#### Results:

A 62 year-old man with left-dominant PD diagnosed at age 37, and continuously on carbidopa-levodopa since age 38, developed dyskinesias of the head which were first documented at age 44, and were followed over the next several years by disabling foot cramping, athetoid/dystonic hand movements, facial grimacing, and prominent retrocollis. At age 48, he demonstrated abnormal involuntary eye movements (AIEMs), characterized by difficulty with visual fixation. Current examination found intermittent upward and rightward gaze deviation, however, he was able to make contra-verse voluntary saccades after prolonged saccadic latency. His AIEMs occur exclusively during ON periods, and are worsened by periods of emotional and physical stress. Brain MRI revealed no lesions accountable for his AIEMs.

#### Conclusions:

One recent review identified 5 from 32 advanced PD patients with AIEMs occurring exclusively in the ON state. AIEMs were characterized by repeated, stereotyped, upward and/or lateral gaze deviation, sometimes phasic, brief and jerky, sometimes tonic and sustained for several seconds. Gaze deviation typically was to the side more affected by Parkinsonism (in 4/5 patients). AIEMs are modulated by facilitation and inhibitory maneuvers as in other LID, e.g., temporarily suppressed by will and visual fixation and worsened by emotional and physical stress. The severity of AIEMs does not necessarily correlate with the severity of other concomitant LID. AIEMs never occur in isolation, and the onset is delayed by years with respect to the onset of other LID, suggesting that extensive dopaminergic denervation in the caudate nucleus may be a prerequisite for AIEMs, yet the exact pathophysiology remains elusive.

#### References:

1. Grotzsch H, Sztajzel R, Burkhard PR. "Levodopa-induced ocular dyskinesia in Parkinson's disease." *European Journal of Neurology*. 14:1124-1128. 2007.
2. Linazasoro G, Van Blercom N, Lasa A, "Levodopa-induced ocular dyskinesias in Parkinson's disease." *Movement Disorders*. 17:186-220. 2002.

**Keywords:** Levodopa-Induced Dyskinesias, Abnormal Involuntary Eye Movements

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Clinical Features, Diagnostic Findings and Treatment of Adult-Onset Opsoclonus-Myoclonus Syndrome: A Case Series**

Olga Rosenthal<sup>1,2</sup>, Pooja Raibagkar<sup>1,2</sup>, Shamik Bhattacharyya<sup>1,2</sup>, Ivana Vodopivec<sup>1</sup>, Shirley H Wray<sup>1</sup>

<sup>1</sup>Massachusetts General Hospital, Boston, MA, USA, <sup>2</sup>Brigham and Women's Hospital, Boston, MA, USA

**Introduction:**

OMS consists of involuntary saccades with myoclonus, cerebellar ataxia, and/or encephalopathy. Saccadic movements are termed opsoclonus when multidirectional and ocular flutter when in horizontal plane. OMS is thought to be caused by immune-mediated injury to either pontine omnipause cells which inhibit gaze center burst neurons or cerebellar hemispheres which disinhibit the fastigial nucleus. Our goal is to discuss clinical features, diagnostic findings, and treatment of adult-onset opsoclonus-myoclonus syndrome (OMS).

**Methods:**

Retrospective review of six adults with OMS (ages 24-62 years, 3 women), diagnosed and treated at two tertiary care centers from 2011 to 2014. Clinical, laboratory, and imaging findings, including brain magnetic resonance imaging (MRI) and [<sup>18</sup>F]fluoro-2-deoxyglucose positron emission tomography (FDG-PET), were analyzed.

**Results:**

Ocular flutter and ataxia were present in all, while opsoclonus was found in only two patients. Myoclonus and encephalopathy were present in four and two patients, respectively. Infectious pathogen was not found in anyone. CSF analysis showed lymphocytic pleocytosis in five patients. Neoplasms were diagnosed in three: one benign ovarian dermoid tumor and two malignancies – uterine carcinosarcoma and neuroblastoma. Neural-specific autoantibodies were detected in four patients: one with anti-Ri related to uterine-carcinosarcoma, one with anti-GQ1b, and two with anti-GAD65 (one associated with ovarian dermoid tumor). MRI showed T2 hyperintensity in inferior right cerebellum in one patient. FDG-PET of brain showed cerebellar hypometabolism in three (two bilateral and one unilateral). One of two patients with malignant neoplasms died (both treated with immunotherapy). In the three nonparaneoplastic cases, clinical progression occurred with intravenous corticosteroids until intravenous immunoglobulin (IVIg) was administered.

**Conclusions:**

In adults, ocular flutter and ataxia, rather than opsoclonus or myoclonus, are the characteristic clinical features. The syndrome is associated with diverse antibodies, which may be autoimmune or paraneoplastic. Selective cerebellar hypometabolism in three patients supports the hypothesis of cerebellar hemispheric dysfunction. Patients without malignancies may achieve complete remission with IVIg.

**References:** None.

**Keywords:** Opsoclonus-Myoclonus, Paraneoplastic

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 143

### Divergence Palsy due to Antiepileptic Drugs

Marc A. Bouffard<sup>1</sup>, Louis R. Caplan<sup>1</sup>, Nurhan Torun<sup>2</sup>

<sup>1</sup>Beth Israel Deaconess Medical Center, Department of Neurology, Boston, MA, USA, <sup>2</sup>Beth Israel Deaconess Medical Center, Division of Ophthalmology, Department of Surgery, Boston, MA, USA

#### Introduction:

Diplopia is a potential side effect of many anti-epileptic drugs (AEDs). However, it is not always possible to examine the patient while they are symptomatic and thus the characteristics and the specific mechanisms of anti-epileptic drug-induced diplopia have not been well described.

#### Methods:

We describe two patients who had spells characterized by divergence palsy while receiving anticonvulsants.

#### Results:

We describe two cases of AED-associated diplopia, one attributable to oxcarbazepine and the other to divalproex. Both patients were found to have a divergence palsy only during their spells. In both cases, removal of the offending agent resulted in prompt resolution of symptom which has not recurred over the past 6 and 2 years, respectively. In our second case, divalproex was restarted to confirm our hypothesis of a causal relationship and the spells recurred within days only to resolve again after divalproex cessation.

#### Conclusions:

While diplopia is listed as a side effect of many AEDs, a literature search did not identify any detailed description of an examination during an episode of diplopia. To our knowledge, this is the first report where an association between antiepileptics and divergence palsy was confirmed by a rechallenge. Both patients underwent extensive testing for their episodes of diplopia before being diagnosed with episodic divergence palsy. Awareness of the association between antiepileptics and divergence palsy may change management of patients presenting with similar episodes, especially if patients can be examined while they are symptomatic. It is important to consider iatrogenic causes when patients present with intermittent diplopia.

#### References:

1. Arai M, Fujii S. Divergence paralysis associated with the ingestion of diazepam. J Neurol.1990; 237(1):45-6.
2. Remler BF, Leigh RJ, Osorio I, Tomsak RL. The characteristics and mechanisms of visual disturbance associated with anticonvulsant therapy. Neurology 1990; 40:791-796.

**Keywords:** Divergence Palsy, Diplopia, Antiepileptic Drugs, Ocular Motility

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Quantifying The Vestibulo-Ocular Reflex With Video-Oculography: Nature And Frequency Of Artifacts

Ali S Saber Tehrani<sup>1</sup>, Georgios Mantokoudis<sup>2</sup>, Jorge C Kattah<sup>1</sup>, Karin Eibenberger<sup>3</sup>, Cynthia I Guede<sup>1</sup>, David S Zee<sup>2</sup>, David E Newman-Toker<sup>2</sup>

<sup>1</sup>University of Illinois College of Medicine, Department of Neurology, Peoria, IL, USA, <sup>2</sup>Johns Hopkins University School of Medicine, Department of Neurology, Baltimore, MD, USA, <sup>3</sup>Johns Hopkins University School of Medicine, Department of Otolaryngology – Head and Neck Surgery, Baltimore, MD, USA

### Introduction:

Little is known about the impact of disruptive phenomena (corrective saccades, nystagmus, fixation losses, eye-blinks) on quantitative vestibulo-ocular reflex (VOR) assessment. We sought to characterize the frequency, nature, and impact of abnormal physiologic findings and artifacts on quantitative head impulse test (HIT) VOR measures.

### Methods:

From a prospective study of video-oculography (VOG) in 26 patients with acute vestibular syndrome (16 vestibular neuritis, 10 stroke), we classified 1358 HIT traces using a structured coding manual. All disruptive findings were coded based on morphologic similarity to known physiologic patterns or artifact type. HIT traces were classified by a single, masked rater. A second, independent rater re-coded a 10% subsample to assess inter-rater reliability. Outcomes were presence and type of intrusive eye movement or artifact, whether the intrusion rendered the trace difficult to interpret, and whether disruptive phenomena varied by underlying disease. We report descriptive statistics, Cohen's kappa (inter-rater reliability), and Chi2 p-values (comparisons across diseases).

### Results:

Of all HIT traces, 72% had abnormal (but pathophysiologically-appropriate) intrusive saccades, 44% of traces had at least one artifact, and 42% of traces were deemed uninterpretable. The most common intrusions limiting VOR interpretation were fast-phase eye movements (saccades or nystagmus) occurring during VOR response. Inter-rater agreement on attributes varied (0.27-0.79) but was excellent for presence of a normal VOR (0.78) and very good for the presence of one or more artifacts (0.64) or an uninterpretable trace (0.66). Abnormal head impulses in patients with vestibular neuritis were more susceptible to artifacts (57% of ipsilesional neuritis traces uninterpretable vs. 31-34% of contralesional neuritis traces and stroke traces, Chi2  $p < 0.001$ ).

### Conclusions:

Physicians using quantitative recording devices to measure VOR responses to head impulses for clinical diagnosis should be aware of the potential impact of intrusive eye movements and measurement artifacts, especially in patients with an acutely abnormal HIT.

**References:** None.

**Keywords:** Eye Movement Measurements, Artifacts, Vertigo, Vestibular Neuritis, Stroke

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Neuroophthalmic Manifestations Of Intracranial Tumours In South India

Kowsalya Akkayasamy<sup>1</sup>, Dhivya Rajadurai<sup>1</sup>, Mahesh Kumar<sup>1</sup>

<sup>1</sup>Aravind eye Care System, Madurai, India, <sup>2</sup>Aravind eye Care System, Madurai, India, <sup>3</sup>Aravind Eye Care System, Madurai, India

### Introduction:

To study the clinical profile of patients with intracranial tumours presenting to the NeuroOphthalmology department and to correlate the ocular manifestations and site of the intracranial tumour.

### Methods:

A prospective study of 192 consecutive patients including paediatric age group, who were proven to have Intracranial tumour clinically and radiologically from June 2011 to June 2013 for a period of 2 years, who presented to the Department of Neuro-Ophthalmology in a tertiary eye care centre in South India was conducted and followed up for 3 months after treatment. All patients underwent thorough ophthalmological and neurological evaluation.

### Results:

Of 192 patients, 104(54.2%) were men and 88(45.8%) women. Mean age was 41.97 years, ranging from 3 to 75 years. 156 patients(81.3%) had defective vision at presentation, 25% headache, 4.7% double vision. Bitemporal hemianopia was the commonest field defect followed by generalized constriction. Fundus examination showed papilledema in 17.2%, Temporal pallor in 34.9%, primary optic atrophy in 27.6%. Sellar and suprasellar tumours presented with normal fundus or segmental, primary optic atrophy. Posterior fossa tumours presented with papilledema. Abducent nerve was paralysed in 11.5%, oculomotor nerve in 7.8%, Vestibulocochlear nerve in 8.3%. There was a statistically significant association between best corrected visual acuity at presentation and follow up. ( $p < 0.001$ ). Thus the visual prognosis depends upon the vision at presentation. There was a significant association between type of tumour and age group ( $p < 0.001$ ), but no association between gender and type of tumour ( $p = 0.096$ ). Pituitary adenomas (46.9%), Meningiomas (24%), Acoustic neuroma (8.3%) were the commonest tumours in adults and medulloblastoma in children.

### Conclusions:

A simple case of double vision (32 patients in our study), might have an underlying brain tumour. One with a normal fundus (16.1%) can harbour a sellar, suprasellar tumour. So when a patient presents with ocular complaints, careful clinical examination, if it arises a suspicion of brain tumour, has to be imaged immediately, which can reveal a life or vision threatening brain tumour. So, a neuroophthalmologist can save the life and vision of the patient by detecting it at an early stage.

**References:** None.

**Keywords:** Intracranial Tumour, Pituitary Macroadenoma, Headache, Papilledema, Optic Atrophy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Sideline Testing in Youth and Collegiate Athletes: What Does Vision Add to the Concussion Puzzle?

Laura J. Balcer<sup>1</sup>, Avri Bohm<sup>1</sup>, Lisena Hasanaj<sup>1</sup>, John-Ross Rizzo<sup>1</sup>, Liliana Serrano<sup>1</sup>, Rachel Nolan<sup>1</sup>, Nicholas Moehringer<sup>1</sup>, Nikki Webb<sup>2</sup>, Courtney Civitano<sup>3</sup>, Dennis Cardone<sup>1</sup>, Arlene Silverio<sup>4</sup>, Steven L. Galetta<sup>1</sup>

<sup>1</sup>New York University School of Medicine, New York, NY, USA, <sup>2</sup>New York University Athletics, New York, NY, USA, <sup>3</sup>Long Island University Athletics, New York, NY, USA, <sup>4</sup>Hofstra North Shore LIJ School of Medicine New York, NY, USA

### Introduction:

Identification of rapid yet simple diagnostic tests for sports-related concussion is critical. These tests must be age-appropriate and capture relevant aspects of brain function. Vision encompasses nearly 50% of the brain's pathways, yet is not represented in current sideline testing protocols. We examined the King-Devick (K-D) test, a vision-based measure of rapid number naming, as a complement to the Sport Concussion Assessment Tool, 3<sup>rd</sup> Edition (SCAT3/Child-SCAT3) in youth and collegiate athletes.

### Methods:

Members of a youth ice hockey and football league and collegiate athletes from NYU and Long Island University (LIU) participated in a prospective study to examine sideline tests, including the K-D test, Standardized Assessment of Concussion, (SAC, cognitive component of SCAT3/Child-SCAT3) and timed-tandem-gait test (balance component of SCAT3/Child-SCAT3).

### Results:

Youth and collegiate athletes (n=342, age 14.7±4.9 years, range 5-23) underwent pre-season baseline testing during the fall of 2014. Higher (worse) time scores for the vision-based K-D test were associated with lower (worse) scores for the SAC, particularly for the Orientation (p=0.003) and Immediate Memory components (p=0.01, linear regression accounting for age). K-D scores correlated significantly yet moderately with scores for balance (p=0.003). Athlete age was most strongly correlated with K-D scores (r= -0.79, p<0.0001) compared to SAC (r= 0.51, p<0.0001) and timed-tandem (r= -0.49, p<0.0001).

### Conclusions:

The K-D test, a vision-based measure that requires saccades and other eye movements, captures aspects of brain function that are not entirely captured by current sideline testing for balance and cognition. At the same time, associations of K-D scores with working memory and orientation suggest that closely related anatomical structures, such as the dorsolateral prefrontal cortex, underlie saccades and memory. K-D scores showed the greatest associations with age in this young athlete cohort, suggesting that vision may be a key domain for assessing developmental aspects of brain function.

### References:

1. Ventura RE, Galetta SL, Balcer LJ. The neuro-ophthalmology of head trauma. *Lancet Neurol* 2014;13:1006-1013.
2. Marinides Z, Galetta KM, Andrews CN, Wilson JA, Herman DC, Robinson CD, Smith MS, Bentley BC, Galetta SL, Balcer LJ, Clugston JR. Vision testing is additive to the sideline assessment of sports-related concussion. *Neurol Clin Pract* 2014, in press.
3. Leong DF, Balcer LJ, Galetta SL, Liu Z, Master CL. The King-Devick test as a concussion screening tool administered by sports parents. *J Sports Med Phys Fitness* 2014;54:70-77.
4. Galetta MS, Galetta KM, McCrossin J, Wilson JA, Moster S, Galetta SL, Balcer LJ, Dorshimer GW, Master CL. Saccades and memory: baseline associations of the King-Devick and SCAT2 SAC tests in professional ice hockey players. *J Neurol Sci* 2013;328:28-31.
5. Galetta KM, Barrett J, Allen M, Madda F, Delicata D, Tennant AT, Branas CC, Maguire MG, Messner LV, Devick S, Galetta SL, Balcer LJ. The King-Devick test as a determinant of head trauma and concussion in boxers and MMA fighters. *Neurology* 2011;76:1456-72.

**Keywords:** Vision, Concussion, King-Devick (K-D) Test, Eye Movements

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Assessment of Recruitment Patterns in a Neuro-Ophthalmology Registry

Kimberly D Blankshain<sup>1</sup>, Hyo Jin Park<sup>2</sup>, Heather Moss<sup>2</sup>

<sup>1</sup>Rosalind Franklin University of Medicine and Science - Chicago Medical School, North Chicago, IL, USA, <sup>2</sup>UIC College of Medicine - Ophthalmology, Chicago, IL, USA

### Introduction:

A challenge when studying rare diseases is adequate subject recruitment. Prospective research registries are a useful tool for studying rare diseases by forming observational cohorts with which to study disease(1) and serving as a source of subject recruitment for future studies(2). We developed a neuro-ophthalmic patient registry in June, 2014. The purpose of this study is to critically assess recruitment patterns three months after registry implementation.

### Methods:

An IRB approved neuro-ophthalmology patient registry was implemented using REDcap(3). Inclusion criteria were age  $\geq 21$  years and ability to provide informed consent in English. All patients attending outpatient neuro-ophthalmology appointments were screened for inclusion. Age, gender, race, ethnicity, eligibility and enrollment were recorded for all patients. Demographic variables were compared between registry participants and non-participants during the first three months following implementation.

### Results:

182 patients were screened. 29(15%) and 16(8%) were ineligible due to age and inability to provide informed consent, respectively. 23(13%) declined to participate. 114 patients enrolled (age 49+/-16 years, 65% female, 14% Hispanic ethnicity, 40% black race, 34% white race). Patients excluded due to inability to provide consent were more likely to be older (60+/-20 years,  $p=0.01$  t-test) or of Hispanic ethnicity (69%,  $p<0.0005$ , chi square) than enrolled patients. Patients who declined to participate did not differ from enrolled patients by age, gender, ethnicity or race. .

### Conclusions:

75% of adult patients were recruited into a new neuro-ophthalmology registry over three months. This registry forms a foundation for advancing human neuro-ophthalmic research at our institution by establishing an observational cohort and serving as a source from which to recruit subjects for future studies.. The consent requirements disproportionately excluded subjects who were older or of Hispanic ethnicity. Modifying the consent process to include non-English speaking patients will be important to achieve an adequate population sample and avoid recruitment bias.

### References:

1. Fischer, Ljung, Piatokouki, Liesner, Claeysens, Smink, van den Berg, "Prospective observational cohort studies for studying rare diseases: the European PedNet Haemophilia Registry" Haemophilia 20(4) e280-6. 2014.
2. Malek, Stickler, Antao, Horton "The national ALS registry: a recruitment tool for research. Muscle Nerve Aug 11, 2014 ePub ahead of print
3. Harris, Taylor, Thielke, Payne, Gonzalez, Conde. Research electronic data capture (REDCap) – A metadata-driven methodology and workflow process for providing translational research informatics support. J. Biomed Inform. 2009 42(2): 377-81

**Keywords:** Registry, Observational Cohort, Neuro-Ophthalmic Diseases

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** NIH grants K12 EY 021475, K23 EY 024345, P30 EY 001792, UL1RR029879 An unrestricted departmental grant from Research to Prevent Blindness

## Poster 148

### Prediction of Eye Position During General Anesthesia Using Bispectral Index Monitoring

Seung Ah Chung<sup>1</sup>, Kyoung Hoon Kook<sup>1</sup>, Jong Bok Lee<sup>2</sup>

<sup>1</sup>Department of Ophthalmology, Ajou University School of Medicine, Suwon, South Korea, <sup>2</sup>Department of Ophthalmology, Yonsei University College of Medicine, Seoul, South Korea

#### Introduction:

The position of the eyes in general anesthesia is generally considered one of slight divergence and elevation, yet it has not been well characterized to date. We aimed to quantify eye position related to the depth of anesthesia using bispectral index (BIS), a derived electroencephalograph parameter.

#### Methods:

In this prospective blinded study, we investigated the relationship between BIS, hemodynamic parameters during anesthesia and eye positions in 32 healthy children, mean age 5.4 years, who were undergoing surgical correction for epiblepharon using a standardized anesthetic technique with sevoflurane. BIS and hemodynamic parameters were continuously recorded throughout the procedure: from induction to awakening. Eye positions were video-recorded during surgery and analyzed after surgery at one-minute intervals by the examiner blinded to results of BIS.

#### Results:

There was significant negative correlation between BIS and end-tidal sevoflurane concentrations ( $p < 0.001$ ), and positive correlation between BIS and elevation of the eyes (eye position =  $0.014 \times \text{BIS} + 0.699$ ,  $p = 0.011$ ). Elevation of the eyes (83%) mostly occurred when BIS values reached over 65, while down-shoot eye movement (2%) was seen at BIS values less than 35.

#### Conclusions:

A shallower level of anesthesia, indicated by a higher BIS values was associated with a higher position of the eyes, suggesting that BIS monitoring may be beneficial in predicting eye position. In ophthalmic surgery with particular attention to eye position, the anesthesia depth is recommended to be maintained by monitoring BIS values in range of 35-65.

**References:** None.

**Keywords:** Diagnostic Tests, Ocular Motility

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Poster 149**

**Demographic Profile Of The Patients Presenting To A Neuro-Ophthalmology Clinic Of A Tertiary Eye Care Centre**

Shiva Prasad Gantyal<sup>1</sup>, Rebika Dhiman, Vaitheeswaran Lalgud Ganesan, Digvijay Singh, Rohit Saxena, Pradeep Sharma

*All India Institute Of Medical Sciences, New Delhi, In, India*

**Introduction:**

To evaluate the clinical profile of neuro-ophthalmic disease in patients presenting to Neuro-ophthalmology clinic of a tertiary care centre during one year.

**Methods:**

A cross-sectional hospital-based observational study was done at a tertiary level eye care centre during one year. All patients with neuro-ophthalmic disease were referred to the neuro-ophthalmic clinic where they were evaluated with a detailed clinical history and examination.

**Results:**

A total of 30111 patients were evaluated of which 1597 (5%) were referred to neuro-ophthalmic clinic. The mean age of the referred patients was 31 years ( $31 \pm 19.53$ ) with male predominance. Among 1597 patients, optic nerve disorders were noted in 63.8% (n=1020), cranial nerve palsy in 7% (n=114), cortical visual impairment in 6.5% (n=105) and others (disc hypoplasia, blepharospasm, optic nerve head drusen etc) were 6% (n=96). Among the patients with optic nerve disorders, primary optic neuropathy (traumatic optic neuropathy, hereditary, tumor related, retro bulbar neuritis, toxic neuropathy & idiopathic) was noted in 42.8% (n=685) and secondary optic neuropathy (Ischemic optic neuropathy, post papilledema optic neuropathy, post-papillitis, neuroretinitis, inflammatory optic neuropathy) were 20.9 % (n=335). 16.4% (n=263) patients were wrongly referrals.

**Conclusions:**

Neuro-ophthalmic clinic constitutes a significant referral unit in a tertiary eye care centre. Traumatic and Ischemic optic neuropathies are common and most important preventable forms of neuro-ophthalmic diseases.

**References:** None.

**Keywords:** Demography, Optic Neuropathy, Cortical Blindness, Trauma, Ischemia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## **Botulinum Toxin-Augmented Strabismus Surgery versus Conventional Surgery in the Treatment of Large-Angle Infantile Esotropia**

Aubrey L. Gilbert<sup>1</sup>, Michael J. Wan, Melanie Kazlas, Carolyn Wu, David G. Hunter, Iason Mantagos, Ankoor S. Shah

*Departments of Ophthalmology, Boston Children's Hospital and Harvard Medical School, Boston, MA, USA*

### **Introduction:**

The treatment of large-angle infantile esotropia remains a challenge. Surgical treatment traditionally has involved either three-muscle surgery or supra-maximal medial rectus recessions, but both methods have significant drawbacks including risk of over- or undercorrection. An alternative is to use botulinum toxin to weaken the medial rectus muscles, but multiple treatments are often required and the success rate for large deviations is poor. It has been suggested that the best approach may be to combine these treatments. No previous study has directly compared botulinum toxin-augmented surgery to conventional surgery alone.

### **Methods:**

A retrospective, comparative study was completed evaluating baseline characteristics and outcomes of consecutive patients with large-angle infantile esotropia (55 prism diopters( $\Delta$ ) or greater) who underwent either conventional or botulinum toxin-augmented strabismus surgery. The primary outcome measure was post-op deviation and need for retreatment within one year of initial intervention. Success was defined as a deviation  $<10\Delta$  and no retreatment at 1 year after the initial intervention.

### **Results:**

The record review identified 38 patients meeting inclusion criteria, of whom 14 had been treated with botulinum toxin-augmented surgery and 24 with bilateral medial rectus recessions. Prior to surgery, the augmented group had a larger median angle of deviation (65 versus  $60\Delta$ ,  $p = 0.006$ ) and less recession of the medial rectus muscles was performed (5.5 versus 6.0 mm,  $p = 0.005$ ). The augmented group had greater success at 1 year (70% versus 30%,  $p = 0.035$ ). The augmented group also had a significantly smaller median angle of deviation at 1 year (0 versus  $18\Delta$ ,  $p = 0.026$ ). There were no serious complications in either group.

### **Conclusions:**

Botulinum toxin can significantly augment the effect of medial rectus recessions and improve the success of strabismus surgery in patients with large-angle infantile esotropia.

**References:** None.

**Keywords:** Pediatric Neuro-Ophthalmology, Botulinum Toxin, Strabismus, Infantile Esotropia, Bimedial Recession

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 151

### Heidenhain Variant Of Creutzfeldt Jakob's Disease (CJD), In A Patient Who Had Bovine Bioprosthetic Valve Implantation

Haneen Jabaly-Habib<sup>1</sup>, Waleed Saliba<sup>2</sup>, Mazin Elias<sup>2</sup>, George Habib<sup>3</sup>

<sup>1</sup>Ophthalmology Unit, Poria Hospital, Poria, Israel, <sup>2</sup>Internal Medicine Dept, Emek medical Center, Afula, Israel, <sup>3</sup>Internal Medicine Dept, Carmel Hospital, Haifa, Israel

#### Introduction:

82year old woman, referred to neuroophthalmology clinic for unexplained visual loss. One month earlier she started to complain about blurred vision that deteriorated significantly in the last week. The patient and her son denied any behavioral changes or memory impairment. Her past medical history included: hypertension, hypercholesterolemia, peptic disease, and aortic valve replacement using a bovine bioprosthetic valve- 19-mm carpentier-Edwards Perimount

#### Methods:

On examination: Full ocular movements, visual acuity finger count in both eyes, normal anterior segment, RAPD negative, and normal fundus examination. Humphrey visual fields demonstrated complete absolute right homonymous hemianopsia. In her MRI review, performed two weeks earlier, there were nonspecific white matter changes. Extended evaluation was recommended

#### Results:

One week later the patient was admitted due to memory impairment "loosing words" and "unable to complete sentences". The repeated MRI showed no new pathology, total body CT was normal, EEG was compatible with CJD, cerebrospinal fluid was normal but TAU was extremely elevated. Patient developed rapid loss of short and long term memory, disorientation, incontinence, brisk reflexes. She eventually was discharged to hospice facility and died three weeks later. Unfortunately autopsy was not performed

#### Conclusions:

We believe our patient had CJD-the heidenhain variant that first presents with visual disturbances. Ophthalmologists being first to see these patients should be aware of this diagnosis possibility especially when there is a history of major surgery or tissue transplantation. In literature review, no reports of iatrogenic transmission of CJD through bovine valves were found

**References:** None.

**Keywords:** Neuro-Ophth & Infectious Disease (Eg, AIDS, Prion)

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## The Effects of Pediatric Primary Brain Tumors on Vision and Quality of Life

Supharat Jariyakosol<sup>1,2</sup>, Jason H. Peragallo<sup>1,3</sup>, Beau B. Bruce<sup>1,4,5</sup>, Nancy J. Newman<sup>1,4,6</sup>, Valerie Biousse<sup>1,4</sup>

<sup>1</sup>Department of Ophthalmology, Emory University School of Medicine, Atlanta, GA, USA, <sup>2</sup>Department of Ophthalmology, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand, <sup>3</sup>Department of Pediatrics, Emory University School of Medicine, Atlanta, GA, USA, <sup>4</sup>Department of Neurology, Emory University School of Medicine Atlanta, GA, USA, <sup>5</sup>Department of Epidemiology, Emory University Atlanta, GA, USA, <sup>6</sup>Department of Neurological Surgery, Emory University School of Medicine Atlanta, GA, USA

### Introduction:

Brain tumors are the leading cause of death from childhood cancer. Overall survival has improved due to earlier detection, better therapies, and improved surveillance. Permanent sequelae of the tumor and its treatment may cause severe impairment and decreased quality of life (QOL). Visual dysfunction and impaired vision-related QOL (VR-QOL) are often not recognized in children because of examination difficulty and lack of awareness. We reviewed the literature and evaluated visual impairment and its effects on QOL in an ongoing quality improvement project.

### Methods:

Patients  $\leq 18$  yo,  $\geq 6$  months from diagnosis of primary brain tumor (PBT), excluding primary intrinsic anterior visual pathway tumors, underwent neuro-ophthalmologic examination. Health-related QOL (HR-QOL) questionnaires, using PedsQL Brain Tumor Module,<sup>1</sup> were obtained from patients and parents. VR-QOL questionnaires, using CVFQ (Children's visual function questionnaire)<sup>2</sup> in children  $< 8$ yo, and EYE-Q<sup>3</sup> in children 8-18yo, were obtained. Demographic data, driving status, schooling, and use of low-vision aids were recorded. We reviewed recent studies (2000-2014) of ophthalmologic sequelae, long-term disability, and HR-QOL in pediatric PBT patients.

### Results:

26 patients were evaluated (18 supratentorial, 8 infratentorial tumors). 3/26 patients (11.54%) were legally blind (primarily suprasellar tumors); 12/26(46.15%) had visual impairment; 13/26(50%) had significant visual field defects. 15/26(57.69%) patients had strabismus, cranial nerve palsies, or nystagmus. HR-QOL median score was 68.6(range 28.1-95.8). VR-QOL median score was 3.27(range 0.38-4.0). Twelve studies met our inclusion criteria for review. Prevalence of visual dysfunction ranged from 8% to 79%. 3/12 studies reported the rate of bilateral blindness (3.3% to 16%). Most HR-QOL evaluations showed significantly lower scores in patients compared to normal control/range. VR-QOL was not previously addressed in pediatric PBT studies.

### Conclusions:

Pediatric PBT patients' vision, HR-QOL, and VR-QOL are often severely affected, even when the PBT is considered "cured". Systematic neuro-ophthalmologic examinations in pediatric PBT patients may improve long-term visual outcomes and QOL through earlier interventions.

### References:

1. [www.pedsq.org/](http://www.pedsq.org/)
2. <http://www.retinafoundation.org/questionnaire.htm>
3. Angeles-Han ST, Griffin KW, Harrison MJ, Lehman TJ, Leong T, et al. Development of a vision-related quality of life instrument for children ages 8-18 years for use in juvenile idiopathic arthritis-associated uveitis. *Arthritis Care Res* (Hoboken). 2011 Sep;63(9):1254-61.

**Keywords:** Brain Tumor, Quality Of Life, Visual Loss

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York, and by NIH/NEI core grant P30-EY06360 (Department of Ophthalmology). Dr. Jariyakosol receives support from Chulalongkorn University (Bangkok, Thailand). Dr. Bruce receives research support from the NIH/NEI (K23-EY019341).

## Poster 153

### A Method for Quantifying Off Chart Visual Acuities

Rustum Karanjia<sup>1</sup>, Tiffany J. Hwang<sup>2</sup>, Alexander F. Chen<sup>2</sup>, Matin Khoshnevis<sup>3</sup>, Edward R. Chu<sup>1</sup>, Alfredo A. Sadun<sup>1</sup>

<sup>1</sup>*Doheny Eye Centers, University of California at Los Angeles, Pasadena, CA, USA*, <sup>2</sup>*Keck School of Medicine, Los Angeles, CA, USA*, <sup>3</sup>*University of Los Angeles at Irvine, Irvine, CA, USA*

#### Introduction:

Retrospective chart reviews can provide data to test and generate hypotheses, however, they are limited to the data collected, which in some instances is not sufficiently quantitative. Visual acuity for instance would ideally be collected by ETDRS methods for prospective clinical trials. This is often not the case with retrospective studies, where off chart visual acuities (typically <20/400) are recorded as count fingers (CF) at a set distance, hand motion or light perception. The purpose of this study was to determine by geometric optics the Snellen equivalent visual acuities for count fingers at different distances.

#### Methods:

A random sampling of the finger size and inter-digit distance of dominant hand of 50 males and 50 females was collected from volunteers in an academic center. The average finger size was calculated and the mean total size of the index, middle and ring finger was determined. This distance was used to determine a decimal acuity by calculating the numbers of minutes of arc subtended by 3 fingers, which would represent the letter E. This was converted to Snellen equivalent (x) as follows:  $20/x = 8.75 \text{ mm/y}$  (y=3 fingers with spaces).

#### Results:

The average size of the index (15.9±1.5mm male 13.8±1.9mm female), middle (16.2±1.3mm male 14.2±2.0mm female) and ring finger (15.1±1.2mm male 13.3±2.2mm female) was used with the average inter-digit distance (21.8±7.2 male, 19.9±6.1 female) to calculate the Snellen equivalent acuity for count finger distances from 1 to 6 feet. For example CF@6ft = 0.029, 20/560 for males. Similar data was generated for females.

#### Conclusions:

It is possible to generate an approximate Snellen equivalent visual acuity for count finger acuities using geometric optics. A website was developed to allow other researchers to input their own finger widths and determine a custom table for use in future retrospective chart reviews.

**References:** None.

**Keywords:** Diagnostic Testing

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 154

### Differential Functions Mediated By Melanopsin Assessed In Subjects With Healthy And Diseased Eyes

Aki Kawasaki<sup>1</sup>, Lorette Leon<sup>1</sup>, Sylvie Collomb<sup>1</sup>, Miriam Munch<sup>2</sup>

<sup>1</sup>University of Lausanne/Hopital Ophtalmique Jules Gonin, Lausanne, Switzerland, <sup>2</sup>Charite University, Institute of Physiology, Berlin, Germany

#### Introduction:

Two central monosynaptic tracts originating from melanopsin-expressing retinal ganglion cells (MGCs) are the retinotectal tract (pupil light reflex) and the retinohypothalamic tract (entrainment of circadian rhythm). This study aims to assess the function of these two non-visual, light-dependent systems in patients who have optic atrophy and visual loss.

#### Methods:

Patients with bilateral optic atrophy due to hereditary optic neuropathy (HON; n=11) or open-angle glaucoma (GL; n=11) underwent testing with chromatic pupillometry, light-induced suppression of nocturnal melatonin, visual analogue scale for subjective sleepiness and reaction time at night. Main outcome parameters were the post-illumination pupil response (PIPR) to a blue light stimulus (a marker for intrinsic MGC signalling through the retinotectal tract) and nocturnal melatonin suppression to acute light exposure (a marker of retinohypothalamic integrity). Secondary measures were subjective sleepiness and reaction times. Results were compared to age-matched controls (n=22).

#### Results:

The PIPR and nocturnal melatonin suppression were positively correlated in patients and controls ( $p < 0.05$ ). Patients (HON and GL) showed similar nocturnal melatonin suppression to controls. The PIPR was significantly reduced in the GL group but not HON group, compared to their controls ( $p < 0.05$ ). Light exposure at nighttime acutely reduced subjective sleepiness in patients with HON but not GL, when compared to controls. In addition, GL patients showed slower reaction times during and after nocturnal light exposure.

#### Conclusions:

Glaucomatous, but not hereditary, type of optic atrophy is associated with reduced acute light effects mediated mainly via MGCs. This is detected in the pupil but not hormonal marker of the melanopsin light input pathway. This may indicate differential involvement of MGCs subtypes or MGC photoreceptive sensitivity from certain types of ganglion cell pathology. In addition, such changes in MGC function can influence other cognitive functions.

**References:** None.

**Keywords:** Melanopsin Ganglion Cells, Melanopsin, Pupil, Melatonin, Circadian Rhythm

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Swiss National Science Foundation and Velux Foundation Switzerland

### Familial Papillitis And Macular Cystoid Edema: A Genetic Autoimmune Disorder?

Chiara La Morgia<sup>1,2</sup>, Piero Barboni<sup>3</sup>, Antonio P Ciardella<sup>4</sup>, Alfredo A Sadun<sup>5</sup>, Valerio Carelli<sup>1,2</sup>

<sup>1</sup>IRCCS Istituto delle Scienze Neurologiche di Bologna, Bologna, Italy, <sup>2</sup>Dipartimento di Scienze Biomediche e Neuromotorie, Università di Bologna, Bologna, Italy, <sup>3</sup>Studio Oculistico d'Azeglio, Bologna, Italy, <sup>4</sup>Ophthalmology Unit, Policlinico S.Orsola-Malpighi Bologna, Italy, <sup>5</sup>Ophthalmology, UCLA, Doheny Eye Institute Los Angeles, CA, USA

#### Introduction:

To describe the phenotype, the diagnostic challenge and the therapeutic options of a rare familial neuro-ophthalmological disorder, characterized by papillitis and macular cystoid edema.

#### Methods:

Two case reports are reported.

#### Results:

The proband is a 18-year-old female, who presented at 13 years with progressive loss of vision OS, with occasional eye pain. Ophthalmological evaluation at onset revealed visual acuities OD 20/20; OS 20/60, mild disc edema OD and moderate OS and cystoid macular edema (CME) OS. IV corticosteroids led to partial visual recovery OS to 20/60. After steroid withdrawal vision worsened OS to 20/200. Autoimmune panel including aquaporin4 antibodies was negative except for elevated immune-complexes and positive GM1 antibodies. Cerebrospinal fluid (CSF) examination demonstrated increased proteins and abnormal brain barrier index without oligoclonal bands. HLA revealed DRB1\*15 haplotype. Screening for sarcoidosis, lyme disease and tuberculosis was negative. LHON mutations were absent. Brain MRI failed to reveal demyelinating lesions. The patient was treated with Acetazolamide, oral steroids and three iv Immunoglobulin courses and adalimumab without benefit. Intravitreal triamcinolone injection was effective in resolving CME for 2 and 8 months respectively. The mother (59 years) presented at 18 years with bilateral progressive loss of vision with occasional eye pain and bilateral disc edema with CME, completely resolved after steroids. VA worsened after steroid discontinuation evolving to severe optic atrophy OD with persistent disc edema and CME OS. Autoimmune screening disclosed GM1 antibodies. CSF exam showed high protein levels (88 mg/dl) with abnormal brain barrier index.

#### Conclusions:

We here present two familial papillitis cases, associated with CME responsive to steroid treatment. The strict resemblance of the phenotype points to a genetic basis of the disorder. The positivity of GM1 antibodies, the elevated immune complexes levels, the HLA haplotype and the steroid response suggest an autoimmune disorder. Exome sequencing is ongoing.

**References:** None.

**Keywords:** Genetic Disease, Optic Neuropathy, Neuro-Ophth & Systemic Disease ( Eg. MS, MG, Thyroid), Retina

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

***New Daily Persistent Headache Triggered by Cataract Extraction?***

Julia E. Mallory<sup>1</sup>, James Lin, Jeffrey G. Odel

*Columbia University / Ophthalmology, New York, NY, USA*

**Introduction:**

New daily persistent headache (NDPH) was first described by Vanast (1986), who characterized it as a benign headache syndrome that typically remits without treatment.<sup>1</sup> Subsequently, however, it has been found to be a bilateral headache syndrome that is often persistent. Patients are able to identify the exact time their headache began.<sup>2</sup>

NDPH may be responsive to intravenous migraine medications, high dose corticosteroids, doxycycline, gabapentin, intravenous acyclovir or physical therapy.<sup>2,3</sup> Studies have identified such triggers for NDPH including infection, chemical exposure, stressful life event and surgery involving intubation.<sup>2, 4-7</sup> We introduce a patient whose bilateral cataract extraction appears to have triggered her NDPH.

**Methods:**

Case report.

**Results:**

An 82 year-old white female presented with a constant, bilateral, supraorbital, pressure-like headache of 5 months' duration. Pain began seven days after the second of two sequential cataract extractions that she found stressful. It never subsided, and periodically extended from the vertex down the back of her head. She denied diplopia, photophobia, phonophobia, nausea, autonomic symptoms and positional influence. Her only medication was valsartan for mild hypertension, and she denied illicit drugs and chronic analgesics. She had no viral infections, toxic exposures, other psychological stressors or other surgeries prior to the onset of headache. Initial exam was entirely unremarkable including CT, MRI and exam of neck movement. The 2014 International Classification of Headache Disorders (ICHD-III-beta) diagnostic criteria defines NDPH as headache having a distinct onset with pain that becomes unremitting within 24 hours and persists for more than 3 months. Furthermore, the headache must not be better explained by another ICHD-III-beta diagnosis.<sup>8</sup>

**Conclusions:**

This case suggests that cataract surgery may be a possible trigger for NDPH. Neuro-ophthalmologists should keep this in mind when evaluating patients with headache following cataract surgery.

**References:**

1. Vanast WJ. New daily persistent headaches: definition of a benign syndrome. *Headache*, 26, 317, 1986.
2. Rozen TD. New daily persistent headache: an update. *Curr Pain Headache Rep*, 18(7), 431, 2014.
3. Joshi SG, Mathew PG, Markley HG. New daily persistent headache and potential new therapeutic agents. *Curr Neurol Neurosci Rep*, 14(2), 425, 2014.
4. Li D, Rozen TD. The clinical characteristics of new daily persistent headache. *Cephalalgia*. 22(1):66-69, 2002.
5. Meineri P, Torre E, Rota E, Grasso E. New daily persistent headache: clinical and serological characteristics in a retrospective study. *Neurol Sci.*, 25 Suppl 3, S281-282, 2004.
6. Takase Y, Nakano M, Tatsumi C, Matsuyama T. Clinical features, effectiveness of drug-based treatment, and prognosis of new daily persistent headache (NDPH): 30 cases in Japan. *Cephalalgia*, 24(11), 955-959, 2004.
7. Robbins MS, Grosberg BM, Napchan U, Crystal SC, Lipton RB. Clinical and prognostic subforms of new daily-persistent headache. *Neurology*, 74(17), 1358-1364, 2010.
8. The International Classification of Headache Disorders, 3rd edition (beta version). *Cephalalgia*, 33(9), 629-808, 2013.

**Keywords:** Headache

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Jeffrey Odel, MD is a consultant to Bayer Health Care and Regeneron. Julia Mallory, BA is a research fellow supported by a Summer Student Fellowship grant from Fight for Sight and a grant from the J.T. Tai & Co. Foundation, Inc.

**Poster 157**

**Ten Years Of Temporal Artery Biopsies In Ontario, Canada: A Population-Based Study On Practice Patterns And The Incidence Of Giant Cell Arteritis**

Jonathan A Micieli<sup>1</sup>, Robert Micieli, Edward Margolin

*Department of Ophthalmology and Vision Science, University of Toronto, Toronto, ON, Canada*

**Introduction:**

The goal of this study was to determine the incidence of giant cell arteritis (GCA) in Ontario, Canada and determine the factors influencing which surgical specialists perform temporal artery biopsies (TABs). Significant geographic variability in referral patterns may lead to residents in surgical specialties such as ophthalmology with inadequate exposure to this procedure.

**Methods:**

This was a population-based study including all physicians in Ontario from 2002 to 2013. Using comprehensive physician services billing data from the Intellihealth Medical Services database, physicians performing TABs were categorized by specialty and geographic Local Health Integration Unit. The rate of positive TABs was retrieved from the literature from an Ontario sample during the study period.

**Results:**

The number of TABs was declining over the 10-year study period and the incidence of GCA was determined to be 3.0 per 100,000 people over 50 years of age. Of the 9,958 TABs performed over 10 years, most were performed by general surgeons (38%) followed by ophthalmologists (31%) and plastic surgeons (23%). Ophthalmologists performed significantly more TABs per person compared to general surgeons but significantly more general surgeons performed at least one biopsy. There was significant variation based on geographic location with plastic surgeons performing the most biopsies in regions with more than 1 million people and general surgeons performing most biopsies in rural areas. Among areas with residency training programs there was substantial variation to TAB exposure with many programs in plastics, general surgery and ophthalmology not performing enough procedures to attain competency in this area.

**Conclusions:**

The incidence of GCA in Ontario is consistent with the demographics of this province. Geographic location influences which specialty is most likely to perform TABs and trainees in general surgery, ophthalmology and plastic surgery have variable exposure to TABs and may be unprepared to serve their future practice population.

**References:** None.

**Keywords:** Diagnostic Tests, Giant Cell Arteritis

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## A Method For Recognizing Colorblind Malingering

Andrew E Pouw<sup>1</sup>, Rustum Karanjia<sup>2</sup>, Alfredo A Sadun<sup>2</sup>

<sup>1</sup>Keck School of Medicine, University of Southern California, Los Angeles, CA, USA, <sup>2</sup>Doheny Eye Institute, Los Angeles, CA, USA

### Introduction:

Standard tests of colorblindness screen for organic disease. This new test attempts to recognize malingerers of colorblindness.

### Methods:

An online survey was distributed to 48 self-reported and verified colorblind participants and 81 participants instructed to simulate colorblind malingering. The survey contained three sets of six color-adjusted versions of the standard Ishihara color plates, as well as one set of six unmodified plates. The color-adjusted set demonstrating the highest differences in mean accuracy and group modes between both groups became the basis for a diagnostic test. Statistical measures of the test (sensitivity, specificity, and Youden index) were assessed at each possible cut-off threshold, and a receiver operating characteristic (ROC) function with its area under the curve (AUC) charted.

### Results:

The redshift-adjusted plate set had the highest mean difference between groups (-50%, CI: -42% to -58%) as well as the largest mode difference. Statistical measures showed an optimal cut-off of at least 2 missed redshift plates to identify a colorblind malingerer (Youden index: 0.71, sensitivity: 75.31%, specificity: 95.83%). A stricter cut-off of at least 4 missed redshift plates showed a sensitivity of 43.21% and specificity of 100% (Youden index: 0.43). The AUC of the ROC was 0.89.

### Conclusions:

Our proposed test for recognizing colorblind malingering demonstrates a high degree of reliability. A cut-off threshold of 4 missed redshift plates identified a colorblind malingerer with 100% specificity.

**References:** None.

**Keywords:** Color Blindness, Malingering, Psychophysics, Diagnostic Tests, Pseudo-Isochromatic Plates

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Mycotic Aneurysms Of Intracavernous Internal Carotid Artery

Kumudini Sharma<sup>1</sup>, Vikas Kanaujia<sup>2</sup>, Priyadarshini Mishra<sup>3</sup>, Rachna Agarwal<sup>4</sup>, Alka Tripathy<sup>5</sup>

<sup>1</sup>Department of Ophthalmology, Sanjay Gandhi Postgraduate Institute of Medical Sciences,, Lucknow, India, <sup>2</sup>Department of Ophthalmology, Sanjay Gandhi Postgraduate Institute of Medical Sciences,, Lucknow, India, <sup>3</sup>Department of Ophthalmology, Sanjay Gandhi Postgraduate Institute of Medical Sciences,, Lucknow, India, <sup>4</sup>Department of Ophthalmology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India, <sup>5</sup>Department of Ophthalmology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India

### Introduction:

Mycotic aneurysms of intracavernous segment of internal carotid artery are extremely rare. Early diagnosis is important as they have propensity to rupture.

### Methods:

Case I - is a ten year old boy who had a boil at the tip of the nose which was incised and drained. A week later he developed preseptal cellulitis, fever and headache. He was given systemic antibiotics, fever and headache subsided but was unable to open his left eye. Examination showed complete third and fourth nerve palsy on the left side. Haemogram showed leucocytosis with increased ESR & C-reactive protein. MRI revealed an aneurysm in the region of the left cavernous sinus. CT angiogram demonstrated a lobulated aneurysm arising from cavernous segment of the left ICA, there was another small aneurysm arising from left ICA at the petrous apex. Case II - A three years old boy had multiple boils in the scalp, following which he developed preseptal cellulitis in the both eyes. He was given a long course of antibiotics, later on he developed complete ophthalmoplegia in both eyes. Haemogram showed leucocytosis, raised ESR and C-reactive protein. MRI showed aneurysms in both the cavernous sinuses. DSA confirmed the bilateral ICA aneurysms in cavernous sinus.

### Results:

Case I - The parent artery occlusion using coils was performed. After 3 months he had marked improvement in third nerve palsy. Case II - On antibiotics the ophthalmoplegia on right side improved completely while there was partial improvement in ophthalmoplegia on left side. Endovascular procedure is planned for left ICA aneurysm.

### Conclusions:

Mycotic aneurysm of cavernous segment of ICA presents as cavernous sinus syndrome with features of underlying infection. It results from direct invasion of vascular wall from the nearby infection such as cavernous sinus thrombophlebitis.

### References:

1. Ghali MGZ, Ghali EZ. Intracavernous internal carotid artery Mycotic aneurysms: Comprehensive review and evaluation of the role of endovascular treatment. Clinical Neurology and Neurosurgery. 2013; 115 (10): 1927-1942.
2. Yen PS, Teo BT, Cehn SC, Chiu, TL. Endovascular treatment for bilateral Mycotic intracavernous carotid aneurysm. Case report and review of the literature. J Neurosurg. 2007 Oct; 107(4):868-72.
3. Tomita T, McLone DG. Mycotic aneurysm of the intracavernous portion of the carotid artery in childhood. Case report. J Neurosurg. 1981 May;54(5):681-4.

**Keywords:** Mycotic Aneurysms, Cavernous Sinus, Internal Carotid Artery, Third Nerve Palsy, Preseptal Cellulitis

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 160

### Platelet-Mediated Microvascular Ischemia Mimicking Migraine in Patients with Thrombocythemia

Melissa C Tien<sup>1</sup>, Su Ann Lim

*National Healthcare Group Eye Institute, Department of Ophthalmology, Tan Tock Seng Hospital, Singapore, Singapore*

#### Introduction:

Patients with essential thrombocythemia (ET) can present with migraine-like typical or atypical transient ischaemic cerebral attacks. We describe a case where migraine-like attacks of fortification spectra without headache were the presenting symptoms in a patient with essential thrombocythemia.

#### Methods:

Case report

#### Results:

A 63-year-old cab driver presented with a 4-month history of seeing zig-zag lines, each episode lasting 5-10 minutes. These visual phenomena were experienced in alternate visual fields, were not associated with headache and were unrelated to any identifiable triggers. He had no past history of migraine but suffered from motion sickness. The neuro-ophthalmic examination was completely normal; in particular, there was no visual field defect by confrontation visual field testing. A preliminary diagnosis of late onset migraine aura without headache was entertained; however, the patient declined investigations despite his atypical presentation. A month later, he was admitted with right sided headache, fortification spectra, nausea and giddiness. A complete blood count revealed an elevated platelet level at  $904 \times 10^9$  per litre. Bone marrow aspirate showed a hypercellular marrow with increased megakaryocytes, polyploidy and clustering. Molecular studies found a heterozygous mutation for *JAK2* but no t[9;22] mRNA transcript was detected. Treatment with aspirin and hydroxyurea resulted in immediate resolution of visual symptoms without recurrence.

#### Conclusions:

This rare but well recognized syndrome of atypical transient ischemic attacks in thrombocythemia is likely caused by hypersensitive platelet production leading to ischemic and thrombotic processes in cerebral end-arterial microvasculature. It is important that neuro-ophthalmologists, neurologists as well as haematologists consider this diagnosis in any patient presenting with migraine-like visual symptoms.

**References:** None.

**Keywords:** High Intracranial Pressure/Headache

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Ganglion Cell Damage And Functional Recovery After Optic Neuritis

Alexander U Brandt<sup>1</sup>, Timm Oberwahrenbrock<sup>1</sup>, Friedemann Paul<sup>1</sup>, Fiona Costello<sup>2</sup>

<sup>1</sup>NeuroCure Clinical Research Center, Charité - Universitätsmedizin Berlin, Berlin, Germany, <sup>2</sup>University of Calgary, Hotchkiss Brain Institute, Calgary, AB, Canada

### Introduction:

Optic neuritis (ON) is characterized by inflammation of the optic nerve and retrograde axonal and neuronal damage in the retina. In optical coherence tomography (OCT) the retinal nerve layer swells during the acute phase. In the post-acute phase RNFL and ganglion cell layer thin as a surrogate for axonal loss and neurodegeneration. During the acute phase, ON regularly leads to severe vision loss in the affected eye. Recovery from optic neuritis is defined as reconstitution of visual function after an acute ON. The objective was to describe ON recovery based on neuronal damage in the retina.

### Methods:

Prospective study in patients with acute optic neuritis. 56 patients (46 female and 10 male, age 36±9 years) with acute ON were followed over median 371 (25 – 975) days. Spectral domain OCT was performed on each visit, usually a few weeks apart. Best-corrected visual acuity (BCVA) was measured monocularly by the means of Snellen charts. Neuronal damage was determined as ganglion cell and inner plexiform layer (GCIPL) loss.

### Results:

26 patients showed no GCIPL loss after acute ON, defined as in the range of 2x the standard deviation of unaffected eyes. Eyes with GCIPL loss were divided by the median (-15 µm) in eyes with moderate (n=16) and eyes with severe GCIPL loss (n=14). Male sex was associated with a higher GCIPL loss, whereas there was no influence of age. During the acute phase, eyes with later severe GICPL loss had worse BCVA, but the BCVA between moderate and no-loss eyes was similar. Eyes with more severe ON tended towards higher residual reduction of visual function.

### Conclusions:

Recovery of ON can be defined by objective measurement of GCIPL thickness in the retina. It will be important to define factors predicting ON severity based on this classification.

**References:** None.

**Keywords:** OCT, Optic Neuritis, Demyelinating Disease, Retina

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## To Evaluate The Retinal Nerve Fiber Layer Loss In Optic Neuritis Using Spectral Domain Optical Coherence Tomography

Kowsalya Akkayasamy<sup>1</sup>, Anand Ravikumar<sup>1</sup>, Mahesh Kumar<sup>1</sup>

<sup>1</sup>aravind Eye Care System, Madurai, India, <sup>2</sup>aravind Eye Care System, Madurai, India, <sup>3</sup>aravind Eye Care System, Madurai, India

### Introduction:

To assess the Retinal nerve fibre layer(RNFL) loss in optic neuritis patients using Spectral Domain Optical Coherence Tomography (SDOCT- Heidelberg Engineering software version 5.4.6) and to compare the RNFL loss with the visual acuity and visual outcome after treatment and also to evaluate the quadrant wise retinal nerve fibre layer loss in optic neuritis.

### Methods:

A Cross sectional Observational, prospective study of 20 patients (27 eyes) with optic neuritis, from January 2013 to May 2014, in the department of Neuroophthalmology in a tertiary care centre in South India was conducted. Optic neuritis from 3 months to one year, aged less than 60 years, both unilateral and bilateral were included. Recurrent optic neuritis, extreme ages were excluded. SDOCT imaging was done in all 27 eyes. It was done in both affected and unaffected eyes. Assessment was done in six quadrants Superotemporal, Superonasal, Inferotemporal, Inferonasal, Nasal, Temporal and average of all six quadrants was calculated. RNFL thickness of affected eye was compared with unaffected eye and normative data of device.

### Results:

Average of RNFL thickness in 6 quadrants of affected eyes (80.77 $\mu$ m) with unaffected eyes (97.92 $\mu$ m) showed significant loss (p 0.031). Comparison of the unilaterally affected eye (80.77 $\mu$ m) with normative data (98.00 $\mu$ m), showed significant reduction in RNFL thickness (p 0.031). Bilaterally affected eyes (67.71 $\mu$ m), when compared with normative data (98.43 $\mu$ m), showed clinically significant reduction (p <0.001). Unilaterally affected eyes had less RNFL loss than bilaterally affected eyes. Presenting visual acuity was associated with average RNFL loss, but statistically significant results (beta coefficient = -10.00, p value = 0.357 using regression analysis) could not be obtained. Inferotemporal quadrant was more affected. Temporal sector had more RNFL loss.

### Conclusions:

From our study, spectralis SDOCT is an effective and convenient tool in calculating the RNFL thickness. Reduced thickness correlates with the poor visual function.

**References:** None.

**Keywords:** Optic Neuritis, Retinal Nerve Fiber Layer, Spectral Domain Optical Coherence Tomography, Quadrants, Visual Acuity

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 163

### Retinal Ganglion Cell Injury In Early Pediatric Onset MS

Jennifer Graves<sup>1</sup>, Hardeep Chohan, Benjamin Cedars, Samuel Arnow, Emmanuelle Waubant, Ari Green

*University of California, San Francisco, San Francisco, CA, USA*

#### Introduction:

We sought to document retinal ganglion cell injury in a pediatric onset multiple sclerosis (MS) cohort and evaluate if differences in neuronal levels of injury can be seen by sex in eyes with or without history of optic neuritis (ON).

#### Methods:

This is a retrospective study of consecutive subjects with pediatric onset MS (<18 years at first symptom) who presented for visual evaluation to a pediatric MS center. Spectral domain optical coherence tomography (OCT) was performed. Segmentation of retinal layers was performed using automated software algorithm (Heidelberg). Generalized estimating equations were used to measure associations of sex, optic neuritis and age with retinal nerve fiber layer (RNFL) thickness and ganglion cell layer (GCL) volumes.

#### Results:

Fifty-five subjects (65% female; n=108 eyes) were included. Mean age of MS onset was 12.9 years (SD 3.9); mean age at time of exam was 15.1 years (SD 3.5) with average disease duration of 1.9 years (SD 2.7). In multivariable analyses, history of ON was associated with both reduced RNFL thickness (-16.2  $\mu$ m, 95% CI -25.3, -7.0, p=0.001) and 24% lower ganglion cell layer volumes (-0.097 mm<sup>3</sup>, 95% CI -0.13, -0.065, p<0.001). Older age at time of scan was associated with 2- $\mu$ m thicker measurement of RNFL per year (95% CI 0.68, 3.43, p=0.001). In eyes with history of ON (n=28), males had 12 $\mu$ m lower RNFL thickness, though this did not reach statistical significance (95% CI -29.4, 4.62, p=0.15).

#### Conclusions:

Substantial cell soma loss occurs after optic neuritis even in pediatric onset MS. The observed trend towards greater neuroaxonal loss in males in early MS should be confirmed in larger studies.

**References:** None.

**Keywords:** Pediatric Neuro-Ophthalmology, Demyelinating Disease, Diagnostic Testing (OCT), Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## The Correlation of Critical Flicker Fusion Function and P100 latency of Visual Evoked Potential with Luminance

YanJun Chen<sup>1</sup>, Ashley Lundin, James Ver Hoeve

*Department of Ophthalmology and Visual Sciences, University of Wisconsin-Madison, Madison, WI, USA*

### Introduction:

Two frequent manifestations of optic nerve disease are loss of the sensation of brightness and blurred vision. Here we study the relationship between several measures of optic nerve function, Critical Flicker Fusion (CFF) function, implicit time of the P100 of Pattern Reversal Visual Evoked Potential (PRVEP), log MAR acuity and color function in subjects with visual dysfunction simulated by neutral density filters.

### Methods:

Four healthy adult subjects underwent PRVEP, CFF, color vision (Ishihara plates), and log MAR acuity testing under various levels of luminance. Neutral density filters of 0.6 to 3.0 log unit were used to control stimulus luminance. CFF thresholds, log MAR acuity, Ishihara color plate measurements and P100 latencies and amplitudes of VEP responses were plotted as a function of luminance.

### Results:

CFF decreased and P100 latency increased linearly with the logarithm of stimulus luminance in each of the 4 subjects. P100 latency increased by 9-22 ms, and CFF decreased by 4.7 Hz per log unit decrement in stimulus luminance. Color vision was least affected by changes in log stimulus luminance, and log MAR acuity decreased minimally as luminance increased up to 1.2 log, above which acuity declines precipitously.

### Conclusions:

CFF and P100 implicit time are linearly related to log luminance. The slopes of the linear regression are similar across individuals despite having different baseline thresholds or latencies. The rate of change in CFF or P100 with luminance thus may be useful as a clinical test of optic nerve function. CFF is faster to administer and has low variability on repeat testing.

### References:

1. del Romo GB, Douthwaite WA, Elliott DB. Critical flicker frequency as a potential vision technique in the presence of cataracts. *Investigative ophthalmology & visual science* 2005;46:1107-1112.
2. Douthwaite WA, Halliwell JA, Lomas AM, Yan Muk WK, Topliss JN. Critical fusion frequency in the central visual field. *Ophthalmic Physiol Opt* 1985;5:15-21.
3. Hecht S, Schlaer S, Verrijp CD. INTERMITTENT STIMULATION BY LIGHT : II. THE MEASUREMENT OF CRITICAL FUSION FREQUENCY FOR THE HUMAN EYE. *The Journal of general physiology* 1933;17:237-249.
4. Schneider CW. Electrophysiological analysis of the mechanisms underlying critical flicker frequency. *Vision research* 1968;8:1235-1244.
5. Froehlich J, Kaufman DI. Effect of decreased retinal illumination on simultaneously recorded pattern electroretinograms and visual-evoked potentials. *Investigative ophthalmology & visual science* 1991;32:310-318.

**Keywords:** Critical Flicker Fusion Function, Visual Evoked Potential, Neutral Density Filter, Optic Nerve Function

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** The project described was supported in part by the Clinical and Translational Science Award (CTSA) program, through the NIH National Center for Advancing Translational Sciences (NCATS), grant UL1TR000427. The content is solely the responsibility of the authors and does not necessarily represent the official views of the NIH. This project was supported in part by an unrestricted research grant to the Department of Ophthalmology and Visual Sciences at the University of Wisconsin from Research to Prevent Blindness, Inc.

## Poster 165

### The Effects of Amblyopia on Visual Evoked Potentials

Florin Grigorian<sup>1</sup>, Michael Smit, Adriana P Grigorian

*University Hospitals Case Medical Center, Cleveland, OH, USA*

#### Introduction:

Amblyopia is attributed to cortical changes, therefore Visual Evoked Potentials (VEP) testing is well positioned in the diagnosis, prognosis and management of this condition

#### Methods:

In this retrospective study we investigate how useful VEP can be for amblyopia management. We analyzed the amplitude, latency and morphology of VEP on the amblyopic eye compared to the fellow eye and the presence of changes in the normal eye due to patching. Pattern reversal VEP with 3 channels was used.

#### Results:

We retrospectively analyzed 25 VEP studies of pediatric patients with unilateral amblyopia (refractive and strabismic). Age ranged from 5 to 17 years. Visual acuity in the amblyopic eye ranged from 20/30 to 20/400. In the nonamblyopic eye the patch effect (prolonged implicit time) was rarely seen. In the amblyopic eye the majority of the patients had abnormal VEP in all area analyzed: implicit time, amplitude and morphology compared with the sound eye. Majority of patients had P100 prolonged less than 130ms. Only a minority of patients had P100 prolonged over 130ms. For the latest group, only maintenance patching was indicated.

#### Conclusions:

VEP is a helpful tool for the management as well as the understanding of the pathophysiology of amblyopia.

#### References:

1. Watts, Neveu, Holder, Sloper Visual evoked potentials in successfully treated strabismic amblyopes and normal subjects. JAAPOS 6(6):389-92 2002
2. Henc-Petrinovic, Deban, Gabric, Petrinovic Prognostic value of visual evoked responses in childhood amblyopia. Eur J Ophthalmol 3(3):114-20 1993

**Keywords:** Amblyopia, VEP

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 166

### Doubling Method for Papilledema and Pseudopapilledema

Bokkwan Jun<sup>1</sup>

*Mason Eye Institute/Department of Ophthalmology, Columbia, MO, USA*

#### **Introduction:**

To demonstrate clinical usefulness of doubling method for distinguishing low grade papilledema (Modified Frisen Scale (MFS) grade 1 or 2), pseudopapilledema and normal optic disc by spectral-domain optical coherence tomography (SD-OCT).

#### **Methods:**

The databases were reviewed for patients with papilledema (Modified Frisen Scale grade 1 or 2), patients with pseudopapilledema and patient with normal optic disc (control) who undergone for SD-OCT and stereoscopic optic disc color photography. The doubled thickness of temporal quadrant (Tdt) and the doubled average of temporal and nasal quadrants, which was the sum of them, (Ttn) were calculated and compared to the thickness of inferior quadrant (Ti) in each group. Statistical significance was analyzed.

#### **Results:**

The data were obtained from 37 cases of papilledema, 37 cases of pseudopapilledema and 23 cases of control. In group of papilledema (MFS grade 1 or 2), the average thickness was 126.7 $\mu$ m, Tdt was 170.2 $\mu$ m, Ttn was 179.3 $\mu$ m and Ti was 190.5 $\mu$ m. In group of pseudopapilledema, the average thickness was 113.8 $\mu$ m, Tdt was 157.2 $\mu$ m, Ttn was 165.0 $\mu$ m and Ti was 146.8 $\mu$ m. In control group, the average thickness was 86.1 $\mu$ m, Tdt was 120.2 $\mu$ m, Ttn was 125.0 $\mu$ m and Ti was 117.9 $\mu$ m. In 81% of papilledema, 23% of pseudopapilledema and 37% of control group, Ti was greater than Tdt and Ttn. The difference between group of papilledema and pseudopapilledema and the difference between group of papilledema and control group were statistically significant ( $p < 0.05$ ). (Further data collection and final analysis are pending at the time of submission.)

#### **Conclusions:**

The differentiation of low grade papilledema and pseudopapilledema needs to be determined based on clinical setting. As an adjunct method, the doubling method by SD-OCT is a simple way to help to distinguish between low grade papilledema and pseudopapilledema. During the early stage of papilledema, swelling occurs more in inferior quadrant than temporal or nasal quadrants.

**References:** None.

**Keywords:** OCT, Pseudotumor cerebri, Papilledema, Pseudopapilledema

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 167

### Correlation Between Structural And Functional Changes In Retina In Parkinson's Disease

Manpreet Kaur<sup>1</sup>, Rohit Saxena<sup>1</sup>, Digvijay Singh<sup>1,3</sup>, Madhuri Behari<sup>2</sup>, Pradeep Sharma<sup>1</sup>, Vimla Menon<sup>1</sup>

<sup>1</sup>Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, Delhi, India, <sup>2</sup>Department of Neurology, Neurosciences Center, All India Institute of Medical Sciences, Delhi, India, <sup>3</sup>Division of ophthalmology, Medanta, Gurgaon, India

#### Introduction:

To evaluate structural changes in retina and correlate those with visual function changes in Parkinson's disease (PD).

#### Methods:

A cross-sectional comparative study with 20 cases of PD and 20 age matched healthy controls was conducted. Visual acuity, color vision, contrast sensitivity, visual fields, pattern VER and multifocal ERG were recorded to determine functional change, while structural changes were evaluated as retinal nerve fibre layer (RNFL) thickness, macular thickness, macular volume and ganglion cell-inner plexiform layer complex (GCL-IPL) thickness using SD-OCT.

#### Results:

PD patients were in stage 2 (H&Y stage) of disease (range 1-3), with mean UPDRS III score of  $19 \pm 10.42$ , having average disease duration of  $5.8 \pm 2.78$  years. Visual acuity, color vision and visual fields were unaffected but contrast sensitivity was significantly worse than controls ( $p < 0.001$ ). Multifocal ERG values in the central 2° field revealed decreased foveal electrical activity, with increased pattern VER amplitude and latency. Significant RNFL thinning was observed in the average RNFL ( $p=0.033$ ), superior ( $p=0.018$ ) and temporal ( $p=0.036$ ) quadrants. Significant ganglion cell layer loss was captured on OCT with average, minimum GCL-IPL and all six sectors showing thinning ( $p \text{ values} \leq 0.003$ ). The functional changes correlated significantly with structural changes, disease duration and severity. No correlation was seen between structural alterations in retina and disease duration or severity.

#### Conclusions:

Subclinical visual dysfunction was observed in PD patients with good structural-functional correlation. GCL-IPL thinning may be a more reliable parameter than RNFL thickness for structural alterations in retina.

**References:** None.

**Keywords:** Parkinson's Disease, Retinal Nerve Fibre Layer, Ganglion Cell Layer, Multifocal ERG, Optical Coherence Tomography

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Relationship Between Visual Acuity And Retinal Nerve Fiber Layer Thickness Measured By Spectral Domain Optical Coherence Tomography In Patients With Optic Neuropathy**

Sungeun Kyung<sup>1</sup>, Jiwoong park<sup>1</sup>, Moohwan chan<sup>1</sup>

<sup>1</sup>Dankook University, Cheonan, South Korea, <sup>2</sup>,

**Introduction:**

The structural axonal loss in the optic nerve can be quantified by measuring the peripapillary retinal nerve fiber(RNFL) thickness using optical coherence tomography(OCT). Measuring the RNFL thickness in optic neuropathy provides a structural assessment of the optic nerve, enables prognostic evaluations. We assessed the relationship between RNFL measured by spectral domain optical coherence tomography (SD-OCT) and visual acuity in optic neuritis and traumatic optic neuropathy.

**Methods:**

In this cross-sectional observational study, 40 patients were included. RNFL thickness and visual acuity (VA) in both optic neuritis and traumatic optic neuropathy were measured at least 6 months of the event. The correlations between best-corrected visual acuity (BCVA) and OCT parameters were evaluated using regression analysis. (GraphPad Software, San Diego, CA)

**Results:**

40 subjects received systemic steroids. After at least 6 months, the attack eye had significant RNFL thinning in 4 (superior, inferior, temporal, nasal) quadrants ( $p \leq 0.05$ ) compared to fellow normal eye. When logMAR BCVA was plotted against RNFL thickness, Pearson's correlation tests showed superior ( $p=0.033$ ), temporal( $p=0.9272$ ), inferior( $p=0.0035$ ), and nasal( $p=0.5972$ ) quadrant. Linear regression analysis of logMAR BCVA and RNFL thickness in optic neuropathy patients indicated a relationship of  $y = -33.05X + 111.2$  in superior quadrant, and  $Y = -56.04X + 115.1$  ( $x$ : logMAR VA,  $y$ : RNFL thickness) in inferior quadrant, which was statistically significant.

**Conclusions:**

The relationship between BCVA and SD-OCT parameters were significantly noted in superior and inferior quadrant of eyes with optic neuritis and traumatic optic neuropathy. The lower and superior RNFL thickness may predict visual acuity after optic neuropathy, and correlate with impaired visual function.

**References:**

1. K. Kallenbach and J. Frederiksen, "Optical coherence tomography in optic neuritis and multiple sclerosis: a review," European Journal of Neurology, vol. 14, no. 8, pp. 841–849, 2007.
2. Kim JH, Lee HS, Kim NR, Seong GJ, Kim CY, relationship between visual acuity and retinal structures measured by spectral domain optical coherence tomography in patients with open-angle glaucoma. Invest Ophthalmol Vis Sci. Jul 17; 55(8):4801-11, 2014

**Keywords:** Visual Acuity, Optical Coherence Tomography, Retinal Nerve Fiber Layer Thickness

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Ganglion Cell Layer Thinning Detected By Optical Coherence Tomography As A Sign Of Early Optic Atrophy In Pediatric Papilledema**

Andrew R. Lee<sup>1</sup>, Pradeep Mettu, Kim Jiramongkolchai, Evan Silverstein, M. Tariq Bhatti, Sharon F. Freedman, Mays A. El-Dairi

*Department of Ophthalmology, Duke University, Durham, NC, USA*

**Introduction:**

Recent Spectralis (Heidelberg, Germany) spectral domain optical coherence tomography (OCT) research software can quantify the thickness of each individual retinal layer. Analyzing the macula may give clues to differentiate resolution of papilledema from superimposed atrophy. We sought to characterize and compare the thickness of each macular layer in children with resolved papilledema versus normals.

**Methods:**

Data was obtained in an ongoing retrospective, IRB-approved observational study including eyes of children with resolved papilledema due to idiopathic intracranial hypertension (IIH) and normals who had optic nerve and macular imaging with Spectralis OCT. The average thickness of each of the seven retinal layers in the macula (central 6 mm) and peripapillary retinal nerve fiber layer (pRNFL) were compared between these groups using a two-sided t-test with Bonferroni correction applied as necessary.

**Results:**

Included were 68 eyes from 68 children: 25 with resolved papilledema due to IIH and 43 normals. Eyes with papilledema had significantly thinner ganglion cell layer (GCL) and inner plexiform layer (IPL) than normals: (mean±SD (mm<sup>3</sup>)): 0.99±0.18 vs. 1.13±0.10, p<sub>Bonferroni</sub>=0.0091; and 0.83±0.12 vs. 0.91±0.07, p<sub>Bonferroni</sub>=0.0329, respectively. Area under receiver operating curve (ROC-AUC) showed higher discrimination ability of papilledema vs. normal for GCL and IPL than pRNFL (AUC=0.72, 0.67, and 0.50 respectively). The pRNFL was not different between eyes with pediatric papilledema and normals (mean±SD (microns)): 102.96±28.16 vs. 102.71±10.01, p=0.9667.

**Conclusions:**

In pediatric eyes with resolved papilledema, the macular GCL and IPL may be more sensitive than pRNFL for the detection of early optic atrophy. This finding may help distinguish the resolution of papilledema from superimposed early optic atrophy.

**References:** None.

**Keywords:** Diagnostic Tests (ERG, VER, OCT, HRT, Mferg, Etc), Pseudotumor Cerebri, Pediatric Neuro-Ophthalmology, Retina, High Intracranial Pressure/Headache

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 170

### Peripapillary Retinal Nerve Fiber Layer Thickness Corresponds To Drusen Location And Extent Of Visual Field Defects In Patients With Optic Disc Drusen

Lasse Malmqvist<sup>1</sup>, Birgit Sander, Marianne Wegener, Steffen Hamann

*Department of ophthalmology, Glostrup Hospital, University of Copenhagen, Glostrup, Denmark*

#### **Introduction:**

Optic disc drusen (ODD) are hyaline deposits located within the optic nerve head. While a minority of ODD patients are affected by complications such as anterior ischemic optic neuropathy and retinal hemorrhages, optic coherence tomography (OCT) reveals thinning of the peripapillary retinal nerve fiber layer (RNFL) in most patients. Peripapillary RNFL thinning is thought to be associated with the high frequency of visual field defects seen in these patients. The goal of this study was to investigate the basic characteristics of patients with ODD and to compare the peripapillary RNFL thickness to the anatomic location of ODD, the risk of complications and the extent of visual field defects.

#### **Methods:**

Records from patients, that in the period from January 2014 until October 2014 were either diagnosed with ODD or visited the hospital because of previously identified ODD, were reviewed in this retrospective study.

#### **Results:**

155 eyes of 86 ODD patients were evaluated. 65 % were female and 75 % had bilateral ODD. Presenting complications were seen in 22 % of all inquiries including anterior ischemic optic neuropathy (10,5 %), optic neuritis (5,8%), retinal hemorrhage (4,7%) and central retinal artery occlusion (1,2%). Peripapillary RNFL thinning was seen in 83,6 % of eyes with OCT performed (n =61). Patients with superficial ODD had greater mean peripapillary RNFL thinning than patients with buried ODD ( $P < 0.0001$ ). There was a correlation between mean RNFL thinning and visual field defects as measured by perimetric mean deviation ( $R = -0,72$ ;  $P < 0.0001$ ). No significantly reduced mean peripapillary RNFL thickness was seen in patients with one or more complications compared to patients without complications.

#### **Conclusions:**

Basic characteristics for ODD patients correspond well with the previously reported. Anterior ischemic optic neuropathy is the most frequent severe complication of ODD. Peripapillary RNFL thickness correlates with the anatomic drusen location and the extent of visual field defects in ODD patients.

**References:** None.

**Keywords:** Optic Disc Drusen, Retinal Nerve Fiber Layer, Visual Fields, Perimetry

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Progressive Retinal Structure Abnormalities in Multiple System Atrophy

Carlos E. Mendoza-Santiesteban<sup>1,2</sup>, Jose A. Palma<sup>1</sup>, Jose MARTinez<sup>1</sup>, Lucy Norcliffe-Kaufmann<sup>1</sup>, Thomas R. Hedges<sup>2</sup>, Horacio Kaufmann<sup>1</sup>

<sup>1</sup>New York University, Department of Neurology, NYC, NY, USA, <sup>2</sup>Tufts University, Department of Ophthalmology, Boston, MA, USA

### Introduction:

To study the retinal nerve fiber layer (RNFL) and ganglion cell complex (GCC) thickness in patients with MSA, assess changes over time, and determine whether these measurements could be used as biomarker of disease severity.

### Methods:

We studied 31 eyes from 16 patients with MSA, 24 eyes from 12 Parkinson disease (PD) patients and 44 eyes from 22 controls. Subjects underwent complete ophthalmological evaluation including high definition-optic coherence tomography (HD-OCT) to measure RNFL and GCC thickness. The Unified Multiple System Atrophy Rating Scale (UMSARS) was used to evaluate disease severity and progression.

### Results:

All MSA and control subjects had normal visual acuity and color discrimination. Average RNFL and GCC thickness were significantly reduced in MSA ( $p=0.029$ ,  $p=0.002$ ) and PD ( $p=0.004$ ,  $p<0.001$ ) when compared to controls. In 11 MSA patients, prospective measurements were available. There was a significant reduction of the global RNFL and global GCC thicknesses between the initial and the follow-up measurements ( $10\pm 7$  months). Significant inverse correlations were found between the inferior RNFL thickness and disease duration, and between nasal and inferior RNFL thicknesses and UMSARS scores. From these results, we defined regression equations that predict clinical impairment ( $UMSARS-I = 0.11 * Disease\ duration + 0.828 * age - 0.356 * inferior\ RNFL + 0.537 * global\ RNFL - 9.5$ ;  $UMSARS-II = -0.702 * nasal\ RNFL + 0.837 * global\ RNFL - 0.28 * inferior\ RNFL + 30.6$ )

### Conclusions:

There is a progressive reduction in the RNFL thickness in visually asymptomatic MSA patients. This abnormality can be detected by HD-OCT, a non-invasive, easy-to-perform technique.

### References:

1. Wenning GK, Geser F, Krismer F, et al. The natural history of multiple system atrophy: a prospective European cohort study. *Lancet Neurol* 2013;12(3):264-274.
2. Bodis-Wollner I, Kozlowski PB, Glazman S, Miri S. alpha-synuclein in the inner retina in parkinson disease. *Ann Neurol* 2014;75(6):964-966.
3. Lee JY, Kim JM, Ahn J, Kim HJ, Jeon BS, Kim TW. Retinal nerve fiber layer thickness and visual hallucinations in Parkinson's Disease. *Mov Disord* 2014;29(1):61-67.
4. Schneider M, Muller HP, Lauda F, et al. Retinal single-layer analysis in Parkinsonian syndromes: an optical coherence tomography study. *J Neural Transm* 2014;121(1):41-47.
5. Fischer MD, Synofzik M, Kernstock C, et al. Decreased retinal sensitivity and loss of retinal nerve fibers in multiple system atrophy. *Graefes Arch Clin Exp Ophthalmol* 2013;251(1):235-241.

**Keywords:** OCT, RNFL, MSA, Parkinson

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This work was supported by the Dysautonomia Foundation Inc., (to HK, JM, LNK, CMS, and JAP), the National Institutes of Health (U54-NS065736-01 to HK and LNK) and The Massachusetts Lions Clubs/Research to Prevent Blindness Challenge Grant (to New England Eye Center, CMS & TH).

## Poster 172

### OCT shows Consistent Relationships between Macular Layers and Peripapillary RNFL in Chronic Demyelinating and Compressive Optic Neuropathies

Mark J Morrow<sup>1</sup>, Fawzi Abukhalil

*Harbor-UCLA Department of Neurology, Torrance, CA, USA*

#### **Introduction:**

Optical coherence tomography (OCT) demonstrates thinning of the peripapillary retinal nerve fiber layer (P-RNFL) and inner macular layers in patients with chronic optic neuropathy, including those with multiple sclerosis (MS) and a history of optic neuritis (ON). We sought relationships between each layer of the macula and the P-RNFL in demyelinating and compressive optic neuropathy.

#### **Methods:**

We analyzed spectral-domain OCT results from chronic patients with MS or neuromyelitis optica (NMO) with and without a history of ON (128 MS eyes and 10 NMO eyes), idiopathic ON (13 eyes) and compressive optic neuropathy (14 eyes). No patient had a history of significant visual change within the past 6 months and none had additional eye pathology. We performed regression analysis for each patient group, comparing P-RNFL and individual macular layer values.

#### **Results:**

There were highly significant positive correlations between mean P-RNFL thickness and the volumes of each of the three inner retinal layers (RNFL, GCL and IPL); these layers in turn correlated closely with each other. Similar relationships existed with the temporal quadrant of P-RNFL. These linear correlations were virtually superimposable between any of the demyelinating or compressive neuropathy groups. In contrast, the P-RNFL did not correlate, or demonstrated a weak negative correlation, with the retinal layers from the INL outward.

#### **Conclusions:**

Chronic optic atrophy induces a predictable degree of retrograde degeneration that includes the axons, cell bodies and dendrites of the retinal ganglion cell, regardless of its cause. We found no evidence of trans-synaptic degeneration involving the outer retinal layers; if anything, there was a slight thickening of these layers as the inner retinal layers thinned.

**References:** None.

**Keywords:** Diagnostic Tests (OCT), Optic Neuropathy, Demyelinating Diseases, Tumors

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**OCT Shows Different Relationships Between Macular Layers And Peripapillary RNFL In Acute Versus Chronic Papillitis And Papilledema**

Fawzi Abukhalil<sup>1</sup>, Mark J Morrow

*Harbor-UCLA Department of Neurology, Torrance, CA, USA*

**Introduction:**

Optical coherence tomography (OCT) demonstrates swelling of the optic nerve head (ONH) and peripapillary retinal nerve fiber layer (P-RNFL) in patients with acute papilledema due to intracranial hypertension and in anterior optic neuritis (papillitis). As these conditions become chronic, atrophy of retinal ganglion cell (RGC) axons develops. We sought relationships between the P-RNFL and each layer of the macula in acute and chronic papillitis and compared them to eyes with papilledema.

**Methods:**

We analyzed spectral-domain OCT in eyes with papillitis during the acute and chronic phases (13 eyes) and compared results to a large cohort of eyes with papilledema associated with intracranial hypertension (88 eyes). We performed regression analysis for each patient group, comparing P-RNFL and individual macular layer values.

**Results:**

There were highly significant positive correlations between mean P-RNFL thickness and the volumes of each of the three inner retinal layers (RNFL, GCL and IPL) in chronic eyes with normal or thinned P-RNFLs, but not in eyes with acute ONH swelling. In the latter, there were trends toward positive correlation only with the macular RNFL, which was weakly significant only in the larger papilledema group. In both papillitis and papilledema, there was no correlation between P-RNFL and the outer retinal layers. Relationships of P-RNFL and retinal layer volumes were very similar in papillitis and papilledema eyes.

**Conclusions:**

Conditions that cause swelling of RGC axons at the ONH may induce slight thickening of the macular RNFL, but not of the underlying GCL and IPL that contain RGC cell bodies and dendrites, nor of the outer retinal layers. In contrast, loss of RGC axons associated with chronic papilledema or papillitis causes retrograde degeneration in the macular RNFL, GCL and IPL. Recognition of the normal, close relationship between the P-RNFL and inner retinal layers may allow early detection of RGC swelling.

**References:** None.

**Keywords:** Diagnostic Tests (OCT), Optic Neuropathy, Demyelinating Diseases, High Intracranial Pressure

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## The Photopic Negative Response In Idiopathic Intracranial Hypertension

Heather E. Moss<sup>1,2</sup>, Jason C. Park<sup>1</sup>, J. Jason McAnany<sup>1</sup>

<sup>1</sup>University of Illinois at Chicago/Ophthalmology and Visual Sciences, Chicago, IL, USA, <sup>2</sup>University of Illinois at Chicago/Neurology and Rehabilitation, Chicago, IL, USA

### Introduction:

There is a need for non-invasive biomarkers that will improve diagnosis and management of idiopathic intracranial hypertension (IIH). The photopic negative response (PhNR) is a slow negative component of the full-field electroretinogram with specificity for inner retinal dysfunction associated with retinal ganglion cell injury. It is abnormal in other optic neuropathies and has not been previously studied in IIH or papilledema. Our purpose was to evaluate the PhNR as an index of visual function in IIH.

### Methods:

The PhNR of the photopic full-field, brief flash electroretinogram was recorded in ten subjects with IIH using standard stimulation and recording techniques. PhNR amplitude in IIH subjects was compared 15 visually normal control subjects. In IIH subjects, visual function was assessed using standard automated perimetry mean deviation (SAP-MD) scores and current and prior optic nerve structures were evaluated using the Frisén papilledema grading scale. Associations between PhNR amplitude, SAP-MD and papilledema grade were evaluated.

### Results:

The mean PhNR amplitude was significantly lower in IIH subjects compared with control subjects ( $p = 0.015$  Mann-Whitney rank sum). Log PhNR amplitude and SAP-MD were correlated significantly (pearson correlation,  $r = 0.8$ ,  $p = 0.01$ ). All IIH subjects with reduced PhNR amplitude ( $n=6$ ) had optic atrophy or history of high-grade papilledema, while those with normal PhNR ( $n=4$ ) did not have history of high-grade papilledema ( $p=0.0005$  Fisher's exact). SAP-MD severity was not associated with history of high-grade papilledema ( $p=0.52$  Fisher's exact).

### Conclusions:

We demonstrate that the PhNR amplitude can be decreased in IIH subjects and that the decrease correlates with compromise in a clinical measure of visual function (SAP-MD). The observation that high-grade papilledema is associated with PhNR abnormalities but not SAP-MD severity supports literature implicating PhNR association with pre-perimetric optic nerve injury. Further studies are needed to determine the clinical utility of PhNR as a marker for diagnosis and monitoring of IIH.

**References:** None.

**Keywords:** Diagnostic Tests, High Intracranial Pressure, Optic Neuropathy, Pseudotumor Cerebri, Perimetry

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** NIH K12 EY021475, K23 EY024345, R00 EY019510, P30 EY01792, Illinois Society for the Prevention of Blindness Research Grant, unrestricted departmental grant from Research to Prevent Blindness



## Poster 175

### Attenuated Hunter Syndrome: A Rare Cause of Simultaneous Retinal and Optic Nerve Disease

Kannan Narayana<sup>1</sup>, Lau Heather, Rachel Nolan, Rucker Janet, Balcer Laura, Galetta Steven

*NYU School of Medicine, Dept of Neurology, New York, NY, USA*

#### Introduction:

Hunter Syndrome (HS) is a multisystemic disorder characterized by glycosaminoglycan (GAG) accumulation due to enzyme deficiency of iduronate 2-sulfatase. This case report characterizes visual pathway involvement in HS.

#### Methods:

Case report.

#### Results:

A 44-year-old man with HS presented with headaches, nausea, and peripheral vision loss. Visual acuities were 20/25 OU with normal color vision. There was no APD; bilateral optic disc edema was noted. Humphrey visual fields (VF) showed pericentric ring scotomas. Full field ERG was normal. Intracranial pressure monitoring was normal. Orbital MRI demonstrated bilateral scleral thickening with a suggestion of scleral canal crowding. Over the next 16 months, VF defects mildly progressed, despite stable acuities and disc appearance. OCT demonstrated microcystic macular edema, thinning of the retinal nerve fiber layer (RNFL: OD 100 / OS 129 microns), ganglion cell layer (GCL), outer nuclear and photoreceptor layers. At 15 months, RNFL thickness declined in OD (by 13 microns), increased in OS (by 9 microns), while macular and retinal changes were stable.

#### Conclusions:

This patient illustrates the complex mechanisms of visual impairment in HS. We hypothesize that the retinal changes and scleral canal crowding with optic disc edema both resulted from GAG deposition. Likewise, the ring scotomas of our patient were pericentral and a full field ERG was normal. Segmentation of the GCL demonstrated a pattern of loss consistent with the field defect. GCL loss could have resulted from trans-synaptic degeneration of the photoreceptor layer, but we think primary injury to the GCL is most likely. The visual acuity preservation and symmetrical pattern of VF loss make a primary optic nerve mechanism unlikely. These findings demonstrate that a combination of retinal and optic nerve changes, including optic disc swelling, optic atrophy and retinopathy, occur in HS. This case emphasizes the potential role for OCT to help delineate the areas of visual pathway involvement.

**References:** None.

**Keywords:** Optical Coherence Tomography (OCT), Hunter Syndrome, Disc edema, Retina, Eye Abnormalities

**Financial Disclosures:** K. Narayana: None. H.Lau:Consultant for Pfizer, Contracted Research; Roles for Self; Shire, Genzyme, Ultragenyx, Amicus, Glaxo Smith Kline, Pfizer. R.Nolan: None. J. Rucker: None. L.Balcer: received personal compensation from Biogen Idec and consulting for Biogen Idec, Vaccinex and Genzyme. She is on a clinical trial advisory board for BiogenIdec.S.Galetta: has received consulting honoraria from Biogen Idec, Genzyme and Vaccinex

**Grant Support:** None.

## Poster 176

### Ganglion cell and retinal nerve fiber layer analysis in a case of PION

Joshua Pasol<sup>1</sup>, Yevgeniya Libershteyn<sup>2</sup>, Allison McClellan<sup>2</sup>

<sup>1</sup>Bascom Palmer Eye Institute, Miami, FL, USA, <sup>2</sup>Miami Veteran's Hospital, Miami, FL, USA

#### Introduction:

Posterior Ischemic Optic Neuropathy (PION) is a retro bulbar optic nerve ischemia presenting with sudden, painless visual loss with the presence of decreased vision, visual field defect, an afferent pupillary defect, yet the retina and optic nerve appear normal initially. PION can be seen in isolation, as part of giant cell arteritis, or post surgical. A PubMed search does not reveal any cases of OCT in the phases of PION. Ganglion cell OCT can help determine when ganglion cell thinning occurs in ischemic optic neuropathy. A case of PION and OCT is presented.

#### Methods:

Single case study of a 64-year-old woman with sudden, painless loss of vision in the right eye. She had no symptoms of GCA and there was no recent surgical history. The visual acuity showed count fingers at 3 feet in the right eye and 20/20 left eye. HVF right eye (Figure 1) showed a superior altitudinal defect. There was a large right afferent pupillary defect. The optic nerve, macula and peripheral retina appeared normal (Figure 2). Initial OCT of the GCL and RNFL were normal (Figure 3). MRI of the orbits showed no optic nerve enhancement (Figure 4). Numerous blood tests were normal including ESR and CRP. Three days of IV methylprednisolone followed by oral prednisone taper were given. Follow up GCL and RNFL OCT were obtained 4 weeks (Figure 5) and 9 weeks (Figure 6) after initial visit.

#### Results:

RNFL OCT is initially normal in PION followed by later thickening likely due to axoplasmic stasis. RNFL thins in the later phase of optic nerve ischemia. GCL OCT is initially normal in PION and shows thinning at 4 weeks and 9 weeks in this case.

#### Conclusions:

GCL OCT thins rapidly following PION suggestive of rapid loss of ganglion cells. There is a short time window in preventing ganglion cell death in potential optic nerve ischemia treatment.

**References:** None.

**Keywords:** optic neuropathy, OCT

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Perimacular Ganglion Cell Complex Thinning Detected By Spectral-Domain OCT Useful In Detecting Optic Tract Syndrome

Fannie Petit<sup>1,2</sup>, Frédéric Bédard Dallaire<sup>3</sup>, Sébastien Gagné<sup>1</sup>, Laurent Létourneau Guillon<sup>4</sup>, Jacinthe Rouleau<sup>1</sup>

<sup>1</sup>Department of Ophthalmology, Notre-Dame Hospital, Montreal, QC, Canada, <sup>2</sup>Department of Ophthalmology, Maisonneuve-Rosemont Hospital, Montreal, QC, Canada, <sup>3</sup>Faculty of medicine and health sciences, Sherbrooke University, Sherbrooke, QC, Canada, <sup>4</sup>Department of Radiology, Notre-Dame Hospital Montreal, QC, Canada

### Introduction:

Unilateral lesions to the optic tract lead to optic nerve atrophy (bow-tie atrophy) and concurrent macular ganglion cell complex (GCC) thickness reduction in a reproducible pattern in both eyes. This is called optic tract syndrome and it is reflected clinically as an incongruous homonymous hemianopia and a contralateral relative afferent pupillary defect (RAPD).

### Methods:

Case report description of a patient with an optic tract lesion and its characteristic findings at the Spectral-Domain OCT (SD OCT) using the OD-OS asymmetry analysis.

### Results:

The OD-OS asymmetry analysis can be very useful to detect ipsilateral peri-macular temporal thinning and a contralateral peri-macular nasal thinning of the GCC which are characteristic signs of this syndrome. It compares the thickness of cells between eyes by highlighting in grayscale cells that are thinner than corresponding cell in the opposing eye. The grayscale of this analysis, by enhancing the pattern resembling the homonymous hemianopia, makes it easier to detect optic tract lesions and to differentiate them from glaucoma.

### Conclusions:

It is very useful clinically when a patient is erroneously first referred for suspicion of glaucoma and when bow-tie atrophy is not evident.

### References:

1. Yamashita T, Miki A, Iguchi Y, Kimura K, Maeda F, Kiryu J. Reduced retinal ganglion cell complex thickness in patients with posterior cerebral artery infarction detected using spectral-domain optical coherence tomography, *Jpn J Ophthalmol*, 56:502-510, 2012.
2. Jindahra P, Petrie A, Gordon T. P. Retrograde trans-synaptic retinal cell loss identified by optical coherence tomography. *Brain*, 132; 628–634, 2009.
3. Kanamori A, Nakamura M, Yamada Y, Negi A. Spectral-domain optical coherence tomography detects optic atrophy due to optic tract syndrome. *Graefes Arch Clin Exp Ophthalmol*, 251:591–595, 2013.
4. Spectral domain optical coherence tomography (SD-OCT). [Heidelberg engineering website]. Available at: <http://www.heidelbergengineering.com/us/products/spectralis-models/imaging-modes/spectral-domain-oct/>. Accessed July 14, 2014.
5. Romero R, Gutierrez I, Wang E, Reder A, Bhatti T, Bernard J, Javed A. Homonymous Hemimacular Thinning: A Unique Presentation of Optic Tract Injury in Neuromyelitis Optica. *Journal of Neuro-Ophthalmology*, 32, 150–153, 2012.

**Keywords:** Spectral-Domain Optical Coherence Tomography, Macular Ganglion Cell Complex, Homonymous Hemianopsia, Optic Tract Syndrome

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**To Evaluate Changes In Retinal Nerve Fiber Layer And Ganglion Cell Layer On Cirrus HD-OCT In Cases Of Multiple Sclerosis (With And Without Optic Neuritis) And Optic Neuritis**

Ganesh Pillay<sup>1</sup>, Rohit Saxena<sup>1</sup>, Rohit Bhatia<sup>2</sup>

<sup>1</sup>*dr.Rajendra Prasad Centre Of Ophthalmic Sciences,Aiims, New Delhi, India,* <sup>2</sup>*cardio-Neuro Centre, All India Institute Of Medical Sciences, New Delhi, India*

**Introduction:**

Purpose: To prospectively examine relation of visual functions to retinal nerve fibre layer(RNFL) thickness and ganglion cell layer(GCL) complex changes over 6 months as a structural biomarker for axonal loss in multiple sclerosis and optic neuritis patients.

**Methods:**

Patient with Multiple sclerosis without optic neuritis (MS) (n=20; 40 eyes), Optic neuritis (ON) (n=26; 58 eyes), Multiple sclerosis with optic neuritis (MS+ON) (n=24; 48), and disease-free controls (n=20; 40 eyes). Visual testing was performed for each eye at baseline and 6 months which included Snellens visual acuity (in Log MAR units), Pelli-Robsons contrast sensitivity, Ishiharas colour vision, visual evoked response, Retinal nerve fibre layer and ganglion cell layer complex thickness was measured using Cirrus HD-OCT. EDSS score was calculated for multiple sclerosis patients.

**Results:**

Average RNFL thickness was reduced significantly [with respect to controls (86.1µm) ] among the eyes of patients with MS+ON (56.8µm) (P<0.005) as well the unaffected fellow eye (67.3µm) (P<0.005) and ON (52.8µm) (P<0.005) but no significant difference was found in MS patients (80.6µm) (P=1). GCL complex thickness was reduced significantly [with respect to controls (84.3µm) ] among eyes of MS+ON (56.1µm) (P<0.005), the fellow unaffected eye of this group(66.2µm) (P<0.005), ON (54µm) (P<0.005) as well as MS patients (65.8µm) (P<0.005). GCL complex thickness significantly correlated clinically with visual acuity (P=0.0032), colour (P<0.001), EDSS (P=0.0047), VER amplitude (P=0.0078) and latency (P=0.0001) in MS patients and significantly clinically correlated with visual acuity (P=0.0042), colour vision (<0.001) and contrast (p=0.02) in ON patients.

**Conclusions:**

Ganglion cell layer complex thickness measurement is a better clinical marker than retinal nerve fibre layer thickness (Cirrus HD-OCT) in Multiple sclerosis and optic neuritis patients. Although eyes with history of optic neuritis demonstrate the greatest reduction in GCL complex thickness, MS non-ON eyes have less GCL complex thickness than controls, suggesting the occurrence of chronic axonal loss separate from acute attacks in MS patients.

**References:** None.

**Keywords:** Demyelinating Disease, Optic Neuropathy, Diagnostic Tests, Neuro-Ophth & Systyemic Disease, Higher Visual Functions

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Examination of Visual Evoked Potential (VEP) in a Pediatric Population with Newly Diagnosed Elevated Intracranial Hypertension

Ahmara G Ross<sup>1</sup>, Valeria Fu<sup>3</sup>, Gabrielle Bonhomme<sup>1,2</sup>, Islam Zaydan<sup>1,2,4</sup>, Kenwal Nishcal<sup>3</sup>, Leanne Lope<sup>3</sup>, Christin Sylvester<sup>3</sup>, Ellen Mitchell<sup>1,2,3</sup>

<sup>1</sup>UPMC/ University of Pittsburgh, Pittsburgh, PA, USA, <sup>2</sup>UPMC/ University of Pittsburgh Department of Neuro-Ophthalmology, Pittsburgh, PA, USA, <sup>3</sup>UPMC/Children's Hospital of Pittsburgh, Pittsburgh, PA, USA, <sup>4</sup>UPMC/ University of Pittsburgh Department of Neurology Pittsburgh, PA, USA

### Introduction:

Pattern VEP has been demonstrated in adults (>16 years of age) to have severely delayed latencies demonstrating a significant correlation between the affects of intracranial pressure and subsequent health of the visual system. Most studies involve adult populations, with pediatric populations uninvestigated. We hypothesize that VEP, similar to adult populations, can be used to predict dysfunction of the visual pathway from elevated ICP.

### Methods:

32 children (12.08 years, SD 4.40) diagnosed with IIH (37.41 mmHg $\pm$ 7.16) using the Modified Dandy Criteria were examined. Pattern VEPs were recorded monocularly. Va LogMar (Mean 0.1291 $\pm$ 0.252), OCT average RNFL (Mean 131.08 $\mu$ m $\pm$ 4.56) will be compared and associated with opening intracranial pressure were recorded at presentation. These parameters were used to compare outcomes of VEP Latency and Amplitude compared with an age matched control population obtained within 4 months of documented elevated opening pressure.

### Results:

The average age (male 9.696 $\pm$ 0.8396; female 13.914 $\pm$ 1.0423, p=0.002) and BMI (male 23.323 $\pm$ 2.14; female 34.646 $\pm$ 2.34, p=0.003) differed significantly between the population of pediatric patients evaluated. The presence of optic disc edema showed no association with elevated ICP. Average RNFL was positively correlated with elevated intracranial pressure, while RNFL (131.08 $\pm$ 4.56, r<sup>2</sup>=1.676, p=0.04).

VEP was analyzed in this population. Total p100 latency was significantly reduced in patients presenting with elevated ICP (101 $\pm$ 1.31, p=0.03), while amplitude was elevated (25.16 $\pm$ 2.32, p=0.02). While latency was not associated with elevations in intracranial pressure (r<sup>2</sup>=0.01), elevation in amplitude was found to be positively correlated (r<sup>2</sup>=0.84, p=0.02).

### Conclusions:

Age, BMI, Visual acuity, color vision, and the presence of optic disc edema are inaccurate measures to be used to determine visual pathway dysfunction elevated ICP. Optical Coherence Tomography and Visual fields obtained can indicate pathology. VEP can be used to determine elevation in ICP, and might represent a easily obtainable and reliable method of monitoring changes in intracranial pressure.

**References:** None.

**Keywords:** Pediatric IIH, Idiopathic Intracranial Hypertension, Visual Evoked Potential

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Retinal Nerve Fiber Layer Thickness in a Population-Based Study of Elderly Subjects: The Alienor Study

Marie B Rougier<sup>1</sup>, Jean F Korobelnik<sup>1,2,3</sup>, Cedric Schweitzer<sup>1</sup>, Marie N Delyfer<sup>1,2,3</sup>, Jean F Dartigues<sup>2,3,4</sup>, Cecile Delcourt<sup>2,3</sup>, Catherine Helmer<sup>2,3,5</sup>

<sup>1</sup>University Hospital, Ophthalmology, Bordeaux, France, <sup>2</sup>INSERM, ISPED, INSERM Unit 897, Bordeaux, France, <sup>3</sup>University Bordeaux, Bordeaux, France, <sup>4</sup>University Hospital, memory consultation Bordeaux, France, <sup>5</sup>INSERM, CIC 1401 Bordeaux, France

### Introduction:

To establish normative data of retinal nerve fiber layer (RNFL) thickness in the elderly and to determine the factors influencing its thickness.

### Methods:

Peripapillary RNFL thickness was measured with spectral domain OCT (SD-OCT) in 210 elderly participants from the Alienor population-based study who were aged 75 years or older. The measure was assessed in 6 segments. RNFL data were analyzed across age and sex strata in non-glaucoma participants. Mixed linear models were used to evaluate the associations of RNFL thickness with age, sex, ocular parameters (axial length, cataract), and vascular risk factors (blood pressure, diabetes, body mass index, smoking status, heart disease and stroke).

### Results:

The mean global RNFL thickness was 91.4  $\mu\text{m}$  (SD: 12.6), ranging from 55 to 122; the highest values were found in the inferotemporal and superotemporal segments. After adjustment for sex and ocular parameters, including axial length, increasing age was significantly associated with lower thickness globally (mean thinning per decade = 5.6  $\mu\text{m}$ ,  $p=0.003$ ), in the superotemporal (-12.7  $\mu\text{m}$  per decade,  $p<0.0001$ ) and inferotemporal (-8.1  $\mu\text{m}$  per decade,  $p=0.022$ ) segments. RNFL thickness tended to be higher in women than in men, but this trend was significant only in the inferotemporal segment (+6.6  $\mu\text{m}$  for women,  $p=0.012$ ). The axial length was associated with RNFL thickness globally and in most segments. RNFL thickness did not differ according to cataract extraction. There were no association between vascular factors and RNFL thickness.

### Conclusions:

In concordance with previous studies in adult population, we found a decrease of RNFL thickness with age, even after 75 years. The decrease was stronger in the supero and inferotemporal segments. Moreover, the study demonstrated that RNFL tended to be higher in women, particularly in the inferotemporal segment. Finally, as RNFL thickness did not differ according to cataract extraction, it can be used in both people with and without cataract extraction.

**References:** None.

**Keywords:** OCT, Retinal Nerve Fiber Layer, Epidemiology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## **OCT-Derived Retinal Capillary Density is Decreased in Corresponding Areas of Retinal Neuron Loss in Optic Neuropathy**

Min Wang<sup>1</sup>, Randy H. Kardon<sup>2</sup>

<sup>1</sup>Eye and ENT Hospital of Fudan University/Department of Ophthalmology, Shanghai, China, <sup>2</sup>Iowa City VA Center for Prevention and Treatment of Visual Loss, Department of Veterans Affairs Hospital, University of Iowa Hospital and Clinics, Iowa City, IA, USA

### **Introduction:**

To evaluate the chronic effect of optic neuropathy on retinal capillary density using a new OCT-based method of angiography that does not require dye injection. We hypothesized that loss of retinal ganglion cells and their axons would reduce metabolic demand of the inner retina, causing loss of capillary blood flow.

### **Methods:**

Retinal capillaries were visualized using en face OCT angiography, based upon the detection of intravascular movement of light-scattering particles. Fourteen eyes were studied from 8 patients with optic neuritis (ON; n=2), neuromyelitis optica (NMO; n=1), nonarteritic anterior ischemic optic neuropathy (NAION; n=2), Leber hereditary optic neuropathy (LHON; n=2) and compressive optic neuropathy (pituitary adenoma; n=1).

### **Results:**

In ON and NMO patients, retinal capillary density derived from blood flow was decreased significantly in the macula and around the optic disc that corresponded to the areas with loss of retinal nerve fiber layer (RNFL) and ganglion cell-inner plexiform complex (GCC). Following resolution of disc edema, NAION eyes demonstrated lack of retinal capillary filling in altitudinal sectors corresponding to the same areas with loss of RNFL, GCC, and visual field. In LHON patients, maculo-papillary bundle defects and macula areas of GCC loss were observed using en face SD-OCT and retinal capillary flow was absent in the corresponding macula and retinal areas. In one patient with compressive optic neuropathy from a pituitary adenoma the structural loss in the maculo-papillary bundle, nasal RNFL and GCC was associated with lack of corresponding retinal capillaries rendered by OCT angiography.

### **Conclusions:**

Loss of retinal ganglion cells and their axons appears to result in sufficient reduction in metabolic requirements of the retina to result in significant loss of capillary blood flow in corresponding locations. OCT angiography may help in understanding the link between metabolic demand and blood flow in eyes with optic neuropathy.

**References:** None.

**Keywords:** Diagnostic Tests, Optic Neuropathy, Retina, Capillary Blood Flow, Ganglion Cell Complex

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 182

### Adaptive Optics Imaging With Histopathologic Correlation in Cancer-Associated Retinopathy

Zoë R Williams<sup>1</sup>, Ethan A Rossi<sup>2</sup>, David A DiLoreto, Jr.<sup>1</sup>

<sup>1</sup>Flaum Eye Institute, University of Rochester, Rochester, NY, USA, <sup>2</sup>Center for Visual Science, University of Rochester, Rochester, NY, USA

#### Introduction:

We describe the first report of in vivo adaptive optics imaging in cancer-associated retinopathy as correlated with histopathology.

#### Methods:

A patient with cancer-associated retinopathy (positive anti-retinal autoantibody testing and markedly diminished ERG) was imaged with fundus photography, confocal scanning laser ophthalmoscopy (cSLO) and high-density spectral domain OCT. cSLO images were obtained using infrared (IR) reflectance, blue light fundus autofluorescence (FAF) and IR FAF imaging modes. Adaptive optics scanning light ophthalmoscopy (AOSLO) images were obtained of a ~900  $\mu\text{m}^2$  region centered on the foveola, and strips extending ~2.5 mm nasally, 2.4 mm temporally, 2.4 mm superiorly and 2 mm inferiorly. One month later, images were acquired of a ~1.3 mm<sup>2</sup> region centered on the foveola. Post-mortem PO sections of formalin-fixed globe were obtained and processed for H&E.

#### Results:

Relatively normal cone photoreceptors were visible in an elliptical area within the foveola. A narrow band of hypo-reflectivity containing sparse cones separated this central region from a surrounding annular area that contained both normal and abnormal appearing cones. Cone structure was extremely abnormal at 300  $\mu\text{m}$ , with large areas of hypo-reflective cones interspersed with hyper-reflective cones exhibiting irregular morphology. The surrounding retina was relatively hypo-reflective with very few identifiable cones, and clumps of hyper-reflective material extending into the inner retina. Histopathology revealed complete loss of photoreceptor cell nuclei except at the fovea where a single layer of nuclei was seen in the outer nuclear layer with attached cone inner segments. There were migrated RPE cells and melanophages in the atrophic inner nuclear layer.

#### Conclusions:

This is the first report of in vivo adaptive optics imaging with histopathologic correlation in any disease entity. Our findings validate the ability of AOSLO to identify cone photoreceptor loss in vivo in patients with cancer associated retinopathy.

**References:** None.

**Keywords:** Adaptive Optics Imaging, Cancer Associated Retinopathy, Histopathology

**Financial Disclosures:** Ethan Rossi: 1) University of Rochester - Patent 2) Canon, Inc. - Financial Zoe Williams: no disclosures David DiLoreto, Jr: no disclosures

**Grant Support:** This work was supported by the Research to Prevent Blindness (Flaum Eye Institute, Rochester, New York).



## Robust Optic Nerve Head Analysis Based On 3D Optical Coherence Tomography

Ella M Kadas<sup>1</sup>, Hanna Zimmermann<sup>1</sup>, Janine Mikolajczak<sup>1</sup>, Wolf Lagrèze<sup>2</sup>, Friedemann Paul<sup>1,3</sup>, Alexander U Brandt<sup>1</sup>

<sup>1</sup>NeuroCure Clinical Research Center, Charité – Universitätsmedizin Berlin, Berlin, Germany, <sup>2</sup>University Eye Hospital Freiburg, Freiburg, Germany, <sup>3</sup>Department of Neurology, Charité - Universitätsmedizin Berlin, Berlin, Germany

### Introduction:

Many conditions such as glaucoma (GD), optic neuritis (ON), multiple sclerosis (MS) or idiopathic intracranial hypertension (IIH) affect the optic nerve head (ONH). Optical coherence tomography (OCT) allows 3D ONH imaging. However, current ONH analysis methods work in only atrophic or in swollen ONH but not both. Manual steps for masking Bruch's membrane opening (BMO) or ONH center are regularly required. Our objective was to develop a robust and fully automatic ONH quantification algorithm applicable in atrophic, normal and swollen conditions.

### Methods:

Algorithm development based on the following steps. Motion artifacts were corrected. The region of the retinal pigment epithelium (RPE) as leading structure to detect BMO was estimated. The RPE was enhanced and blood vessels suppressed during a filtering sequence. The ONH centroid was then established from the BMO. The optic nerve head volume (ONHV) was derived from the borders of RPE, BMO and inner limiting membrane (ILM) within a 3.4 mm diameter circle around the ONH centroid. The algorithm was tested on ONH volume scans acquired with a Spectralis HD-OCT.

### Results:

Scans from 148 eyes of healthy controls, GD patients, MS patients with and without previous ON and IIH patients were processed. The algorithm failed in 4 eyes (2.7%) due to unpropitious position of blood vessels. ONHV showed excellent test-retest reliability in a sub-study including 3 repeated measurements of 11 healthy eyes (intra-class correlation coefficient: 0.984). Finally, ONHV correlated well to the retinal nerve fiber layer thickness provided by the device ( $p < 0.001$ ).

### Conclusions:

Our algorithm was able to reliably determine BMO dimensions and estimate the ONH centroid. ONHV estimations were robust in both atrophic and swollen ONH conditions. The presented method should therefore serve as a source for several ONH parameters and as a reliable tool to follow-up ONH changes in diseases involving papilledema and ONH atrophy.

### References:

 None.

**Keywords:** Optical coherence tomography, Optic nerve head, Multiple sclerosis, idiopathic intracranial hypertension, Glaucoma

**Financial Disclosures:** HZ received travel grant from Novartis Pharma and speaker honoraria from Teva. AUB is cofounder and director of Motognosis and reports grants, personal fees and non-financial support from Novartis Pharma, personal fees and non-financial support from Biogen Idec, personal fees from Heidelberg Engineering, personal fees from Bayer, personal fees from TEVA, outside the submitted work. FP received research support from the German Ministry for Education and Research (Competence Network Multiple Sclerosis), Guthy Jackson Charitable Foundation and National Multiple Sclerosis Society, research grants and speaker honoraria from Bayer, Teva, Genzyme, Merck, Novartis, MedImmune and is member of the steering committee of the OCTIMS study (Novartis). All other authors report nothing to disclose. Conflict of Interest The ONHV algorithm was filed as a patent application naming inventors EMK, FP and AUB from this poster among others as inventors. All other authors report no conflict of interest.

**Grant Support:** This study was supported by BMWi grant ZIM-KF KF2291305AK3.

## Ocular Motility Defects with Concordance Neuroimaging

Nagham AlZubidi<sup>1</sup>, Constance L. Fry<sup>2</sup>, John E. Carter<sup>1,2</sup>, Bundhit Tantiwongkosi<sup>3</sup>

<sup>1</sup>Department of Neurology, University of Texas Health Science Center, San Antonio, TX, USA, <sup>2</sup>Department of Ophthalmology, University of Texas Health Science Center, San Antonio, TX, USA, <sup>3</sup>Department of Radiology, University of Texas Health Science Center, San Antonio, TX, USA

### Introduction:

Ocular motility defects (OMDs) have various etiologies and comprise one of the most comprehensive differential diagnoses in neuro-ophthalmology. The differential diagnosis of (OMDs) ranges from a benign to a life-threatening cause. MRI or CT of orbit/brain are complementary to each other and play a significant role in neuro-ophthalmology and neurology evaluation. Typically, MRI is considered the imaging of choice for neuro-ophthalmological assessment, with CT being reserved for disorders involving bone, calcification or those conditions requiring a surgical approach to the bone. We herein present a retrospective review of a series of patients with (OMDs) with concomitant neuro-imaging findings.

### Methods:

This is a retrospective review of neuro-imaging studies of patients with ( OMDs) seen in our institution and reviewed by a single neuro-radiologist. Patients with (OMDs) secondary to myasthenia gravis, intraocular pathology, functional vision loss and thyroid orbitopathy were excluded.

### Results:

Sixty-five patients with (OMDs) with abnormal neuroimaging were identified. Of the 65 patients, 23 (35.4 %) patients had involvement of a single cranial nerve. There were 6 patients (9.2 %) with isolated third nerve palsy due to *Guillain-Barré syndrome (GBS)* variant, acute cerebrovascular accident (CVA) and *B-cell lymphoma* . Two patients (3.07%) had isolated fourth nerve palsy due to metastatic renal cell carcinoma and trauma. Fifteen patients (23.07 %) had isolated sixth nerve palsy from CVA, *B-cell lymphoma*, *leptomeningeal* carcinomatosis (LC), metastasis, glioblastoma multiforme (GBM), Chiari I malformation, multiple sclerosis and neurosarcoidosis. Twenty-six patients (40%) had multiple cranial nerve involvement due to CVA, trauma, cavernous meningioma, *lymphoma*, LC, metastasis, and GBM. Seven patients (10.76%) had nystagmus and 6 patients (9.23%) had gaze palsies due to CVA, trauma, and pineoblastoma. There were 3 patients (4.61%) with idiopathic muscle atrophy associated with trauma.

### Conclusions:

Our findings highlight the value of advanced neuro-imaging in the early diagnosis and localization of the various causes of (OMDs).

### References:

1. Mehta S, Loevner LA, Mikityansky I, Langlotz C, Ying GS, Tamhankar MA, Shindler KS, Volpe NJ. The diagnostic and economic yield of neuroimaging in neuro-ophthalmology. *J Neuroophthalmol.* 2012;32(2):139-44.
2. Lee AG, Johnson MC, Policeni BA, Smoker WR. Imaging for neuro-ophthalmic and orbital disease - a review. *Clin Experiment Ophthalmol.* 2009;37(1):30-53.
3. Weiss RA, Haik BG, Saint-Louis LA, Ellsworth RM. Advanced diagnostic imaging techniques in ophthalmology. *Adv Ophthalmic Plast Reconstr Surg.* 1987;6:207-63.
4. Chou KL, Galetta SL, Liu GT, Volpe NJ, Bennett JL, Asbury AK, Balcer LJ. Acute ocular motor mononeuropathies: prospective study of the roles of neuroimaging and clinical assessment. *J Neurol Sci.* 2004;219(1-2):35-9.
5. Murchison AP, Gilbert ME, Savino PJ. Neuroimaging and acute ocular motor mononeuropathies: a prospective study. *Arch Ophthalmol.* 2011;129(3):301-5.

**Keywords:** Neuro-Imaging, Ocular Motility Deficit, Neuro-Rdiology And Neuro-Ophthalmology, Concordance Neuroimaging, Diplopia And Neuro-Imaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Mural Enhancement of the Intracranial Internal Carotid Artery in Giant Cell Arteritis

Sidney M. Gospe III<sup>1</sup>, Pradeep Mettu<sup>1</sup>, M. Tariq Bhatti<sup>1,2</sup>, Samuel Kuzminski<sup>3</sup>, Timothy J. Amrhein<sup>3</sup>, Mays A. El-Dairi<sup>1</sup>

<sup>1</sup>Duke University Medical Center/Department of Ophthalmology, Durham, NC, USA, <sup>2</sup>Duke University Medical Center/Department of Neurology, Durham, NC, USA, <sup>3</sup>Duke University Medical Center/Department of Radiology, Durham, NC, USA

### Introduction:

While autopsy studies of patients with giant cell arteritis (GCA) have demonstrated inflammatory involvement of the internal carotid artery (ICA), radiologic signs of ICA inflammation have not been a prominent feature in the published literature. We present three patients with biopsy-proven GCA who demonstrated evidence of mural enhancement of the intracranial ICAs by magnetic resonance imaging (MRI).

### Methods:

Retrospective review of 64 patients with biopsy-proven GCA. Contrast-enhanced cranial MRI was performed in 11 of these patients. The appearance of the ICAs was compared to normal controls.

### Results:

Three GCA patients demonstrated bilateral mural enhancement of the cavernous portion of the ICA. Patient 1 was a 64-year-old woman who presented with jaw pain and headaches, followed by vision loss to no light perception (NLP) in the left eye due to anterior ischemic optic neuropathy (ION) and visual field loss in the right eye due to posterior ION. She concurrently suffered a myocardial infarction, likely related to GCA. Patient 2 was a 79-year-old female who suffered anterior ION of the left eye and subsequently developed anterior and posterior ION of the right eye several days after initiation of high-dose steroid treatment, resulting in bilateral NLP vision. Patient 3 was a 75-year-old male who presented with bilateral sixth nerve palsies and headaches. In addition to the abnormal ICA findings, all patients demonstrated bilateral enhancement of the orbital fat or optic nerve sheaths.

### Conclusions:

Mural ICA enhancement on MRI is not a typical finding in GCA, but its presence may portend a more severe disease course and warrant aggressive treatment.

**References:** None.

**Keywords:** Vascular Disorders, Neuroimaging, Neuro-Ophth & Systemic Disease, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Morning Glory Disk Anomaly Associated with Absence of Intracranial Internal Carotid Artery

Adriana P Grigorian<sup>1</sup>, Florin Grigorian

*University Hospitals Eye Institute, Cleveland, OH, USA*

### Introduction:

Morning glory disk anomaly (MGDA) has been associated with numerous other congenital malformations including intracranial vascular abnormalities(1). The latest have been noticed in as many as 45% of cases (2), ranging from segmental narrowing of the Circle of Willis to complete stenosis of the internal carotid artery. We report a case of MGDA associated with agenesis of the intracranial internal carotid artery (ICA).

### Methods:

A 13 year old female presented for progressive visual loss in the left eye. Her vision was best corrected to 20/20 OD and 20/25 OS (-1.25D OU). She had +1 RAPD OS with full color vision and normal, symmetrical anterior segment examinations. Fundus examination was normal except for an anomalous optic disk OS, funnel shaped with peripapillary atrophy and central gliosis overlying the central retinal vessels. The visual field was full OD and had an enlarged blind spot OS. A diagnosis of morning glory disk anomaly (MGDA) OS was made and MRI/MRA was ordered.

### Results:

MRI/MRA demonstrated complete absence of the right intracranial ICA with the right middle cerebral artery receiving flow directly from the basilar artery and the right anterior cerebral artery from the left anterior circulation. The rest of the circulation was normal without evidence of vascular stenosis or hypertrophied lenticulostriate vessels to suggest moyamoya.

### Conclusions:

To our knowledge this is the first case report of a congenitally absent intracranial portion of ICA associated with MGDA. Due to the high risk of vascular and structural brain anomalies and risk of stroke, bleeding and seizures, all patients with MGDA should undergo MRI/MRA or computerized tomographic angiography.

### References:

1. Hanson MR, Price RL, Rothner AD, Tomsak RL. Developmental anomalies of the optic disc and carotid circulation. A new association. J Clin Neuroophthalmol. 5(1):3-8, 1985.
2. Lenhart P.D., Lambert S.R., Newman N.J., et al: Intracranial vascular anomalies in patients with morning glory disc anomaly. Am J Ophthalmol. 142: pp. 644-650, 2006

**Keywords:** Congenital Disk Anomaly, Intracranial Vascular Malformation

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Measurement of Optic Nerve Sheath Diameter by CT, MRI and Ultrasound

Klara Landau<sup>1</sup>, Christian Giger-Tobler<sup>1,6</sup>, Johannes Eisenack<sup>1</sup>, David Holzmann<sup>2</sup>, Athina Pangalu<sup>3</sup>, Veit Sturm<sup>4</sup>, Hanspeter E Killer<sup>5</sup>, Gregor P Jaggi<sup>1,7</sup>

<sup>1</sup>University of Zurich, Department of Ophthalmology, Zurich, Switzerland, <sup>2</sup>University of Zurich, Department of ENL, Zurich, Switzerland, <sup>3</sup>University of Zurich, Department of Neuroradiology, Zurich, Switzerland, <sup>4</sup>Cantonal Hospital St.Gallen, Department of Ophthalmology St.Gallen, Switzerland, <sup>5</sup>Cantonal Hospital Aarau, Department of Ophthalmology Aarau, Switzerland, <sup>6</sup>Rücken-&Schmerz-Praxis Guemligen, Switzerland, <sup>7</sup>Augenarzt Obwalden Sarnen, Switzerland

### Introduction:

Quantification of the optic nerve sheath diameter (ONSD) is a promising approach for detection of elevated intracranial pressure. The comparability of current methods is unclear. The objective of this study was to assess the relationship between ONSD as measured with CT, MRI and ultrasound (US) in patients without known optic nerve disease or increased intracranial pressure.

### Methods:

15 patients (60.8 [y]  $\pm$ 16.73 SD; 7 female) with paranasal sinus pathology in whom CT and MRI were performed underwent ONSD measurements by US, as well as an ophthalmological examination. US-, CT- and MRI-derived maximal ONSD values 3mm behind the globe were compared.

### Results:

ONSD measured (n=30) by US (mean 6.2 [mm]  $\pm$ 0.84 SD) were significantly ( $p < 0.01$ ) higher than ONSD in CT (5.2  $\pm$ 1.11) or MRI (5.3  $\pm$ 1.14). There was no significant ( $p = 0.24$ ) difference but good correlation ( $p = 0.854$ ,  $p < 0.01$ ) between ONSD measured in CT and MRI. Those of US and CT ( $p = 0.662$ ,  $p < 0.01$ ) and US and MRI ( $p = 0.615$ ,  $p < 0.01$ ) showed a modest but significant correlation.

### Conclusions:

The comparability of ONSD measurements in patients without known optic nerve disease and assumed normal intracranial pressure appears to be given between CT and MRI while comparability between US and CT or MRI seems to be less reliable.

### References:

1. Soldatos T, Chatzimichail K, Papathanasiou M, et al. Optic nerve sonography: a new window for the non-invasive evaluation of intracranial pressure in brain injury. Emerg Med J 2009;26:630-4.
2. Watanabe A, Kinouchi H, Horikoshi T, et al. Effect of intracranial pressure on the diameter of the optic nerve sheath. J Neurosurg 2008;109:255-8.
3. Sutherland AI, Morris DS, Owen CG, et al. Optic nerve sheath diameter, intracranial pressure and acute mountain sickness on Mount Everest: a longitudinal cohort study. Br J Sports Med 2008;42:183-8.
4. Kimberly HH, Noble VE. Using MRI of the optic nerve sheath to detect elevated intracranial pressure. Crit Care 2008;12:181.
5. Killer HE, Jaggi GP, Flammer J, et al. Cerebrospinal fluid dynamics between the intracranial and the subarachnoid space of the optic nerve. Is it always bidirectional? Brain 2007;130:514-520.
6. Maude RR, Hossain MA, Hassan MU, et al. Transorbital Sonographic Evaluation of Normal Optic Nerve Sheath Diameter in Healthy Volunteers in Bangladesh. PLoS ONE 2013;8(12)
7. Bäuerle J, Schuchardt F, Schroeder L, et al. Reproducibility and accuracy of optic nerve sheath diameter assessment using ultrasound compared to magnetic resonance imaging. BMC Neurology 2013;13:187
8. Bland JM, Altman DG. Statistical methods for assessing agreement between two methods of clinical measurement. Lancet 1986;307-10.
9. Brown B. St. John. How safe is diagnostic ultrasonography. Can Med Assoc J, Vol. 131, August 15, 1984
10. Weigel M, Lagrèze WA, Lazzaro A, et al. Fast and quantitative high-resolution magnetic resonance imaging of the optic nerve at 3.0 tesla. Invest Radiol 2006, 41:83-86

**Keywords:** Optic Nerve Sheath Diameter, Computer Tomography, Magnetic Resonance Imaging, Ultrasound

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Occipital Partial Status Epilepticus With Abnormal MRI Imaging

Sonalee Kulkarni, John Cochran. *Inova Medical Group, Fairfax, VA, United States*

**INTRODUCTION:** A healthy female presented with acute visual phenomenon suspicious for complex migraine. Further work up revealed an occipital simple partial status epilepticus on EEG and changes on MRI brain.

**METHODS:** Case presentation.

**RESULTS:** A 30 year old woman presented with a three day history of seeing a large colorful and spinning beach ball intermittently appearing in her right peripheral vision associated with frontal headache, nausea and vomiting. Visual field testing revealed a right homonymous hemianopia. MRI brain showed subtle diffuse abnormal T2 and FLAIR signal involving the left occipital cortex and extending to the left parieto-occipital and temporo-occipital junctions which was more obvious on a follow up scan done about 10 days after symptom onset. Video-EEG monitoring confirmed electrographic seizures arising from the left posterior quadrant. Detailed laboratory tests including CSF studies were unremarkable. She was treated with levetiracetam. She stopped having clinical seizures. The EEG normalized about a month and a half since onset of symptoms. Abnormalities noted on the MRI brain completely resolved.

**CONCLUSIONS:** Complex migraine versus transient ischemic attack versus seizure is always in the differential of any transient neurologic deficit. An abnormal EEG helped in the diagnosis. MRI findings due to status epilepticus often suggest a combination of cytotoxic and vasogenic edema, however it is unclear why only certain patients show MRI changes.

### REFERENCES:

1. Walker MC, Smith SJ, Sisodiya SM, Shorron SD Case of simple partial status epilepticus in occipital epilepsy misdiagnosed as migraine: clinical, electrophysiological an MRI characteristics
2. *Epilepsia* 36(12): 1233-6, Dec1995
3. Tracy A. Milligan, Amir Zamani, Edward Bromfield Frequency and patterns of MRI abnormality due to status epilepticus Seizure, Volume 18 Issue 2 104-108, Mar 2009
4. Panayiotopoulos CP, Visual phenomenon and headache in occipital epilepsy: a review, a systematic study and differentiation from migraine 1(4) 205-6, Dec 1999

**Keywords:** none

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Neuro-Imaging Characteristics of Common Extraocular Prosthetic Devices

Samantha F Lu<sup>1</sup>, Banafsheh Salehi<sup>2</sup>, Karen Tong<sup>3</sup>, Ali Sepahdari<sup>2</sup>, An Huynh<sup>1</sup>, Sarah Beck<sup>4</sup>, Beatrice Wong<sup>1</sup>, Samantha Perea<sup>1</sup>, Terry Wood<sup>1</sup>

<sup>1</sup>Department of Ophthalmology, Loma Linda University Medical Center, Loma Linda, CA, USA, <sup>2</sup>Department of Radiology, University of California, Los Angeles, Los Angeles, CA, USA, <sup>3</sup>Department of Radiology, Loma Linda University Medical Center, Loma Linda, CA, USA, <sup>4</sup>Department of Pathology, University of Michigan Ann Arbor, MI, USA

### Introduction:

With more implantable ophthalmic devices available, there is an increased risk that implanted ophthalmic hardware will be misidentified on imaging studies performed for non-ophthalmic indications. The purpose of this study is to describe the neuroimaging characteristics of common extraocular prosthetic devices, including gold weight eyelid implants and common glaucoma drainage devices, and the complications associated with these devices.

### Methods:

A retrospective chart review was performed to identify patients who had undergone both an ophthalmic procedure involving an implanted device and a neuro-imaging study performed for a non-ophthalmic condition.

### Results:

**CT:** 1) Axial CT shows prominent beam hardening artifact resulting from the implanted gold eyelid weight. 2) Axial and coronal NECT displays a Molteno implant. 3) Axial NECT in a Ahmed valve implant. Note the plate as a dense curvilinear structure, and the valve as a small focus of low density. 4) Axial NECT of a Baerveldt drainage implant. Note the endplate as a dense curvilinear plate with fenestration.

**MRI:** 1) Sagittal and coronal MRI shows a gold weight sutured to the anterior tarsal plate of the left eyelid. 2) Coronal and axial MRI with and without contrast in a case of Ahmed valve show hypointense fluid collection around the the plate of the valve within the conjunctival pocket. No abnormal enhancement is seen.

**Complications:** 1) Infection is a complication of implanted valves. MRI is the best imaging modality for this diagnosis. Axial MRI show abnormal enhancement involving the preseptal and retrobulbar regions. 2) Giant reservoir formation has been reported with Ahmed valve. Large reservoirs may cause mass effect and impaired extraocular muscle movements. Coronal and sagittal NECT images show a relatively large fluid collection surrounding the Ahmed valve.

### Conclusions:

These cases demonstrate the different appearances of gold weight eyelid implants and multiple glaucoma drainage devices. Increased communication and cross-specialty educational events between ophthalmologists and radiologists may result in more accurate interpretation of neuro-imaging studies that involve implanted ophthalmic devices.

### References:

1. Reiter M, Schwoppe R, Walker K, et al. Imaging of glaucoma drainage devices. J Comput Assist Tomogr 2012; 36(2): 277-9
2. Kemp PS, Allen RC, Kwon YH. Giant Reservoir Formation from Ahmed Seton Valve. <http://webeye.ophth.uiowa.edu/eyeforum/cases/150-Giant-Seton-Reservoir.htm>
3. Pirouzian A., Demer J.L. Clinical findings following Ahmed glaucoma valve™ implantation in pediatric glaucoma. Clin. Ophthalmol. 2008;2(1):123-127
4. Elizenda M, Ceballos and Richard K. Parrish II. Plain Film Imaging of Baerveldt Glaucoma Drainage Implants. AJNR 2002 23: 935-937

**Keywords:** Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## A Lion In The Bush

Padmaja Sudhakar<sup>1</sup>, Muhammad Zafar<sup>1</sup>, Kimberly Jones<sup>1</sup>, Flavius Raslau<sup>2</sup>, Janna Neltner<sup>4</sup>, Sachin Kedar<sup>3</sup>

<sup>1</sup>University Of Kentucky, Dept Of Neurology, Lexington, KY, USA, <sup>2</sup>University Of Kentucky, Dept of Neuroradiology, Lexington, KY, USA, <sup>3</sup>University Of Nebraska Medical Center, Dept of Neurology, Omaha, NE, USA, <sup>4</sup>University of Kentucky, Department of Pathology Lexington, KY, USA

### Introduction:

Extrarenal rhabdoid tumors of the thorax and mediastinum are rare, highly aggressive neoplasms with a poor prognosis. We describe a 3 month old child who presented with left arm weakness and left Horner's syndrome from an extrarenal rhabdoid tumor at the left thoracic inlet.

### Methods:

Single case report

### Results:

A 3 month old girl presented with left arm weakness noticed 4 days earlier and left upper eyelid swelling noted 11 days earlier. CT head and left clavicle Xray elsewhere did not reveal abnormalities. She was born full term via difficult labor and vacuum suction. Developmental milestones were appropriate for age. An inflammatory verrucous epidermal nevus (ILVEN) involving her left gluteal region was noticed two weeks after birth. At presentation the infant had flaccid left arm weakness with reduced deep tendon reflexes. Neuro-ophthalmic exam revealed central, steady and maintained fixation in both eyes, a left Horner's pupil without heterochromia and normal eye exam otherwise. When crying, flushing was noted only on the right half of her face. A general exam revealed a palpable non-tender firm left supraclavicular neck mass that was missed by the emergency room doctors. Contrast enhanced neck MRI revealed a large solid enhancing mass measuring 5.5 x 4 X 7 cm at the thoracic inlet that encroached the left C7-T1 neural foramen causing mild cord deformity, engulfed the left common carotid artery and large pulmonary metastasis. Carcinoembryonic antigen, alpha fetoprotein, B- HCG, urine homovanillic acid and vanillylmandelic acid levels were normal. Biopsy showed extra-renal malignant rhabdoid tumor with lymphovascular invasion. Despite starting chemotherapy she developed progressive respiratory failure from pneumothorax with increasing tumor burden. Care was withdrawn and she passed away after terminal extubation.

### Conclusions:

Extrarenal Rhabdoid tumors are highly aggressive tumors which may have metastatic disease at the time of presentation. It remains uncertain if earlier referral could have changed the prognosis.

### References:

1. Sparano A, Kreiger P, Kazahaya K. Malignant rhabdoid tumor of the parapharyngeal space. Ear Nose Throat J. 2009 Mar;88(3):E24-6.
2. Fridley JS, Chamoun RB, Whitehead WE, Curry DJ, Luerssen TG, Adesina A, Jea A. Malignant rhabdoid tumor of the spine in an infant: case report and review of the literature. Pediatr Neurosurg. 2009;45(3):237-43.
3. Garcés-Iñigo EF, Leung R, Sebire NJ, McHugh K. Extrarenal rhabdoid tumours outside the central nervous system in infancy. Pediatr Radiol. 2009 Aug;39(8):817-22. doi: 10.1007/s00247-009-1288-4.
4. Mahoney NR, Liu GT, Menaeker SJ, Wilson MC, Hogarty MD, Maris JM. Pédiatric horner syndrome: etiologies and roles of imaging and urine studies to detect neuroblastoma and other responsible mass lesions. Am J Ophthalmol 2006; 142: 651-659.

**Keywords:** Extrarenal Rhabdoid Tumor, Horner's Syndrome, Inflammatory Verrucous Epidermal Nevus, Metastasis, Pneumothorax

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Why the Delay in Diagnosis? Increased Time From Symptom Onset to Diagnosis in Blepharospasm: A Prospective, Clinic-Based Study

Kristen E Dunbar<sup>1</sup>, Michael M Johns III<sup>2</sup>, Hyder A Jinnah<sup>3</sup>, Ami R Rosen<sup>3</sup>, Laura J White<sup>2</sup>, Ted H Wojno<sup>4</sup>

<sup>1</sup>Tufts Medical Center/New England Eye Center/Ophthalmology, Boston, MA, USA, <sup>2</sup>Emory University Hospital/Otolaryngology, Atlanta, GA, USA, <sup>3</sup>Emory University Hospital/Neurology, Atlanta, GA, USA, <sup>4</sup>Emory University Hospital/Emory Eye Center/Ophthalmology Atlanta, GA, USA

### Introduction:

Blepharospasm is a debilitating dystonia shown to affect quality of life, mood, and sleep quality [1,2,3]. With proper diagnosis, early treatment with botulinum toxin dramatically improved symptoms and quality of life [4,5]. Older studies showed unacceptable lag times from symptom onset to diagnosis prolonging the time to acceptable treatment [6,7]. Despite improved education, we noticed many patients presenting with incorrect diagnoses or on inappropriate treatments. The aim of this study was to explore time from symptom onset to correct diagnosis to evaluate how physician education has changed since initial studies in the United States.

### Methods:

A structured questionnaire was administered to consecutive patients with blepharospasm presenting to outpatient tertiary care clinics. Information was gathered from each participant including length of time from symptom onset to diagnosis, types and numbers of providers seen prior to diagnosis, and treatments administered prior to receiving botulinum toxin.

### Results:

The study enrolled 72 patients with blepharospasm. Average age was 66.2, (F:M=2.4:1). Mean time from symptom onset to diagnosis was 25.6 months--the delay was over 3 years in 21% of patients. At initial presentation with symptomatology, only 2 patients (3%) were diagnosed correctly, mean number of physicians seen before diagnosis was 3.1. For those diagnosed in the 5 years prior to enrollment, the lag time was 25.5 months with no statistically significant difference before this time (t-value=.193, t>.001).

### Conclusions:

Despite classic presenting signs easily identifiable by simple history and examination, a significant lag time to diagnosis exists. It is common for patients with this disease to see multiple providers, receive ineffective treatments, and suffer from symptoms for over a year prior to correct diagnosis. Improved education and awareness with help patients obtain correct diagnosis and treatment more efficiently.

### References:

1. Adams WH, Digre KB, Patel BC, et al, The evaluation of light sensitivity in benign essential blepharospasm, *Am J Ophthalmol*, 142(1), 82-87, 2006.
2. Paus S, Gross J, Moll-Muller M, et al, Impaired sleep quality and restless legs syndrome in idiopathic focal dystonia: a controlled study, *J Neurol*, 258(10), 1835-40, 2011.
3. Hall TA, McGwin G, Jr., Searcey K, et al, Health-related quality of life and psychosocial characteristics of patients with benign essential blepharospasm, *Arch Ophthalmol*, 124(1), 116-119, 2006.
4. Costa PG, Aoki L, Saraiva FP, Matayoshi S, Botulinum toxin in the treatment of facial dystonia: evaluation of its efficacy and patients' satisfaction along the treatment, *Arq Bras Oftalmol*, 68(4), 471-474, 2005.
5. Hall TA, McGwin G, Jr., Searcey K, et al, Health-related quality of life and psychosocial characteristics of patients with benign essential blepharospasm, *Arch Ophthalmol*, 124(1), 116-119, 2006.
6. Jankovic J, Ford J, Blepharospasm and orofacial-cervical dystonia: clinical and pharmacological findings in 100 patients, *Ann Neurol*, 13(4), 402-411, 1983.
7. Kowal L, Davies R, Kiely P, Facial muscle spasms: an Australian study, *Aust N Z J Ophthalmol*, 26(2), 123-128, 1998.

**Keywords:** Blepharospasm, Dystonia

**Financial Disclosures:** Grants as discussed in the abstract.

**Grant Support:** 1. U54 NS065701 from the Office of Rare Disease Research and the National Institutes of Neurological Disorders & Stroke 2. Unrestricted grant from Research to Prevent Blindness, Inc., New York, USA

### Atypical Presentation of Orbital Lymphangioma.

Haydée S Martinez<sup>1,2</sup>, Macarena Clementi<sup>1,2,4</sup>, Mirta Arana<sup>1,2</sup>, Maria L Braccia Gancedo<sup>1,2,3</sup>, Mariana de Virgiliis<sup>1,3</sup>, Pablo I Perez Vega<sup>1,3</sup>, Luciana Iacono<sup>3</sup>

<sup>1</sup>Hospital de Clínicas José de San Martín, Buenos Aires, Argentina, <sup>2</sup>Universidad de Buenos Aires, Buenos Aires, Argentina,

<sup>3</sup>Hospital Oftalmológico Dr. Pedro Lagleyze, Buenos Aires, Argentina, <sup>4</sup>Clinica de Ojos Dr Nano San Miguel, Argentina

#### Introduction:

Orbital lymphangiomas are vascular orbital anomalies, representing less than 4% of all space-occupying orbital lesions, which most frequent forms of presentation are proptosis and hemorrhagic episodes.

#### Methods:

Review of medical history of a patient with pulsatile progressive enophthalmos associated with compressive optic neuropathy and diplopia. Review of the literature.

#### Results:

A 29-year-old woman who developed a long-term left orbital enophthalmos, but in the last 4-months became progressive pulsating enophthalmos, positive to Valsalva manoeuvre, associated with restrictive diplopia and compressive optic neuropathy, with best corrected visual acuity (VA) 1.0 in the right eye, and 0.7 in her left eye with a clear RAPD and color vision impairment. Orbital CT and MRI scanning were performed, obtaining lytic signs in orbit, in communication with anterior cranial cavity, with an isodense extra and intraconal contrast-enhanced tissue with tortuous structure and intralesional calcifications. Due to the intensification of the symptoms and signs in the physical examination, RAPD and worsening in VA due to the optic nerve compression, she was remitted to neurosurgery to undergo surgical treatment immediately. Since it was a great diameter lesion, associated with oculomotor impairment and optic neuropathy, transcranial access was performed. The patient rapidly recovered from her compressive optic neuropathy, esthetical improvement of her pulsatile enophthalmos, but diplopia persisted. The histopathology exam revealed the tumor was a cystic lymphangioma, and CT scanning performed after surgery revealed residual lesional tissue.

#### Conclusions:

Although orbital lymphangiomas are benign tumors, and the most frequent form of presentation is with proptosis, in this case the onset was atypical. The early diagnosis of this entity should prevent further complications not only in the VA, but in the risk of morbidity that they may lead to if not treated.

#### References:

1. Lagrèze W1, Metzger M, Rössler J. Sclerotization of orbital lymphangioma with OK-432. *Ophthalmology*. 111(5):479-81. 2014
- Gooding C1, Meyer D. Intralesional Bleomycin: A Potential Treatment for Refractory Orbital Lymphangiomas. *Ophthalm Plast Reconstr Surg*. 30(3):e65-7. 2013
2. Gandhi NG1, Lin LK, O'Hara M. Sildenafil for pediatric orbital lymphangioma. *JAMA Ophthalmol*.;131(9):1228-30. 2013
3. Mowatt L1, Crossman G. Orbital lymphangioma in a child: a diagnostic dilemma. *West Indian Med J*. 2012 Oct;61(7):764-6.
4. Simas N1, Farias JP2. Orbital Lymphangiomas: Surgical Treatment and Clinical Outcomes. *World Neurosurg*. 81(5-6):842.e5-10. 2013

**Keywords:** Orbit/Ocular Pathology, Tumors, Ocular Motility, Optic Neuropathy, Adult Strabismus With A Focus On Diplopia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 193

### Prospective Assessment of Peri-Oral Weakness Following Peri-Orbital Botulinin Toxin for Blepharospasm.

Alexander J Hartmann<sup>1</sup>, Michael S Lee, Andrew R Harrison

University of Minnesota, Minneapolis, MN, USA

#### Introduction:

In our experience performing peri-orbital injections of botulinin toxin for benign essential blepharospasm (BEB) we have encountered three patients who have experienced persistent ptosis of the upper lip. Notably, these patients have not had injection of any of the apparently weakened peri-oral musculature. In addition, this appears to be possibly chronic, as the individuals affected have continued to experience lip ptosis even as their blepharospasm symptoms return and they require additional injection, typically at 3-4 month intervals. There are reports of botulinin toxin unmasking previously subclinical myasthenia, although these patients we are describing have never developed any other symptoms of myasthenia. There is a single case series identified in our literature review of three patients who experienced transient lip ptosis following injections done for cosmetic treatment of "crow's feet." (Matarasso 2001). To our knowledge, a prospective study to assess for the incidence of this particular side effect has not been undertaken.

#### Methods:

Patients are enrolled from the eye clinic during routine visits. Patients with conditions other than BEB are included as controls. Patients with myasthenia, history of stroke, history of Bell's Palsy, and other conditions affecting the facial muscles are excluded from the study. Photographs of the patients at rest and in six different facial expressions are taken, to examine frontalis, orbicularis oculi, risorius, zygomaticus, and orbicularis oris muscles. The SunnyBrook scale (Neely 2010) is used in accompaniment of the photos to assess each patient's degree of facial weakness qualitatively. Quantitative assessment is graded using the Lip Length and Snout Indices (Janson, 1990). Observers will be blinded to which patients are from the BEB populations and which are from non BEB controls.

#### Results:

This study is presently ongoing.

#### Conclusions:

No conclusions are known yet. Our goal is to catalogue the incidence and severity of this as yet largely unreported side effect.

#### References:

1. Matarasso SL, Matarasso A. "Treatment guidelines for botulinum toxin type A for the periocular region and a report on partial upper lip ptosis following injections to the lateral canthal rhytids." *Plast Reconstr Surg.* July 108(1):208-14. 2001.
2. Janson C, Jennekens FG, Wokke JH, Leppink GJ, Wignne HJ. "Lip-length and snout indices: methods for quantitative assessment of peri-oral facial muscle strength." *J Neurol Sci.* July 97(2-3):133-42. 1990.
3. Neely JG, Cherian NG, Dickerson CB, Nedzelski JM. "Sunnybrook Facial Grading System: Reliability and Criteria for Grading." *Laryngoscope.* May 120:1038-1045. 2010.

**Keywords:** Blepharospasm, Botulinin, Peri-Oral, Weakness

**Financial Disclosures:** Andrew Harrison and Michael Lee each own 1/3 of a company they formed called Neuro-Ophthalmix, LLC

**Grant Support:** None.

## Poster 194

### Vertical Diplopia And Ptosis From Removal Of The Orbital Roof In Pterional Craniotomy

Jonathan C. Horton<sup>1</sup>, Michael T. Lawton<sup>2</sup>, Michael W. McDermott<sup>2</sup>, Shilpa J. Desai<sup>1</sup>

<sup>1</sup>UCSF/Ophthalmology, San Francisco, CA, USA, <sup>2</sup>UCSF/Neurosurgery, San Francisco, CA, USA

#### Introduction:

We describe a newly recognized clinical syndrome consisting of ptosis, diplopia, and limited vertical ductions that occurs following orbital roof removal during orbital-zygomatic-pterional craniotomy.

#### Methods:

Review of patient records of a single neuro-ophthalmologist from 1998 to 2013 identified 8 patients with persistent vertical diplopia after surgery. In all cases the orbital roof was removed to gain better access to the anterior cranial fossa. The operative report, CT or MRI imaging, intracranial pathology, neurovisual exam, ocular deviation, and degree of ptosis were recorded. Subsequent treatment and patient outcome were documented.

#### Results:

Eight patients had neuro-ophthalmic findings after pterional craniotomy for meningioma removal or aneurysm clipping. The cardinal features were ptosis, limited elevation and hypotropia. Three patients also had limitation of downgaze and two had limitation of abduction. Imaging showed loss of the fat layers which normally envelop the superior rectus/levator palpebrae superioris. The muscles appeared attached to the defect in the orbital roof. Ptosis and diplopia developed in two patients despite Medpor titanium mesh implants. Deficits in all patients showed spontaneous improvement. Two patients underwent a levator advancement for ptosis repair. In three patients an inferior rectus recession using an adjustable suture was performed to treat vertical diplopia. Follow-up a mean of 6.5 years later revealed that all patients had a slight residual upgaze deficit, but alignment was orthotropic in primary gaze.

#### Conclusions:

After pterional craniotomy, ptosis, diplopia and vertical gaze limitation can result from tethering of the superior rectus/levator palpebrae superioris complex to the surgical defect in the orbital roof. Lateral rectus function is sometimes compromised by muscle attachment to the lateral orbital osteotomy. This syndrome occurs in about 1% of patients after removal of the orbital roof and can be treated, if necessary, by prism glasses or surgery.

**References:** None.

**Keywords:** Ptosis, Diplopia, Pterional Craniotomy, Gaze Restriction

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** National Eye Institute

**Poster 195**

**Treatment of Rivaroxaban Associated Orbital Hemorrhage**

Marilyn C Kay<sup>1</sup>

*University of Wisconsin, Madison, WI, USA*

**Introduction:**

An elderly man awoke with severe pain and bruising of right periocular area. He had taken his usual rivaroxaban the night before and reported no trauma. CT scanning showed an orbital hemorrhage. His ocular pressure was in the mid thirties and did not respond to glaucoma medications or subsequent canthotomy. Treatment with an antagonist to rivaroxaban was carried out after transfer to my institution. Discussion of the mechanism of rivaroxaban's efficacy and the use of an antagonist drug that is usually used to reverse cardiac-related anticoagulation will be discussed.

**References:** None.

**Keywords:** Rivaroxaban And Orbit Hemorrhage

**Financial Disclosures:** The author had no disclosures.

**Grant Support:** None.

## **Stonewalled: Bilateral Sequential Vision Loss in a Peritoneal Dialysis Patient**

Angelina Espino Barros Palau<sup>1</sup>, Michal L Morgan<sup>2</sup>, Patricia Chevez-Barrios<sup>2</sup>, Sushma Yalamanchili<sup>2</sup>, Andrew G. Lee<sup>2</sup>.

<sup>1</sup>ITESM, Monterrey, Mexico; <sup>2</sup>Houston Methodist Hospital, Houston, TX, United States

### **Introduction:**

We present a patient with biopsy proven calcific uremic arteriopathy causing a bilateral anterior ischemic optic neuropathy (AION).

### **Methods:**

A 66-year-old white man who was hospitalized for aortic stenosis leading to congestive heart failure presented with bilateral sequential painless vision loss of the left eye and then the right eye separated by 6 weeks. He had a history of end-stage renal disease on peritoneal dialysis secondary to essential hypertension and type 2 diabetes mellitus as well as left lower extremity deep vein thrombosis, secondary hyperparathyroidism, hyperlipidemia, hypothyroidism and benign prostatic hypertrophy. Outside ophthalmology found erythrocyte sedimentation rate (ESR) of 150 and treated with intravenous and then oral corticosteroids following loss of left eye vision. He had also developed a necrotic penile ulcer 2 weeks prior to admission and firm skin nodules of the lower extremities while admitted with X-rays showing extensive vascular calcifications.

### **Results:**

Neuro-ophthalmological examination was remarkable for acuity 20/70 OD and count fingers OS with a right relative afferent pupillary defect. Visual field testing revealed inferior altitudinal and superior nasal step loss OD with an inferior nasal step OS. Fundus examination was remarkable for right optic nerve edema. ESR was 82 mm/hr with C-reactive protein of 1.42 mg/dL. Prednisone was increased to 80 mg daily empirically. Temporal artery biopsy could not be performed due to other illnesses. MRI of the brain and orbits and lumbar puncture were unremarkable. Review of laboratory results revealed worsened phosphatemia compared to one year prior with a calcium-phosphorus product of 63 mg\*mg/dL/dL on admission. The patient died from heart failure while hospitalized. Subsequent autopsy showed extensive vascular calcification. These findings confirmed the presumptive diagnosis of calcific uremic arteriopathy imitating giant cell arteritis.

**Conclusions:** Calcific uremic arteriopathy should be suspected in chronic kidney disease patients presenting with anterior ischemic optic neuropathy.

### **References:**

1. Shah MA, Roppolo MW. Calciphylaxis: temporal artery calcification preceding widespread skin lesions and penile necrosis. *Case Rep Nephrol.* 2012;2012:309727.
2. Korzets A, Marashek I, Schwartz A, Rosenblatt I, Herman M, Ori Y. Ischemic optic neuropathy in dialyzed patients: a previously unrecognized manifestation of calcific uremic arteriopathy. *Am J Kidney Dis.* 2004 Dec;44(6):e93-7.

**Keywords:** Calcific Uremic Arteriopathy, Chronic Kidney Disease, Vascular Calcification, Ischemic Optic Neuropathy, Giant Cell Arteritis.

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 197

### Allergic Fungal Sinusitis Mimicking Thyroid Orbitopathy

Lina Nagia<sup>1</sup>, Michael Vaphiades<sup>1</sup>, Bradford Woodworth<sup>2</sup>

<sup>1</sup>University of Alabama Birmingham/Department of Ophthalmology, Birmingham, AL, USA, <sup>2</sup>University of Alabama Birmingham/Department of Surgery, Birmingham, AL, USA

#### Introduction:

22-year-old African-American man presented to our clinic with complaint of tearing and proptosis of both eyes along with occasional blur in the right eye. His medical history is significant for asthma, which is well controlled. His mother has a history of Grave's disease with exophthalmos. Examination was notable for optic neuropathy of the right eye and significant exophthalmos both eyes.

#### Methods:

Case Report.

#### Results:

Thyroid labs were normal. Neuro-imaging revealed expansion and opacification of all paranasal sinuses with intracranial and intraorbital extension. Patient underwent oral steroid treatment followed by nasal endoscopy procedure with sinusotomy, sinusotomy and mucous membrane removal. Mucous samples sent to pathology for analysis. GMS stain highlighted abundant branching fungal hyphae.

#### Conclusions:

The purpose of this submission is to present a case of progressive exophthalmos with associated optic neuropathy caused by advanced allergic fungal sinusitis. This clinical entity, while rare, can cause serious ophthalmic complications that can be reversed if prompt diagnosis and treatment are initiated. The unique radiographic appearance of this process is also highlighted. Allergic fungal rhinosinusitis is a form of fungal sinusitis that is noninvasive and has higher incidence in the southern and southwestern US. While diagnostic criteria have been established, treatment both medical and surgical remains a challenge.

**References:** None.

**Keywords:** Orbit Pathology, Allergic Fungal Sinusitis, Optic Neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 198

### Optic Nerve Sheath Meningioma. No Longer Impossible But Still Difficult.

T. Ben Ableman<sup>1</sup>, Steven A. Newman

*University of Virginia, Charlottesville, VA, USA*

#### Introduction:

In 1977 while talking about the difficulty in identifying small meningiomas involving the orbital apex, optic nerve sheath, and canal Susac, Smith, and Walsh wrote an article entitled "The Impossible Meningioma." The advent of CT scanning (allowing identification of enlarged optic nerve sheath and calcification) substantially improved the situation and the subsequent advent of MRI scanning with gadolinium and fat sat further advanced diagnostic armamentarium. In spite of these advances, optic nerve sheath meningiomas are often discovered after substantial delay. The fact that fractionated radiation therapy may substantially improve the prognosis underlies the importance of timely diagnosis and referral.

#### Methods:

Retrospective analysis of 7 cases of optic nerve sheath meningiomas with substantial diagnostic delay

#### Results:

In several cases significant progressive visual loss occurred during the 1-10 year delay in diagnosis. Some of these reversed with radiation therapy. Several conclusions could be drawn regarding the reason for the diagnostic delay.

#### Conclusions:

1. Lack of ordering imaging studies often due to an alternative diagnosis such as AION or papillitis. 2. Lack of appropriately directed imaging (head scans instead of orbital scans, the lack of employment of fat sat and gadolinium). 3. Miss-reading requiring reinterpretation. 4. Misinterpretation of OCT data because blocked axonal transport masks development of optic atrophy.

#### References:

1. Susac JO, Smith JL, Walsh FB "The impossible meningioma". Arch Neurol. 34(1):36-8, Jan 1977.
2. Turbin RE, Thompson CR, Kennerdell JS, Cockerham KP, Kupersmith MJ). "A long-term visual outcome comparison in patients with optic nerve sheath meningioma managed with observation, surgery, radiotherapy, or surgery and radiotherapy". Ophthalmology 109 (5): 890-9; discussion 899-900, May 2002.
3. Jackson A, Patankar T, Laitt RD " Intracranial Optic Nerve Meningioma: A Serious Diagnostic Pitfall" . AJNR Am J Neuroradiol 24:1167-1170, June 2003.
4. Lindblom B, Truwit CL, Hoyt WF "Optic nerve sheath meningioma. Definition of intraorbital, intracanalicular, and intracranial components with magnetic resonance imaging". Ophthalmology 99 (4): 560-6, April 1992.
5. Miller, NR (2004). "Primary tumours of the optic nerve and its sheath". Eye 18, 1026-37

**Keywords:** Optic Nerve Sheath Meningioma, MRI, OCT, Radiation Therapy, Imaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 199

### An Advanced Pancoast Tumor Masquerading as Congenital Hereditary Ptosis

Deborah C Parish<sup>1</sup>, Jenny Yu<sup>2</sup>

<sup>1</sup>UPMC / Oculoplastics, Pittsburgh, PA, USA, <sup>2</sup>UPMC / Oculoplastics, Pittsburgh, PA, USA

#### Introduction:

We present a patient with a stage IV Pancoast tumor which masqueraded as worsening congenital hereditary ptosis.

#### Methods:

A 69-year-old male presented complaining of progressively worsening drooping of his eyelids. He stated that his eyelids had always been droopy and weak, since early childhood. Family history was significant for multiple family members with ptosis and resultant surgical repair. On examination, anisocoria with subtle miosis of the left eye and ptosis of both eyes (left > right), was found, with a measured MRD1 of -1 OD and an MRD1 of -4 OS. Apraclonidine testing confirmed Horner's syndrome. Slit lamp examination showed bilateral nuclear sclerosis with narrow angles by Van Herick's and dilated fundoscopic exam was unremarkable. The patient's past social history was significant for smoking. The past medical history was significant for a recent episode of bronchitis with a negative CXR that was treated successfully with antibiotics, and for a recent palpable nuchal lymph node.

#### Results:

Chest CT was suspicious for an apical sulcus tumor in the medial aspect of the left apex. PET scan was remarkable for FDG avid left apical soft tissue density, several FDG avid thoracic lymphadenopathies, multiple FDG avid bilateral pleural nodules, multiple FDG avid mesenteric/retroperitoneal lymph nodes in the abdomen and pelvis, FDG avid peritoneal nodules, and intensely FDG avid L3 and L4 vertebral body lesions that were all consistent with metastatic disease. A biopsy of the left scalene axillary node revealed a granulomatous adenocarcinoma that was cytokeratin and thyroid transcription factor-1 positive, further suggesting a Pancoast tumor as the cause of this primary pulmonary adenocarcinoma. The patient was determined to have a stage IV pancoast tumor.

#### Conclusions:

This stage IV Pancoast tumor likely presented at such a late stage, due to the clinical picture of this patient with long standing ptosis and dark iris pigmentation that masked his diagnosis.

#### References:

1. Davagnanam I, Fraser CL, Miszkiet K, Daniel CS, Plant GT. Adult Horner's syndrome: a combined clinical, pharmacological, and imaging algorithm. *Eye*.2013;27:291–298. doi: 10.1038/eye.2012.281.
2. Almog Y, Gepstein R, Kesler A. Diagnostic value of imaging in horner syndrome in adults. *J Neuroophthalmol*.2010;30 1:7–11.
3. Walton KA, Buono LM. Horner's syndrome. *Curr Opin Ophthalmol*. 2003;14(6):357–363.
4. Foroulis CN, Zarogoulidis P, Darwiche K, et al; Superior sulcus (Pancoast) tumors: current evidence on diagnosis and radical treatment. *J Thorac Dis*. 2013;5(4):342-358.

**Keywords:** Pancoast, Ptosis, Anisocoria, Adenocarcinoma

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None

## Poster 200

### It's All Going Dark!

Tarek A Shazly<sup>1</sup>, Amanda Way<sup>1</sup>, Lea Ann Lope<sup>1,2</sup>, Ellen B Mitchell<sup>1,2</sup>

<sup>1</sup>University of Pittsburgh, Department of Ophthalmology, Pittsburgh, PA, USA, <sup>2</sup>Children's Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

#### Introduction:

Restricted diffusion on diffusion-weighted imaging (DWI) has been noted in brain parenchymal lymphomas. It has been reported in ischemic, inflammatory and adult compressive optic neuropathies, but has not previously been reported in pediatric lymphomatous optic neuropathy.

#### Methods:

Case report and review of literature.

#### Results:

We present a case of a 6-year-old girl with past medical history of Hemophagocytic lymphohistiocytosis (HLH) and peripheral T-cell lymphoma who reported "blurry vision in her left eye" for 2 weeks. Her corrected VA was 20/20 OD and 20/200 OS with decreased color vision and afferent papillary defect in the left eye. Dilated fundus examination revealed unremarkable fundus appearance in the right eye with florid left optic nerve swelling, diffuse pallor and peri-papillary hemorrhages. MRI of the brain and orbits showed left optic nerve thickening and enhancement extending to the orbital apex. Axial DWI reveals high signal in the left intraorbital optic nerve while ADC map demonstrating a corresponding region of darkness indicating restricted diffusion. Lumbar puncture revealed opening pressure of 38.5 cm H<sub>2</sub>O, negative for malignant cells or infectious agents. Diamox 250 mg bid was initiated. Patient received 6 Gy in 3 fractions to the left orbit and 2-3 weeks later received whole brain irradiation getting 18 Gy in 12 fractions with an additional 6 Gy in 3 fraction boost to the left orbit with weekly intrathecal triple chemotherapy. On 2 week follow up, her VA improved to 20/20 OD 20/60 OS with the persistence of left APD and left dyschromatopsia.

#### Conclusions:

Restricted diffusion in the optic nerve has been previously suggested to augur a poor visual outcome, even in inflammatory or neoplastic conditions. Our case suggests that prompt diagnosis and treatment of infiltrative neuropathy may result in visual recovery in a pediatric patient even in the presence of restricted diffusion.

#### References:

1. Sudhakar, Padmaja, Francisco Rivas Rodriguez, and Jonathan D. Trobe. "MRI restricted diffusion in lymphomatous optic neuropathy." *Journal of Neuro-Ophthalmology* 31.4 (2011): 306-309.
2. Schaefer PW, Grant PE, Gonzalez RG. Diffusion-weighted MR imaging of the brain. *Radiology*. 2000;217:331–345
3. Yamasaki F, Kurisu K, Satoh K, Arita K, Sugiyama K, Ohtaki M, Takaba J, Tominaga A, Hanaya R, Yoshioka H, Hama S, Ito Y, Kajiwarra Y, Yahara K, Saito T, Thohar MA. Apparent diffusion coefficient of human brain tumors at MR imaging. *Radiology*. 2005;235:985–991
4. Guo AC, Cummings TJ, Dash RC, Provenzale JM. Lymphomas and high grade astrocytomas: comparison of water diffusibility and histologic characteristics. *Radiology*. 2002;224:177–183
5. Kitis O, Altay H, Calli C, Yuntun N, Akalin T, Yurtseven T. Minimum apparent diffusion coefficients in the evaluation of brain tumors. *Eur J Radiol*. 2005;55:393–400
6. Chong, Kok Wee, et al. "Hemophagocytic lymphohistiocytosis with isolated central nervous system reactivation and optic nerve involvement." *Journal of child neurology* 27.10 (2012): 1336-1339.
7. Lee, Edward W., and William F. Mieler. "Ocular findings in a patient with hemophagocytic syndrome." *Archives of ophthalmology* 124.11 (2006): 1656-1658.

**Keywords:** Lymphoma, Ischemia, lymphohistiocytosis, Hemophagocytic

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 201

### ***Bartonella Henselae*-Associated Neuroretinitis with Orbital Intraconal and Optic Nerve Sheath Involvement**

Ann Shue<sup>1</sup>, Joseph N Martel, Tarek A Shazly, Ailee M Laham, George Trichonas, Gabrielle R Bonhomme

University of Pittsburgh Medical Center/Department of Ophthalmology, Pittsburgh, PA, USA

#### **Introduction:**

We present clinical, radiological and histopathological findings of a patient with neuroretinitis, with positive *Bartonella henselae* IgM and an indeterminate QuantiFERON-TB Gold In-Tube (QFT-GIT) test, who developed orbital intraconal and optic nerve sheath inflammation. This is the first report of intraconal inflammation prompting optic nerve sheath fenestration (ONSF) in association with *B. henselae* infection.

#### **Methods:**

Retrospective chart and literature review.

#### **Results:**

A 48-year-old male presented with a central scotoma of his left eye (OS). Past medical history included ulcerative colitis, primary sclerosing cholangitis, and recent right eye scleritis and prostatitis. One month prior, an inguinal lymph node biopsy for diffuse lymphadenopathy was negative for malignancy. Visual acuity was count fingers OS, without a frank relative afferent pupillary defect (RAPD). Fundus exam revealed significant disc edema, an inferotemporal perivascular choroidal mass and a serous retinal detachment OS. Subsequent MRI brain demonstrated enhancement of the left intraconal soft tissue and anterior optic nerve sheath. The following day, his left eye exhibited an RAPD, omnidirectional limitation of movement, elevated intraocular pressure, ptosis, proptosis and conjunctival injection with chemosis. Given the patient's severe loss of vision and progression, he underwent ONSF and orbital biopsy. Intraoperatively, there was a paucity of intraconal fat, with dense fibrinous white tissue and cartilaginous-like material in the intraconal orbit. Histopathology revealed dense fibroconnective tissue. Extensive laboratory evaluation revealed elevated *B. henselae* IgM titers (1:40, then 1:80). IgG was negative. Repeated indeterminate QFT-GIT results due to a low response in the mitogen control tube suggested an immunocompromised state. The late fundusoscopic finding of a macular star was consistent with the presence of a neuroretinitis. Previous reports have described inflammatory masses of the optic nerve head and retrobulbar optic neuritis in bartonellosis, however none have reported intraconal soft tissue involvement.

#### **Conclusions:**

Intraconal soft tissue inflammation may be another vision-threatening manifestation of *B. henselae* infection.

#### **References:**

1. Buzzacco DM, Lubow M, Davidorf FH, Cebulla CM. Atypical cat scratch disease with vitritis, serous macular detachment, neuroretinitis, and retrobulbar optic neuritis. Graefes Arch Clin Exp Ophthalmol. 2013;251:1001-2.
2. Cunningham ET Jr., et al. Inflammatory mass of the optic nerve head associated with systemic Bartonella henselae infection. Arch Ophthalmol. 1997;115:1596-7.
3. Kawasaki A, Wilson DL. Mass lesions of the posterior segment associated with Bartonella henselae. Br J Ophthalmol. 2003;87:248-9.

**Keywords:** Neuro-Ophth And Infectious Disease, Orbit/Ocular Pathology, Optic Neuropathy, Retina, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 202

### Impact of Injection Site on Dose Escalation in the Treatment of Blepharospasm

Rachel G. Simpson<sup>1</sup>, S. Khizer Khaderi

*University of California, Davis, Sacramento, CA, USA*

#### **Introduction:**

Blepharospasm is a common debilitating condition that can limit a patient's ability to read, drive or work productively. While paralytic toxoid injections have been considered the treatment of choice for this condition, very little data exists to guide practitioners on the most effective ways to administer this therapy. We hypothesize injection site location may impact the dose amount required to achieve symptom resolution.

#### **Methods:**

We performed a retrospective review based on billing codes for botulinum toxin injections, cross-referenced with codes for patients with blepharospasm on the neuro-ophthalmology service at an academic institution from January 2009 to September 2014. Patients with <8 treatments were excluded from consideration. Toxoid injection sites were then categorized as either superior to the lid crease (Method 1), or between the superior lash line and lid crease (Method 2). Injection site, dose, and date of treatment were documented for each patient.

#### **Results:**

We identified 31 patients who met the selection criteria above. Our results revealed a statistically significant difference ( $p = 0.019$ ) between patients treated by Method 1 (30 dose escalations) compared to Method 2 (6 dose escalations). Furthermore, there was a trend of dose reduction in patients switched from Method 1 to Method 2, though the number of patients switched was too small to be significant.

#### **Conclusions:**

Preliminary data indicates that injection site selection impacts dosages for successful treatment of blepharospasm patients. Patients who were treated with Method 2 were able to maintain or decrease the amount of toxoid needed to achieve symptomatic improvement, whereas patients treated with Method 1 experienced more frequent escalations in injection amount. This study suggests a preferred method for botulinum toxoid injections for the effective treatment of blepharospasm.

**References:** None.

**Keywords:** Blepharospasm, Injections

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 203

### A Case of Nasopharyngeal Carcinoma Masquerading as Primary Orbital Tumour

Shantha Amrith<sup>1</sup>, Benjamin Boon Chuan Tan

*National University Hospital, Singapore, Singapore*

#### **Introduction:**

Nasopharyngeal Carcinoma (NPC), although rare in North America, is a common malignancy in Southeast Asia and Southern China. Ophthalmic manifestations of NPC include isolated or multiple cranial nerve palsies, orbital apex or cavernous sinus syndromes, papilloedema or unilateral optic disc swelling.

#### **Methods:**

We report a rare case of NPC masquerading as a primary orbital tumour.

#### **Results:**

The patient is a 34 year old Chinese gentleman who first presented at a foreign hospital with right sided facial numbness. He was seen by a Neurologist and the MRI (Brain) performed was reported as normal. He gradually developed symptoms of dry eye, diplopia and right proptosis over the next couple of months and an MRI (Orbits) was later performed. MRI (Orbits) showed a lesion in the right orbital apex, extending into the infratemporal fossa, pterygoid muscle and lateral sphenoid wall. A right lateral orbitotomy and biopsy was performed, with histology reported as squamous cell carcinoma. A right orbital exenteration was subsequently planned, following which the patient came to our centre to seek a second opinion. A multidisciplinary team was involved in the management of this patient at our centre. The otolaryngologist in the team performed a nasoendoscope and noted that the posterior nasal space was normal looking. A blind biopsy was nonetheless taken and subsequently returned positive for non-keratinizing undifferentiated NPC. The patient was subsequently treated with chemotherapy and radiotherapy.

#### **Conclusions:**

We report a rare case of NPC masquerading as a primary orbital tumour. A high level of suspicion should always be maintained for atypical presentations of primary orbital tumours.

**References:** None.

**Keywords:** Nasopharyngeal Carcinoma, Orbital Mass

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 204

### Visual Field and Graves Ophthalmopathy

Sabine Defoort<sup>1</sup>, Jean Philippe Woillez, Virginie Rufin

*Chru/Neuro Ophthalmology, Lille, France*

#### **Introduction:**

Whether primary or secondary to thyroid disease, Graves' ophthalmopathy is surprising by its evolution despite correct treatment. Transformation to acute mode may occur very quickly. Focal signs (edema, proptosis) and oculomotor signs (diplopia) are likely to alert physician. Visual field, as the indicator of optic nerve function appears to be an excellent element to monitoring disease progression.

#### **Methods:**

With analysis of several clinical cases we try to assess the risk of transformation of the evolutionary mode to enable a finer monitoring of this disease

#### **Results:**

Visual fields analysis shows that its alterations are often a sign of fat or muscular intra orbital change . Sometimes alterations are linked with superficial corneal involvement (drought, superficial punctate keratitis, corneal abscess). Association with another disease of optic nerve (glaucoma), is possible, though quite rare, while many patients with ocular hypertension are frequent . After initiation of corticosteroid therapy bolus, it is surprising to see a high recovery rate.

#### **Conclusions:**

Graves ophthalmopathy neuropathy remains a dreaded complication. Corticosteroid therapy in bolus is, according to us, the treatment of choice and allows us to get frequent ameliorations. Central visual field is a method for reliable monitoring

**References:** None.

**Keywords:** Graves (Systemic Disease), Optic Neuropathy, Perimetry, Visual Fields

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 205

### Veps To Lateralized Stimuli To Measure The Interhemispheric Transfer Time (IHTT)

Ilie P. Cretu<sup>1,2</sup>, Solange C. Milazzo<sup>1,4</sup>, Pierre D. Betermiez<sup>1</sup>, Michel R. Petitjean<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, University Hospital of Amiens, Amiens, France, <sup>2</sup>Department of Ophthalmology, Hospital of Abbeville, Abbeville, France, <sup>3</sup>Service de Physiologie - Explorations Fonctionnelles, Hôpital Ambroise Paré, Boulogne-Billancourt, France, <sup>4</sup>University of Picardy Jules Vernes Amiens, France

#### Introduction:

To evaluate in a population of normal subjects the VEPs to lateralized stimuli to measure the interhemispheric transfer time (IHTT), and methodological variability factors.

#### Methods:

We recorded in 36 young right-handed women evoked responses of visual occipital regions right (O2) and left (O1) in response to visual stimulation achieved through a screen with checkerboards alternants either in open fields, or half-fields when the line of sight coincides with a central focus. The test is performed in monocular eye test is chosen by lot with balancing choices, not to have any effect of precession. The atmosphere is scotopic total. The reversal of checkerboard is carried out at a frequency of 1.7Hz. Depending on the distance, the size of the checkerboard is 1 °. Were recorded average of 100 traces for each lead, during two successive series.

#### Results:

The results are presented according to three factors analyzed: the eye, the conditions (reproducibility and effect number), the observer. For stimulation in the full field, there is no effect or observer eye for the latencies of waves N75 and P100, and no effects eye or averaging effect for the latency of the N135 wave. For the half-full stimulation, there is no eye effect, or observer reproducibility, but there is a half-full field effect in the left eye. For TTIH, there is no significant difference between half-full fields, or effect or observer reproducibility, but there is an effect eye. Linear regression was highly significant between the P100 and TTIH.

#### Conclusions:

The VEPs to lateralized stimuli seem a good method to calculate the IHTT, with good reproducibility and a minimal observer-effect. Further work on patient disease would be interesting to screen for a prolonged IHTT in inflammatory demyelinating CNS.

**References:** None.

**Keywords:** Visual-Evoked Potentials (Veps), Half-Full Stimulation (To Lateralized Stimuli), Interhemispheric Transfer Time (IHTT), P100, Posterior Afferent Visual Pathway

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

### The Heidenhain Variant of Creutzfeldt-Jakob Disease

Kristen E Dunbar<sup>1</sup>, Geetha K Athappilly<sup>1,2</sup>, Jennifer Renz<sup>1</sup>, Juan E Small<sup>3</sup>, Carl A Geyer<sup>3</sup>, Doreen T Ho<sup>4</sup>, Thomas R Hedges<sup>1</sup>

<sup>1</sup>Tufts Medical Center/New England Eye Center/Ophthalmology, Boston, MA, USA, <sup>2</sup>Lahey Medical Center/Ophthalmology, Burlington, MA, USA, <sup>3</sup>Lahey Medical Center/Radiology, Burlington, MA, USA, <sup>4</sup>Lahey Medical Center/Neurology Burlington, MA, USA

#### Introduction:

Creutzfeldt-Jakob disease (CJD) presents with ataxia, dementia, and myoclonus prior to ophthalmologic symptoms. The Heidenhain variant is different. Below are two cases presenting with initial visual symptoms emphasizing the critical role of ophthalmology.

#### Methods:

A 66-year-old male presented to an optometrist with photophobia. Examination only revealed a right homonymous hemianopsia. Initial work up was for a cerebrovascular accident based on an initial MRI read as a "left parieto-occipital infarct." With progressive cognitive decline, he was referred for further work up. Ophthalmology consultation revealed only a right homonymous hemianopia as before. Ophthalmology's clinical suspicion for CJD, coincided with neurology's findings of rapidly progressive dementia. Repeat MRI revealed faint cortical changes on diffusion weighted imaging in the left frontal temporal and occipital cortex. EEG changes and cerebrospinal fluid (CSF) positive for tau and 14-3-3 protein confirmed the clinical suspicion of the Heidenhain variant of CJD. 53-year-old male presented with "difficulty focusing" prompting referral for cataract surgery. Exam revealed mild central vision loss, color desaturation and perimetry showing a right homonymous field defect. Initial MRI was read as normal. Repeat MRI brain and discussion with the neuroradiologists revealed cortically-based diffusion abnormalities in the left hippocampus, caudate heads, occipital and temporal lobes. Ophthalmic and imaging suspicion for CJD prompted admission, although overt neurologic findings were lacking. EEG changes and CSF positive for tau, 14-3-3 protein, and RT-QuIC confirmed clinical suspicion. Within a month, he passed with rapid decline in neurologic and cognitive status.

#### Results:

Heidenhain variant CJD

#### Conclusions:

Heidenhain variant of CJD is a diagnostic challenge due to the variable visual symptoms, normal structural eye exam, subtle MRI findings and the lack of initial, overt, neurologic abnormalities. Due to the rapidly progressive fatal course, early clinical suspicion by ophthalmology is critical in diagnosis, especially when future treatment is present.

**References:** None.

**Keywords:** Prion Disease, Creutzfeldt-Jakob Disease, Visual Field Defect

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 207

### Six-Year Follow-Up Of The Progression Of Cortical Vision Loss In A Patient With HIV-Related Progressive Multifocal Leukoencephalopathy

Bahareh Hassanzadeh<sup>1</sup>, Norman J. Schatz

*Bascom and Palmer Eye Institute, Ophthalmology Department University of Miami, Miami, FL, USA*

#### Introduction:

A 43- year- old right-handed African American man with 20-year history of HIV presented to with blurred vision in both eyes. He also had right hemiparesis and right arm flailing. Neuro-exam revealed right hemiparesis, hemiataxia and hemibalismus, alexia without agraphia and dysnomia. Ophthalmic exam showed BCVA 20/25 OD, 20/40 OS. EOM was intact. Slit lamp and fundoscopic exam was normal. Humphrey visual field 30-2 demonstrated a left homonymous para-central scotoma.

#### Methods:

His symptoms started 6 years ago as diplopia and progressive right-sided weakness. About 4 months later, he developed headache and blurred vision. MRI/MRA head revealed "hypointense T1 and hyperintense T2 lesions within both occipital lobes of the brain with mild peripheral enhancement. There was a non-enhancing T2 signal in both middle cerebellar peduncles extending into the left medial cerebellum and no diffusion restriction". Blood test showed CD3/CD8 36, HIV1 RNA by PCR 244580 copies/ml, and it was negative for coagulopathy panel. Lumbar puncture was negative for DNA direct probe for JC and BK viruses. Highly active antiretroviral therapy medications were optimized and he was discharged home on antiplatelet.

#### Results:

One year later, he developed kaleidoscopes of colors in front of his eyes. It followed by an episode of grandmal seizure. EEG was normal. Repeat MRI head w/ wo/ gadolinium showed worsening of the confluent lesions. He refused lumbar puncture but agreed with a brain biopsy. After right occipital craniotomy, a 3.5\*3\*1cm tissue was resected. The pathology demonstrated a white matter- centric, focally demyelinating process consistent with PML and immune reconstitution inflammatory syndrome (IRIS) concomitant with an opportunistic infection by JC virus. Immunostaining for John Cunningham (JC) virus (SV40) highlighted several infected oligodendroglial cells at the edge of the lesions.

#### Conclusions:

The diagnosis of PML should be considered in immunocompromised patients with neuro-ophthalmic findings, particularly those with cortical blindness.

**References:** None.

**Keywords:** Cortical Blindness, Progressive Multifocal Leukoencephalopathy, Hemibalismus, Alexia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 208

### Retrograde Degeneration of Retinal Ganglion Cells in Homonymous Hemianopia

Angela M Herro<sup>1</sup>, Byron L Lam, Sean M Gratton

*Bascom Palmer Eye Institute, Miami, FL, USA*

#### Introduction:

To demonstrate the relationship between topographic reduction in macular ganglion cell complex (GCC) thickness as detected with spectral-domain optical coherence tomography (SD-OCT) to visual field defects caused by ischemic occipital cortical injury.

#### Methods:

Retrospective review of all patients who presented to our eye institute between January 2012 and July 2014 with visual field defects secondary to ischemic cortical injury. The visual field defect pattern and mean deviation were analyzed. Retinal nerve fiber layer (RNFL) and macular GCC were both assessed with SD-OCT. Patients with any ocular pathology that could affect these measurements were excluded. The topographic relationship of visual field defect to reduction in GCC was specifically analyzed.

#### Results:

Nine patients met the inclusion criteria. The average age was 54 (range 16-72 years), 88% were men, and 67% had right hemianopsias. The laterality of the visual field defect was used to assign an affected and unaffected side of analysis for RNFL and GCC layer thickness. A right hemianopsia meant that the nasal fibers of the right eye and temporal fibers of the left eye were assigned as "affected side," and the temporal fibers of the right eye and nasal fibers of the left eye were assigned "unaffected." There was no statistically significant difference between affected and unaffected segments of RNFL. However, there was significant difference between the GCC layer reduction in affected and unaffected sides ( $p = 0.03$ )

#### Conclusions:

There may be retrograde trans-synaptic retinal ganglion cell loss in patients with homonymous hemianopsias from cortical visual impairment. This relationship is reflected in the general reduction in RNFL and more specifically in the GCC as the pattern of reduction maintains the topographic relationship of the visual field defect.

#### References:

1. Jindahra, P., Hedges, T. R., Mendoza-Santiesteban, C. E., & Plant, G. T. (2010). Optical coherence tomography of the retina: applications in neurology. *Curr Opin Neurol*, 23(1), 16-23. doi: 10.1097/WCO.0b013e328334e99b
2. Jindahra, P., Petrie, A., & Plant, G. T. (2012). The time course of retrograde trans-synaptic degeneration following occipital lobe damage in humans. *Brain*, 135(Pt 2), 534-541. doi: 10.1093/brain/awr324
3. Park, H. Y., Park, Y. G., Cho, A. H., & Park, C. K. (2013). Transneuronal retrograde degeneration of the retinal ganglion cells in patients with cerebral infarction. *Ophthalmology*, 120(6), 1292-1299. doi: 10.1016/j.ophtha.2012.11.021
4. Yamashita, T., Miki, A., Iguchi, Y., Kimura, K., Maeda, F., & Kiryu, J. (2012). Reduced retinal ganglion cell complex thickness in patients with posterior cerebral artery infarction detected using spectral-domain optical coherence tomography. *Jpn J Ophthalmol*, 56(5), 502-510. doi: 10.1007/s10384-012-0146-3
5. Vanburen, J. M. (1963). TRANS-SYNAPTIC RETROGRADE DEGENERATION IN THE VISUAL SYSTEM OF PRIMATES. *J Neurol Neurosurg Psychiatry*, 26, 402-409.

**Keywords:** Homonymous Hemianopsia, Ganglion Cell Complex, Retrograde Transsynaptic Degeneration

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 209

### Lesions of the Optic Tract: A Review of 35 Cases

Kristopher M Kowal<sup>1</sup>, Jonathan D Trobe

*University of Michigan, Ann Arbor, MI, USA*

#### **Introduction:**

Previous case series reports, (1,2,3,) the largest involving 21 cases, have documented that optic tract lesions are usually tumors. The imaging detail has been sparse. We have encountered 35 cases that demonstrate a much broader range of causes with diverse imaging features.

#### **Methods:**

Retrospective review of case records from a single institution between 2000 and 2014. Inclusion criteria were 1) homonymous hemianopia; 2) lesion demonstrated on MRI in the region of the optic tract; 3) if the imaged lesion extended into the retrogeniculate region, a relative afferent pupil defect had to be present in the eye with temporal field loss.

#### **Results:**

Causes included brain hemorrhage--6, anterior choroidal infarction--5, head trauma--3, aneurysmal coiling--3, temporal lobectomy--2, sarcoidosis--2, craniopharyngioma--2, glioblastoma--2, herpes zoster, lymphoma, aneurysm, demyelination, astrocytoma, deep venous thrombosis, radiotherapy, arteriovenous malformation, cavernoma, congenital infarction--1 each. In some cases, the imaging abnormalities were subtle and the localization of the lesion was therefore challenging.

#### **Conclusions:**

Our series enlarges the number of known causes for optic tract lesions and demonstrates a broad spectrum of imaging abnormalities, some quite subtle.

#### **References:**

1. Savino PJ, Paris M, Schatz NJ, et al. Optic Tract Syndrome: A review of 21 patients. Arch Ophthalmol 1978;96:656-663
2. Newman SA and Miller NR. Optic Tract Syndrome. Arch Ophthalmol 1983;101:1241-12
3. Bender MB, Bodis-Wollner I. Visual dysfunction in optic tract lesions. Ann Neurol 1978;3:187-193

**Keywords:** Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 210

### Hydrocephalus and More!

Tarek A Shazly<sup>1</sup>, Islam Zaydan<sup>1,2</sup>, Ryan Orie<sup>2</sup>, Kathryn McFadden<sup>3</sup>,

<sup>1</sup>University of Pittsburgh, Department of Ophthalmology, Pittsburgh, PA, USA, <sup>2</sup>University of Pittsburgh, Department of Neurology, Pittsburgh, PA, USA, <sup>3</sup>University of Pittsburgh, Department of Pathology, Pittsburgh, PA, USA

#### Introduction:

The differential diagnosis of multifocal central nervous system lesions is quite broad. We present a case of progressive neurological deficits, visual loss and multiple intracranial lesions due to an unexpected etiology.

#### Methods:

Case report and review of the literature.

#### Results:

A 67-year-old female presented with headaches, altered mental status, leg weakness and dysarthria. Exam revealed patient confusion and mild bilateral leg weakness. Brain MRI showed a confluent abnormal flair signal involving the left periventricular and corpus callosum with hydrocephalus. CSF protein was 130, with 4 WBCs, 1 neutrophil, and 78% lymphocytes. Viral PCRs and cytology were negative. Right frontal ventriculostomy was placed with improvement in mental status followed 5 days later by ventriculoperitoneal shunt and left parietal brain biopsy. Biopsy revealed demyelination with relative preservation of axons. Intravenous dexamethasone treatment caused improvement over 3 weeks. One year later, she presented with forgetfulness, headache, and ataxia. Brain MRI showed abnormal flair and enhancement around the left occipital horn. CSF protein was 156, with normal IgG index and no oligoclonal bands. Repeat biopsy revealed no atypical B cells with active demyelination and axonal sparing. ANA, SSA-B, Lyme, Vitamin B<sub>12</sub>, RPR, and toxoplasma were normal. A C-spine and T-spine MRI looking for evidence of similar lesions was normal. Five months later, she presented with acute onset of painless bilateral vision loss. She initially had a dense bitemporal hemianopsia. Two days later, she had complete loss of vision in both eyes except for small right nasal island. Brain MRI showed enhancement of the optic chiasm, subependymal frontal horns, and floor of third ventricle. Chest CT was normal. Aquaporin-4 antibodies were positive in serum and CSF.

#### Conclusions:

Neuromyelitis Optica has a wide spectrum of neurological presentations. It should be considered early on the differential of multifocal CNS lesions especially with chiasmal involvement.

#### References:

1. Bloch O, Auguste KI, Manley GT, Verkman AS. Accelerated progression of kaolin-induced hydrocephalus in aquaporin-4-deficient mice. *J Cereb Blood Flow Metab.* 2006
2. Shen XQ, Miyajima M, Ogino I, Arai H. Expression of the water-channel protein aquaporin 4 in the H-Tx rat: possible compensatory role in spontaneously arrested hydrocephalus. *J Neurosurg.* 2006; 105(6 suppl):459-464 Mao X, Enno TL, Del Bigio MR. Aquaporin 4 changes in rat brain with severe hydrocephalus. *Eur J Neurosci.* 2006; 23:2929-2936
3. Feng X, Papopoulus M, Liu J, Lihua L. Sporadic Hydrocephalus in Aq4 null Mice. *J. Neurosci Res.* 2009 April, 87 (5): 1150-1155.
4. Lennon VAI, Wingerchuk DM, Kryzer TJ, Pittock SJ, Lucchinetti CF, Fujihara K, Nakashima I, Weinshenker BG. A serum autoantibody marker of neuromyelitis optica: distinction from multiple sclerosis. *Lancet.* 2004 Dec 11-17;364(9451):2106-12.
5. Khanna Si, Sharma A, Huecker J, Gordon M, Naismith RT, Van Stavern GP. Magnetic resonance imaging of optic neuritis in patients with neuromyelitis optica versus multiple sclerosis. *J Neuroophthalmol.* 2012 Sep;32(3):216-20. doi:
6. Asavapanumas Nithi, Julien Ratelade, Marios C Papadopoulos, Jeffrey L Bennett, Marc H Levin and Alan S Verkman. Experimental mouse model of optic neuritis with inflammatory demyelination produced by passive transfer of neuromyelitis optica-immunoglobulin G. *Journal of Neuroinflammation* 2014, 11:16

**Keywords:** Optic Neuritis, Neuromyelitis Optica, Hydrocephalus, Demyelination

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Ipratropium-Induced Mydriasis: A Possible Exception to the Current Anisocoria Diagnostic Pathway

Erica L Archer<sup>1</sup>, Robert Shin

University of Maryland Medical Center Department of Ophthalmology, Baltimore, MD, USA

### Introduction:

Pupillary abnormalities often evoke significant clinical concern, especially in ICU settings. We have observed cases of transient anisocoria among critically-ill patients who received nebulized bronchodilators via face mask. Though it is suspected that nebulized ipratropium could induce mydriasis if accidentally administered topically to the eye (23 case reports between 1986 and 2012 have described dilated pupils among patients using ipratropium inhalers and nebulizers)<sup>1-23</sup>, only 3 studies have attempted to reproduce this anecdotal association.<sup>6, 24, 25</sup> The use of topical pilocarpine 1% is considered a definitive diagnostic test to distinguish between pharmacological mydriasis and a third nerve palsy. We have, however, observed cases of presumed ipratropium-induced mydriasis at our institution in which the dilated pupil constricted when challenged with pilocarpine 1%. We suspect that ipratropium-induced mydriasis may be reversed by topical pilocarpine 1%, contradicting the widely-accepted anisocoria diagnostic algorithm.

### Methods:

An interventional case study was performed on a willing volunteer. After an ophthalmological examination revealed no pupillary abnormalities, nebulized ipratropium bromide 0.02% and nebulized ipratropium/albuterol (0.5 mg / 3 mg per 3 mL) solutions were instilled in mist and drop form OD. Serial ophthalmological examinations were performed at 30 minutes, 60 minutes, 6 hours, 12 hours, and 18 hours. Pilocarpine 1% was administered OU at varying times from initial recognition of mydriasis.

### Results:

After instillation of ipratropium, mydriasis gradually developed OD over several hours, lasting greater than 18 hours. After administration of pilocarpine 1% OU, pupillary constriction OU was noted within one hour.

### Conclusions:

Ipratropium bromide can produce mydriasis when administered topically to the eye. This effect appears to be dose-dependent and relatively long-lasting (approximately 18-24 hours). Furthermore, ipratropium-induced mydriasis may be reversed by pilocarpine 1%, challenging the currently accepted anisocoria diagnostic pathway. Pharmacological testing should be interpreted with caution in this clinical context.

### References:

- (1) Brodie T, Adalat S. Unilateral fixed dilated pupil in a well child. Arch Dis Child, 91 (12), 961, 2006; (2) Quinonez ZA, Ravula NR. Anisocoria in a 10-month old girl in the immediate preoperative setting: can you proceed with surgery? J Biomedical Research, 25 (3), 224-226, 2011; (3) Bond DW, Vyas H, Venning HE. Mydriasis due to self-administered inhaled ipratropium bromide. Eur J Pediatr, 161, 178, 2002; (4) Weir REP, Whitehead DEJ, Zaidi FH, Greaves BBG. Pupil blown by a puffer. The Lancet, 363,1853, 2004; (5) Santana-Cabrera L, Fernandez-Tagarro EJ, el Amo-Nolasco B, Jaen-Sanchez N, Caceres-Agra JJ. Unilateral mydriasis secondary to ipratropium bromide in a critically ill patient. J Emerg Trauma Shock, 5 (2), 199-200, 2012; (6) Sharma NS, Ooi JL, Papalkar D, Sharma S, Francis IC. Pharmacological mydriasis secondary to ipratropium bromide: A cause of unilateral dilated pupil. J Clin Neuroscience, 15, 320-321, 2008; (7) Openshaw, H. Unilateral mydriasis from ipratropium in transplant patients. Neurology, 67, 914, 2006; (8) Camkurt MA, Ay D, Akkucuk H. Pharmacologic unilateral mydriasis due to nebulized ipratropium bromide. Am J Emergency Medicine, 29, 576.e5-576.e6, 2011; (9) Krovvidi HP, Thillaivasan A. A benign cause for a unilateral dilated pupil in a critical care patient. European Journal of Anaesthesiology, 25, 689-700, 2008; (10) Udy A. A 10-year-old child with status asthmaticus, hypernatremia and a unilateral dilated pupil. Pediatric Anesthesia, 15, 1120-1123, 2005; (11) Wehbe E, Antoun SA, Moussa JK, Nassif II. Transient anisocoria caused by aerosolized ipratropium bromide exposure from ill-fitting face mask. J Neuro-Ophthalmol, 28 (3), 236-237, 2008; (12) Iosson N. Nebulizer-associated anisocoria. N Engl J, 354, 9, 2006; (13) Goldstein JB, Biousse V, Newman NJ. Unilateral pharmacologic mydriasis in a patient with respiratory compromise. Arch Ophthalmol, 115, 806, 1997; (14) Bisqueria RA, Botz GH, Nates JL. Ipratropium-bromide-induced acute anisocoria in the intensive care setting due to ill-fitting face masks. Respiratory Care, 50(12), 1662-1664, 2005; (15) Lust K, Livingstone I. Nebulizer-Induced Anisocoria. Annals of Int Medicine, 124 (4), 327, 1998; (16) Woelfe J, Zielen S, Lentze M. Unilateral fixed dilated pupil in an infant after inhalation of nebulized ipratropium bromide. The Journal of Pediatrics, 136 (3), 423-424, 2000; (17) Cabana MD, Johnson H, Lee CKK, Helfaer, M. Transient Anisocoria Secondary to Nebulized Ipratropium Bromide. Clin Pediatr, 37, 445-448, 1998; (18) Ryan CA. Ipratropium Bromide Induced Unilateral Mydriasis. Irish Medical Journal, 90 (2), 76, 1997; (19) Ryan, CA. Styrofoam Cup, Ipratropium Bromide, and Anisocoria. Clin Pediatr, 38, 318, 1999; (20) Tuohy PG. Ipratropium and the eye. N Z Med J, 102 (872), 386, 1989; (21) Helprin GA, Clarke GM. Unilateral fixed dilated pupil associated with nebulized ipratropium bromide. Lancet, 2 (8521-22), 1469, 1986; (22) Eustace N, Gardiner C, Eustace P, Marsh B. Nebulised Ipratropium Causing a Unilateral Fixed Dilated Pupil in the Critically Ill Patient: A Report of Two Cases. Crit Care and Resuscit, 6, 268-270, 2004; (23) Geraerts SD, Plotz FB, vGoor C, Duval ELIM, vVught H. Unilateral fixed dilated pupil in a ventilated child with asthma. European Journal of Emergency Medicine, 7, 247 – 248, 2000; (24) Samaniego F, Newman LS. Migratory anisocoria – a novel clinical entity. Am Rev Respir Dis, 134 (4), 844, 1986; (25) Farrow PR, Fancourt GJ. Does Ipratropium bromide by Nebulizer and Face-mask have Local Ocular Effects? Human Toxicol, 5, 53-54, 1986.

**Keywords:** Pupils

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 212

### Automated Pupillograph as a Screening Tool in Ophthalmology Clinic

Ashwin Mohan, Chaithra D Aroor<sup>1</sup>, Rohit Shetty, Luci Kaweri, Narendra KP, Dhanraj Rao, Ramagopal B, Manan Jariwala

*Narayana Nethralaya, Bangalore, India*

#### Introduction:

To evaluate the efficacy of an automated pupillograph as a screening tool in ophthalmology clinic.

#### Methods:

110 subjects in the study were enrolled in the study, 80 normals (72.7%) and 30 (27.3%) patients with Glaucoma. Pupillary reactions were measured using RAPDx Expanded Pupil Diagnostics (Konan Medical USA, Inc., Irvine, CA) and were compared with Neutral Density Filter (NDF) (Gulden Ophthalmics). In addition, 30 Glaucoma patients also underwent analysis of Macular Ganglion cell (mGCC) thickness using a Spectral domain -Optical Coherence Tomography (Optovue RTVue XR AVANTI).

#### Results:

The pupillary reactions in normal 80 patients (57% males, 43 % females) assessed by NDF was found to be less than 0.3 log units. On RAPDx the same was found to be 0.28 log units. Statistically significant correlation ( $p < 0.001$ ) was seen between the two. Mean amplitude of pupillary reactions on RAPDx (0.14 log units) in 30 glaucoma subjects, correlated significantly with the mGCC thickness ( $P < 0.05\%$ ). However mean latency of the pupillary reactions (0.12 msec) showed a weak correlation with mGCC.

#### Conclusions:

This pilot study concludes that RAPDx is comparable to NDF in measuring RAPD and can be used interchangeably. In glaucomatous patients the log-scaled RAPD amplitudes correlated moderately with the mGCC thickness, but the log-scaled RAPD latencies showed a weaker correlation. RAPDx may be used as a screening tool in Ophthalmology clinic.

**References:** None.

**Keywords:** Pupillograph, Macular Ganglion Cell Complex, Pupillary Reactions, Rapdx, Neutral Density Filter

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 213

### The Yield of Diagnostic Imaging in Patients with Isolated Horner's Syndrome

Johanna D. Beebe<sup>1</sup>, Randy H. Kardon<sup>1,2</sup>, Matthew J. Thurtell<sup>1,2</sup>

<sup>1</sup>University of Iowa Department of Ophthalmology and Visual Science, Iowa City, IA, USA, <sup>2</sup> Iowa City VA Medical Center, Iowa City, IA, USA

#### Introduction:

We conducted a retrospective chart review to determine the yield of diagnostic neuroimaging for patients with isolated Horner's syndrome.

#### Methods:

We conducted a retrospective chart review of 123 patients seen by our service between 2000 and 2014, who were coded as having Horner's syndrome with clinical and/or pharmacologic confirmation of the diagnosis. We excluded infants and patients with congenital Horner's syndrome.

#### Results:

Patient age ranged from 6 to 87 years old, with an average age of 50 years old. There were 123 cases of clinically- or pharmacologically-confirmed Horner's syndrome, with 63 male and 60 female patients. On initial presentation, 19 patients had a known cause or constellation of findings that indicated the etiology of their Horner's syndrome. Imaging of the oculosympathetic pathway was pursued in 94 patients (76.4%). Prior to imaging, 5 of the 94 patients (5%) had a known etiology for their Horner's syndrome. There were 89 patients with an isolated Horner's syndrome who underwent neuroimaging in search of a causative lesion. Of these, 17 (19%) were found to have a lesion on neuroimaging that was causative for the Horner's syndrome. The most common finding was carotid artery dissection in 7 patients (8%), with all but one having an acute-onset painful Horner's syndrome. None of the imaged patients were found to have a primary malignancy, but one patient with known metastatic disease was found to have a new metastatic lung lesion. Seven of the patients (7%) had incidental findings on imaging that were unrelated to the oculosympathetic defect.

#### Conclusions:

To our knowledge, this is the largest series evaluating the diagnostic yield of neuroimaging for isolated Horner's syndrome. Imaging identified a causative lesion in 19% of patients with an isolated Horner's syndrome. About 7% of patients had an incidental finding on neuroimaging that was unrelated to the oculosympathetic defect.

**References:** None.

**Keywords:** Pupils, Neuroimaging

**Financial Disclosures:** Dr. Randy Kardon: Funding (grants) from NEI R009040554 R01 EY018853 Funding (grants) Department of Defense TATRC Funding (grants) VA Rehabilitation Research and Development Novartis steering committee OCTiMS

**Grant Support:** Unrestricted grant from Research to Prevent Blindness, New York City, NY

## Evaluation of Pupil Response as Ocular Marker for Pre-Clinical Alzheimer's Disease

Ling Bei<sup>1</sup>, Ying-bo Shui<sup>2</sup>, Fang Bai<sup>2</sup>, Suzanne Nelson<sup>3</sup>, David Beebe<sup>2</sup>, Morris John<sup>4</sup>, Gregory Van Stavern<sup>2</sup>

<sup>1</sup>University of California San Diego/Department of Ophthalmology, San Diego, CA, USA, <sup>2</sup>Washington University in Saint Louis School of Medicine/Department of Ophthalmology and Visual Sciences, Saint Louis, MO, USA, <sup>3</sup>Washington University in Saint Louis School of Medicine/Department of Biostatistics, Saint Louis, MO, USA, <sup>4</sup>Washington University in Saint Louis School of Medicine/Department of Neurology Saint Louis, MO, USA

### Introduction:

Alzheimer's disease (AD) is a neurodegenerative disease characterized by cognitive deficits and visual dysfunction. There is increasing interest in the use of ocular markers as a surrogate for disease activity and progression in AD<sup>1</sup>. Several studies have shown that the pupillary light response (PLR) can differentiate AD patients from healthy controls<sup>2,3</sup>. However, the majority of these studies have assessed the PLR in subjects with established Alzheimer's disease, and it remains unclear whether the PLR is affected in subjects with pre-clinical AD.

### Methods:

We recruited participants from our institution's Alzheimer's Disease Research Center. All participants completed positive emission tomography-Pittsburgh compound B imaging, cerebrospinal fluid analysis and at least 1 neuropsychiatric assessment after their baseline assessment. All participants were assigned a clinical dementia rating (CDR) and underwent a complete neuro-ophthalmic examination. Participants were divided into a dementia biomarker + and – group based on preclinical risk for or presence of dementia. Pupillometry measurements were performed by using the NeuroOptics PLR-200 Pupillometer.

### Results:

A total of 39 participants were recruited with 12 dementia biomarker + and 27 dementia biomarker- individuals. A variety of PLR parameters were assessed. Comparisons between groups were analyzed using Generalized Estimating Equations. The Minimum Pupil Size was significantly smaller in the dementia biomarker + group ( $p = 0.012$ ). The Maximum Pupil Size was also smaller in the biomarker dementia + group, and approached significance ( $p = 0.06$ ). None of the other pupillary parameters showed a significant difference between groups.

### Conclusions:

We found a significant difference in the PLR between dementia biomarker +, pre-clinical AD subjects versus controls. We are continuing to recruit subjects to determine whether our preliminary results are validated. The PLR is a potential candidate for non-invasive screening of pre-clinical AD, and might also be used as a surrogate marker for disease progression in future clinical trials.

**References:** None.

**Keywords:** Pupils, Neuro-Ophth & Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



**A Case Of Horner's Syndrome After Clipping Of The Internal Carotid-Posterior Cerebral Artery Aneurysm Associated With Subarachnoid Hemorrhage**

Tamura Koichiro<sup>1,2</sup>, Chuman Hideki<sup>1</sup>, Nobuhisa Naoi<sup>1</sup>

<sup>1</sup>*ophthalmology, miyazaki, Japan*, <sup>2</sup>*ophthalmology, oita, Japan*

**Introduction:**

Internal carotid-posterior cerebral artery (IC-PC) aneurysm is a cause of parasympathetic nerve paresis of the pupil. We have experienced a case of sympathetic nerve paresis of the pupil after clipping of IC-PC aneurysm associated with subarachnoid hemorrhage. We will report the case and discuss the mechanism.

**Methods:**

A Case report and review of the literature.

**Results:**

A 59-year-old woman had a subarachnoid hemorrhage caused by a rupture of left IC-PC aneurysm in September 2007. She was treated by a clipping procedure with the complication of the rupture of the aneurysm. After the intervention, she noted to have left blepharoptosis. She was referred to us for the treatment of left blepharoptosis in April 2014. Her visual acuity was 20/20 in both eyes. Pupils measured 5mm OD and 3.5mm OS in dark, 3mm OD and 2.5mm OS in light, constrict briskly to light OU. There was no afferent pupillary defect. Her ocular motility was 100% in all directions without nystagmus. She was orthotropic in all directions. Confrontational visual field was normal. Her palpebral fissure width was 9mm OD and 7mm OS. Corneal sensation was normal. Systemic neurological examination revealed a motor aphasia, right hemiplegia, and sensory deficit on the whole right side body including face. After the instillation of 10% cocaine, pupils measured 7mm OD and 4mm OS. After the instillation of 1% hydroxyamphetamine, pupils measured 7mm OD and 5mm OS. After the instillation of 0.1% epinephrine, pupils measured 5mm OD and 7mm OS. Following these results, we diagnosed her to have a postganglionic Horner's syndrome on the left. A CT angiography revealed a left internal carotid artery obstruction.

**Conclusions:**

Similar case has not been found as far as we investigated. Sympathetic nerve paresis of the pupil could occur after clipping of the IC-PC aneurysm associated with subarachnoid hemorrhage.

**References:**

1. Parkinson D, Johnston J, Chaudhuri A. Sympathetic connections of the fifth and sixth cranial nerves. *Anat Rec* ; 191: 221-226.1978
2. Biousse V, TouboulP-J, D'Anglejan-Chatillon J, et.al. Ophthalmologic manifestations of internal carotid artery dissection. *Am J Ophthalmol* ; 126: 565-577.1998
3. Monteiro MLR, Coppeto JR. Horner's syndrome associated with carotid artery arteriosclerosis. *Am J Ophthalmol* ; 105: 93-94.1998
4. Keane JR. Oculosympathetic paresis: Analysis of 100 hospitalized patients. *Arch Neurol* ; 36: 13-16.1979
5. Riley FC, Moter NJ. Oculosympathetic paresis associated with cluster headaches. *Am J Ophthalmol* ; 72: 763-768.1971
6. Thompson HS, Mensher JH. Hydroxyamphetamine test in Horner's syndrome. *Am J Ophthalmol* ; 79: 523-526.1975
7. Trobe JD, Glaser JS, Post JD. Meningiomas and aneurysms of the cavernous sinus. *Arch Ophthalmol* ; 96: 457-467.1978
8. Huddins R, Aneurysms of the posterior inferior cerebellar artery: a clinical and anatomical analysis. *J Neurosurg* ; 58: 381-387.1983

**Keywords:** Horner Syndrome, IC-PC Aneurysm, Subarachnoid Hemorrhage, Anisocoria, Internal Carotid Obstruction

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Test–Retest Reliability of Hemifield, Central-Field and Full-Field Chromatic Pupillometry for Assessing the Function of Melanopsin-Containing Retinal Ganglion Cells**

Shaobo Lei<sup>1</sup>, Herbert C. Goltz<sup>1,3</sup>, Manokaraanathan Chandrakumar<sup>1</sup>, Agnes M.F Wong<sup>1,2,3</sup>

<sup>1</sup>*Program in Neurosciences and Mental Health, University of Toronto, Toronto, ON, Canada,* <sup>2</sup>*Department of Ophthalmology and Vision Sciences, The Hospital for Sick Children, Toronto, ON, Canada,* <sup>3</sup>*Department of Ophthalmology and Vision Sciences, University of Toronto, Toronto, ON, Canada*

**Introduction:**

A sustained pupil constriction can be observed after the offset of a bright blue light stimulus. This post-illumination pupil response (PIPR) is produced by the intrinsically photosensitive melanopsin-containing retinal ganglion cells (ipRGCs), and can be measured by chromatic pupillometry. We previously described a clinically friendly chromatic pupillometry protocol that can induce PIPR with a brief blue light stimulus. The present study is to evaluate the test-retest reliability of current methods of inducing PIPR under hemifield, central-field and full-field stimulation conditions.

**Methods:**

Pupil response was recorded with an eye tracker in 10 visually normal participants. Light stimuli were presented using a Ganzfeld screen with a custom-built device that allows specific regions of the retina to be stimulated. Blue light stimulation at 400 cd/m<sup>2</sup> intensity was presented for 400 ms to the lower and upper halves of the central 30° fields (hemifields), both halves of the central 30° field (central-field) and full-field to induce PIPR. Red light full-field stimulation was also presented with the same intensity and duration as a control condition. Test-retest reliability of the PIPR measures were assessed by calculating the intra-class correlation coefficient (ICC) of 6 repetitions for lower and upper hemifield stimulation, and 3 repetitions for central-field and full-field stimulation.

**Results:**

Hemifield, central-field, and full-field blue light stimulation induced increasingly greater PIPR in ascending order, while full-field red light stimulation induced no PIPR. Mean lower and upper hemifield PIPR were highly symmetric. Mean ICC of blue light PIPR was 0.87 for lower hemifield, 0.88 for upper hemifield, 0.95 for central-field, and 0.94 for full-field stimulation.

**Conclusions:**

We validated a new and repeatable method to measure PIPR induced by hemifield, central-field and full-field light stimulation. Good PIPR measurement reliability was obtained under all conditions. This practical and reliable protocol will facilitate the clinical application of PIPR testing in different disease populations.

**References:** None.

**Keywords:** Pupil Light Reflex, Pupillometry, Test-Retest Repeatability, Melanopsin, Retinal Ganglion Cells

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Supported by the Canada Foundation for Innovation, John and Melinda Thompson Endowment Fund for Vision Neuroscience and the Department of Ophthalmology and Vision Sciences at The Hospital for Sick Children.

## Poster 217

### Unique Presentation of Anti-GQ1b Antibody Syndrome

Tin Yan A. Liu<sup>1</sup>, Vivek R. Patel<sup>2</sup>

<sup>1</sup>Wilmer Eye Institute, Baltimore, MD, USA, <sup>2</sup>University of Southern California Eye Institute, Los Angeles, CA, USA

#### Introduction:

Anti-GQ1b antibody syndrome can present along a spectrum: from acute ophthalmoparesis to Miller Fisher Syndrome to Bickerstaff's brainstem encephalitis to Guillain Barre Syndrome. Here, we report a patient who presented with an interesting combination of neuro-ophthalmological findings.

#### Methods:

Case report.

#### Results:

The patient was an 18 year-old, previously healthy, Caucasian man, who presented with one day of acute onset binocular diplopia. He was in his usual state of health until one week prior to presentation, when he developed upper respiratory tract infection symptoms. On exam, his bilateral pupils were dilated (8mm), didn't constrict to direct light stimulation or accommodation, but constricted readily with 1% pilocarpine drops, thus ruling out direct muscarinic blockade. Right eye was esotropic with a 20% limitation in abduction, consistent with a partial right cranial nerve VI palsy. Left upper eyelid had a 2mm ptosis, with a negative ice test. Left eye was hypotropic with a 10% limitation in upgaze, potentially consistent with a partial left cranial nerve III palsy. Extraocular movements were otherwise full with normal saccadic velocities. His ophthalmic exam was otherwise normal. His neurological exam was otherwise normal, with normal reflexes and no ataxia or dysidiadochokinesia. MRI of the brain and orbits was unremarkable. Acetylcholine receptor binding antibodies and striated muscle antibodies were negative. Single fiber electromyography of the frontalis muscle showed no evidence of myasthenia gravis. Lumbar puncture showed normal basic studies with no albuminocytologic dissociation. CSF infectious work up was negative. His serum GQ1b antibody titer was significantly elevated at 1:3200.

#### Conclusions:

To our knowledge, this is the first reported case in which the patient has the unique combination of bilateral mydriasis with complete internal ophthalmoplegia to both light and near stimuli, in the absence of ataxia and areflexia. It should serve to extend the clinical spectrum of disease associated with anti-GQ1b antibodies.

**References:** None.

**Keywords:** Pupils, Neuro-Ophth & Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Novel Retinal Observations in Genetically Confirmed Kearns Sayre Syndrome

Thomas M Bosley<sup>1</sup>, Darren T Oystreck<sup>2</sup>, Khaled K Abu-Amero<sup>1</sup>, Sawsan R Nowilaty<sup>3</sup>, Emma L. Blakely<sup>4</sup>, Robert Taylor<sup>4</sup>, Igor Kozak<sup>3</sup>

<sup>1</sup>King Saud University/Ophthalmology, Riyadh, Saudi Arabia, <sup>2</sup>University of Stellenbosch/Ophthalmology, Tygerberg, South Africa, <sup>3</sup>King Khaled Eye Specialist Hospital/Ophthalmology, Riyadh, Saudi Arabia, <sup>4</sup>Newcastle University/Wellcome Trust Centre for Mitochondrial Research Newcastle Upon Tyne, United Kingdom

### Introduction:

Kearns Sayre Syndrome (KSS) is a complex mitochondrial syndrome typically caused by a single, large-scale mitochondrial DNA (mtDNA) deletion that presents before the age of 20 years with progressive external ophthalmoplegia, pigmentary retinopathy, and other neurologic and medical problems. We emphasize the retinal changes, including several novel observations, in three clinically and genetically characterized patients with KSS.

### Methods:

Evaluation of three unrelated patients with retinal and neuro-ophthalmologic examinations, medical chart review, and mitochondrial genetic evaluation.

### Results:

All three patients were recognized during early childhood to have progressive external ophthalmoplegia and pigmentary retinopathy bilaterally. All had small stature and other cardiac, endocrinologic, or neurologic signs. Two patients underwent muscle biopsies revealing histopathological changes consistent with mitochondrial myopathy. All had abnormal neuroimaging with hypodense cerebral white matter on CT or bright signal on T2-weighted magnetic resonance images in hemispheric white matter, thalami, and midbrain. All three had modestly reduced visual acuity OU with a pigmentary retinopathy involving predominantly the posterior pole and low amplitude electroretinographic waveforms. One patient had bilateral subretinal fibrosis worse on the left with a full thickness macular hole on the right, neither of which have been reported previously in KSS. All three patients had single, large-scale mtDNA deletions between 5.0-7.6 kb with blood heteroplasmy levels varying between 20% and 58%.

### Conclusions:

Subretinal fibrosis and macular hole are novel clinical observations in KSS. In these three patients, severity of pigmentary retinopathy correlated grossly with retinal electrophysiologic changes but not obviously with ptosis, ocular motility, other clinical and radiologic features, or the severity of the mtDNA rearrangement.

**References:** None.

**Keywords:** Kearns Sayre Syndrome, Mitochondria, Pigmentary Retinopathy, Epiretinal Fibrosis, Progressive External Ophthalmoplegia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** KSA National Program for Science and Technology Grant # 12-MED2621-02

**Autoimmune Retinopathy And Optic Neuropathy In A patient With Anti-GAD And Other Retinal Antibodies**

Sumayya J Almarzouqi<sup>1</sup>, Michael L Morgan<sup>1</sup>, Petros E Carvounis<sup>2</sup>, Andrew G Lee<sup>1-6</sup>

<sup>1</sup>Houston Methodist Hospital/Ophthalmology, Houston, TX, USA, <sup>2</sup>Baylor College of Medicine/Ophthalmology, Houston, TX, USA, <sup>3</sup>Weill Cornell Medical College/ Ophthalmology, Neurology, and Neurosurgery, Houston, TX, USA, <sup>4</sup>UTMB Galveston, TX, USA, <sup>5</sup>Anderson Cancer Center Houston, TX, USA, <sup>6</sup>The University of Iowa Hospitals and Clinics Iowa, IA, USA

**Introduction:**

Autoimmune retinopathy (AIR) is a spectrum of rare acquired retinal diseases, including cancer-associated retinopathy (CAR), melanoma-associated retinopathy (MAR), and presumed non-paraneoplastic autoimmune retinopathy (npAIR). Autoimmune-related retinopathy and optic neuropathy (ARRON) syndrome is characterized by the presence of antibodies against retinal or optic nerve antigens in the absence of cancer. We report a case of acquired autoimmune cone dystrophy and optic neuropathy in a patient without cancer involving an abnormal immunologic reactivity against a 20-kDa optic nerve antigen and 40-kDa and 62-kDa retinal antigens and anti- glutamic acid decarboxylase (GAD) antibodies.

**Methods:**

Single case report

**Results:**

Multifocal electroretinography (MERG) revealed centrally depressed responses, and visually evoked potential (VEP) testing revealed left eye extinguished waveforms at 20/100. Fundus autofluorescence showed central macula hypofluorescence OU and fundus fluorescein angiography showed unusual diffuse patchy hyperfluorescence OU. MRI of the brain and orbits, whole body PET scan, cerebrospinal fluid analysis and testing for Leber hereditary optic neuropathy were unremarkable. Hemoglobin A1C was 8.6%, and serum glucose was 283 mg/dl. Commercial paraneoplastic antibody testing revealed an elevated anti-GAD antibody titer. Experimental auto-antibody testing showed reactivity in the 40-kDa region, consistent with either rhodopsin or the 40-kDa CAR antigen, anti-retinal antibodies in the 62-kDa region and anti-optic nerve antibodies in the region of 20-kDa region.

**Conclusions:**

This case expands the spectrum of autoimmune antibody related disease of the retina and optic nerve. To our knowledge, the presence of anti-GAD antibody as well as anti-retinal and anti-optic nerve antibodies in our patient is unique.

**References:** None.

**Keywords:** Autoimmune Retinopathy, Autoimmune Neuropathy, Anti-GAD Antibody, Anti-Retinal Antibody, Anti-Optic Nerve Antibody

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 220

### GM2-gangliosidosis, AB variant: An Elusive Cause of Neurodegenerative Cherry Red Spots

Michael C Brodsky<sup>1,2,3</sup>, Deborah L Renaud<sup>1,2,4</sup>

<sup>1</sup>Mayo Clinic Department of Neurology, Rochester, MN, USA, <sup>2</sup>Mayo Clinic Department of Pediatrics, Rochester, MN, USA,

<sup>3</sup>Mayo Clinic Department of Ophthalmology, Rochester, MN, USA, <sup>4</sup>Mayo Clinic Department of Medical Genetics Rochester, MN, USA

#### Introduction:

Introduction: GM2-gangliosidosis, AB variant is a rare form of GM2-gangliosidosis due to a deficiency of GM2 activator protein. This autosomal recessive disorder is caused by mutations in GM2A that can only be confirmed by molecular analysis.

#### Methods:

Methods: Retrospective case analysis with cherry red spots confirmed by molecular analysis to be due to GM2 gangliosidosis.

#### Results:

Results: A one-year-old Hmong girl was evaluated for global developmental delay, hypotonia and cherry red spots. The parents were not known to be consanguineous. Her examination was notable for hypotonia with hyperreflexia and excessive startling. The head circumference was normal. An extensive neurometabolic evaluation including hexosaminidase A enzyme assay in white blood cells was negative. Retinal examination at 16 months of age disclosed bilateral cherry red spots with heaped up whitish infiltrate surrounding both foveae but no evidence of optic atrophy or peripheral retinal abnormalities. MR imaging revealed delayed myelination associated with abnormal signal intensity of the bilateral thalami, presenting as T2-hyperintensity of the posterior thalami in the region of the pulvinar nuclei and T2- hypointensity in the anterior thalami. Sequencing of the GM2A gene revealed a homozygous c.160 G>T mutation, predicted to result in a premature protein termination p. Glu54\*.

#### Conclusions:

Conclusions: Deficiency of GM2-activation protein is a rare but important cause of infantile cherry red spots that is not detected by routine neurometabolic screening.

**References:** None.

**Keywords:** Gangliosidosis, Cherry, Red, Spot, Activator

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 221

### Malignant Optic Nerve Glioma Manifesting as Central Retinal Vein Occlusion and Papilledema

Ryan C. Burton<sup>1</sup>, Michael S. Vaphiades<sup>1,2,3</sup>, Jennifer T. Scruggs<sup>1</sup>

<sup>1</sup>University of Alabama at Birmingham, Department of Ophthalmology, Birmingham, AL, USA, <sup>2</sup>University of Alabama at Birmingham, Department of Neurology, Birmingham, AL, USA, <sup>3</sup>University of Alabama at Birmingham, Department of Neurosurgery, Birmingham, AL, USA

#### Introduction:

Malignant optic nerve glioma is a disease of older adults that often progresses to bilateral blindness and death<sup>1</sup>. This is in contrast to optic nerve gliomas of childhood, which are typically benign<sup>2</sup>. It may present under the veil of other diagnoses, including optic neuritis, non-arteritic ischemic optic neuropathy, or retinal vascular occlusion.

#### Methods:

A case report.

#### Results:

A 67-year-old woman presented with decreased Snellen acuity and color vision in the left eye (OS) and pain with eye movement. Fundus exam showed optic disc edema in both eyes and signs of a central retinal vein occlusion OS. Magnetic resonance imaging demonstrated a right thalamic mass extending to the left optic nerve. Intracranial biopsy of this lesion was initially felt to be too dangerous. Vision declined to no light perception (NLP) OS. A left optic nerve biopsy was performed in order to obtain tissue and reach a definitive diagnosis. Unfortunately the biopsy only revealed fibrosis and chronic inflammation. Progression of the radiologic findings, decline in mental status, and development of NLP vision in the right eye prompted a stereotactic thalamic biopsy that revealed glioblastoma multiforme. The patient died three months after her initial presentation.

#### Conclusions:

Malignant optic nerve glioma is a rare condition that suggests a poor visual and overall prognosis, often leading to death within months of the initial presentation. It should be considered as a possible diagnosis in the setting of a central retinal vein occlusion with other intracranial signs.

#### References:

1. Dutton JJ. Gliomas of the anterior visual pathway. *Surv Ophthalmol.* 38, 227-52, 1994.
2. Wabbels B, Demmler A, Seitz J, et al. Unilateral adult malignant optic nerve glioma. *Graefes Arch Clin Exp Ophthalmol.* 242, 741-8, 1994.

**Keywords:** Retina, Neuroimaging, Tumors, Optic neuropathy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This work was supported in part by an unrestricted grant from the Research to Prevent Blindness, Inc. N.Y., N.Y.

## Poster 222

### Comprehensive Postmarketing Review of Visual Defects Reported With Topiramate

Lisa Ford<sup>1</sup>, Jeffrey Goldberg<sup>2</sup>, Fred Selan<sup>3</sup>, Howard Greenberg<sup>1</sup>, Yinqi Shi<sup>3</sup>

<sup>1</sup>Janssen Research & Development, LLC, Titusville, NJ, USA, <sup>2</sup>Shiley Eye Center, University of California, La Jolla, CA, USA,

<sup>3</sup>Janssen Research & Development, LLC, Raritan, NJ, USA

#### Introduction:

Treatment-emergent adverse events (TEAEs) including visual field defects (VFDs) reported with topiramate use, are usually reported as reversible. The objective was to determine possible mechanisms of action (MOA) for VFDs and patterns for development of VFDs with topiramate treatment.

#### Methods:

A comprehensive topiramate database review included: preclinical data – to evaluate topiramate's MOA compared with other antiepileptic drugs (AEDs) associated with VFDs, sponsor's clinical trials database, postmarketing spontaneous reports, and medical literature. All TEAEs suggestive of retinal dysfunction or damage were summarized by system and organ class (SOC) and preferred term (MedDRA version 14.0). A relative risk (RR) analysis was conducted using data from topiramate double-blind, placebo-controlled trials (DBPCTs).

#### Results:

Topiramate-treated patients (N=5679) showed greater frequency of TEAEs suggestive of retinal dysfunction or damage versus placebo-treated (N=1834) (0.3%-0.7% vs. ≤0.1%) patients in DBPCTs for approved indications; similar incidence was noted in open-labels (OLs) and in DBPCTs for investigational indications. In OLs (each investigational indications), incidence was <0.1% (except diabetic peripheral neuropathy [1.4%]). 88% of TEAEs were mild-moderate. Serious events were rare, and most were not treatment limiting and resolved. Most common visual TEAEs were VFD, scotoma and optic atrophy (approved indication trials), and VFD, tunnel vision, retinopathy, retinal hemorrhage and retinal detachment (investigational indication trials). Incidence of TEAEs in DBPCTs (approved and investigational indications) was higher in topiramate-treated (N=9169) versus placebo-treated patients (N=5023) (0.36% vs. 0.24%), RR to placebo-treated patients was 1.51 (95% CI, 0.78-2.91); not statistically significant.

#### Conclusions:

As VFD is not a common TEAE with other AEDs that have a gamma-aminobutyric acid-ergic MOA similar to topiramate; therefore, retinal function is not a class effect. A comprehensive review of topiramate data revealed an increased incidence of visual TEAEs in topiramate-treated versus placebo-treated patients, however, RR was not statistically significant. Topiramate prescribing information includes warnings related to visual TEAEs.

**References:** None.

**Keywords:** Topiramate, Visual Field Defects, Retinal Damage, Retinal Dysfunction, Clinical Trials

**Financial Disclosures:** This study was funded by Janssen Research & Development LLC. Drs. Ford, Selan, Greenberg and Shi are employees of Janssen Research & Development LLC. and hold company stocks. Dr. Goldberg has received compensation for consulting from Janssen Research & Development LLC., Allergan and Theravance.

**Grant Support:** None.



## Sequential Fundus Findings After Platelet Rich Plasma For Facial Rejuvenation

Emely Z Karam<sup>1</sup>, Evlyn Perez<sup>2</sup>, Victor Torres<sup>3</sup>, Ana K Restrepo<sup>4</sup>, Libsen Rodriguez<sup>5</sup>, Herme D Marco<sup>6</sup>

<sup>1</sup>Centro Medico Docente la Trinidad. Unidad Oftalmologica Caracas. AVAO, Caracas, Venezuela, <sup>2</sup>Centro Medico Docente La Trinidad, Caracas, Venezuela, <sup>3</sup>Asociacion venezolana Avance oftalmología AVAO, Caracas, Venezuela, <sup>4</sup>Asociacion Venezolana para avance oftalmología AVAO Caracas, Venezuela, <sup>5</sup>Centro Medico Docente la Trinidad Caracas, Venezuela, <sup>6</sup>Centro Medico Docente La trinidad Caracas, Venezuela

### Introduction:

Injections of dermal filler as autologous fat, collagen, hyaluronic acid, polylactic acid, calcium hydroxylapatite, and polymethylmethacrylate are used for facial rejuvenation. Complications for these procedures are cerebral ischemia and infarction, central retinal artery occlusion, ischemic optic neuropathy, ocular ischemia, palsy nerve ischemia, skin necrosis and even death. The mechanism of these event presumably via high pressure retrograde flow from the supratrochlear, supraorbital, and dorsal nasal artery to central retinal artery and long posterior ciliary artery occlusion. Platelet rich plasma have been used in plastic surgery by scars and skin rejuvenation.

### Methods:

A 61 year old woman developed painful severe visual loss immediately after platelet rich plasma injection into the supraorbital area. Skin redness was observed in the injection site. Her visual acuity was no light perception in the affected eye. The fundus of the eye showed diffused whitening of the retinal, retinal edema, cherry red spot in the fovea, and retinal arterioles attenuation. Fluorescein angiography demonstrated patchy choroidal non perfution and incomplete filling of the retinal arterioles in the later frames. Optical coherence tomography demonstrate retinal edema.

### Results:

Over time the followings fundus findings were observed: retinal edema decreased, diffuse whitening of the retinal persisted until patchy of pigment appeared over peripheral and macular area; lipid filled arterioles with perivascular sheathing until become in a phantom vessel, the optic disc become pale and pigmented in the superior pole. The fluorescein angiography demonstrated incompleted filling of the choroidal and arterial vessels. The optical coherence tomography demonstrate retinal thinning, hiperreflectivity areas, epiretinal membrane, loss of ganglions cells and nerve fiber layer. In the skin, necrosis in the injection area was observed.

### Conclusions:

Dermal injection of cosmetic filler injected in the forehead can cause irreversible blindness. The sequential fundus findings after cosmetic filler injections demonstrated the difficulty to reestablished vision.

### References:

1. Carle MV, Roe R, Novack R, Boyer DS:Cosmetic facial fillers and severe vision loss.JAMA Ophthalmol. 2014 May;132(5):637-9.

**Keywords:** Central Retinas Artera Oclusión, Cosmetic Filler

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Contrast Sensitivity Visual Acuity is Degraded in REM Sleep Behavior Disorder

Matthew J. Khayata<sup>1</sup>, Whitney B. Hough<sup>1</sup>, Peggy H. Vogt<sup>1</sup>, Eric M. Keasler<sup>1</sup>, Jacqueline M. Sanchez<sup>1</sup>, Garrett Barr<sup>1</sup>, David Y. Huang<sup>3</sup>, Charles G. Maitland<sup>1,2</sup>

<sup>1</sup>Florida State University College of Medicine, Tallahassee, FL, USA, <sup>2</sup>TMH Foundation Healthcare, Tallahassee, FL, USA,

<sup>3</sup>Tallahassee Pulmonary Clinic, Tallahassee, FL, USA

### Introduction:

REM Sleep Behavior Disorder (RBD) is a sleep disorder that produces motor enactment during REM sleep. Although pathophysiology is uncertain, studies demonstrate nigrostriatal dopamine deficiencies in RBD patients,<sup>1</sup> a condition frequently linked to alpha-synuclein-associated disorders i.e. Parkinson's disease (PD), characterized by cellular alpha-synuclein aggregation and death in dopamine producing cells. We previously reported contrast sensitivity visual acuity (CSVA) deficits in early-stage Parkinsonism.<sup>2</sup> A recent study identified that misfolded alpha-synuclein, a regulator of dopamine synthesis, is present in the inner retina of PD patients.<sup>3</sup> We therefore speculated that CSVA might be impaired in individuals with RBD.

### Methods:

To analyze CSVA in patients with RBD versus controls. 18 RBD patients and 29 control subjects. Participants completed a visual function questionnaire. Tests included: SLOAN CSVA wall charts at 100%, 2.5%, and 1.25%, and general ophthalmologic and neurologic examinations and Spectral Optical Coherence Tomography (OCT). Exclusion criteria: Visual acuity <20/40, co-morbid ophthalmologic pathologies, and dementia. Results were submitted to independent-samples t-test for statistical analysis.

### Results:

There was a statistically significant difference in mean contrast sensitivity between RBD and controls, with the RBD group scoring lower than the aged matched control group  $6.379 \pm 3.06$  [mean  $\pm$  standard error],  $t(45) = 2.08$ ,  $p = 0.043$ .

### Conclusions:

CSVA is diminished in patients with RBD. It seems plausible that visual deterioration is a consequence of misfolded alpha-synuclein in the inner retinal layers. Loss of CSVA in this population increases risk for falls.

### References:

1. Peever, Luppi, Montplaisir, Breakdown in REM sleep circuitry underlies REM sleep behavior disorder, Trends Neurosci, 37(5), 279-288, 2014
2. Nowalk, Matthews, Walley, Salmasinia, Maitland, Investigational Study on the Degree of Contrast Sensitivity Visual Acuity Defects in Early Stages of Parkinsonism, American Academy of Neurology, San Diego, CA, 2013
3. Bodis-Wollner, Kozlowski, Glazman, Miri,  $\alpha$ -synuclein in the inner retina in parkinson disease, Ann Neurol, 75(6), 964-966, 2014

**Keywords:** REM Sleep Behavior Disorder, Contrast Sensitivity Visual Acuity, Alpha-Synuclein

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 225

### Ophthalmoscopy Vs. MRI – Sometimes Less Is More.

Anton Kolomeyer<sup>1</sup>, Tarek Shazly, Joseph Martel, Gabrielle Bonhomme

*UPMC - Department of Ophthalmology, Pittsburgh, PA, USA*

#### Introduction:

The purpose is to describe a case of retinal disorder mimicking intracranial pathology.

#### Methods:

Case report.

#### Results:

56-year-old female presented to her local ophthalmologist with painless, progressive worsening vision and glare. Vision was 20/50 OD, 20/40 OS. Slit lamp examination showed cataracts and undilated fundus examination revealed macular pigmentary changes bilaterally that were attributed to early macular degeneration. Optical coherence tomography (OCT) showed thin ganglion cell complex and macula bilaterally. Subsequently, she underwent bilateral cataract extraction with intraocular lens placement. Postoperatively, vision improved to 20/25 OD, 20/30 OS. Four months later, she returned with decreased visual acuity of 20/40. Humphrey perimetry revealed dense temporal field loss nearly respecting the vertical midline OS and subtle dampening of superior nasal field OD, resembling an incongruous homonymous defect. Given these findings, MRI brain/orbits was obtained and showed mild microvascular disease but no structural abnormality. The patient was referred to Neuro-Ophthalmology where examination revealed an APD, dyschromatopsia, and left disc pallor, which further suggested optic neuropathy. However, dilated funduscopy revealed a large peripapillary area of retinal atrophy with yellowish peri-foveal deposits and vertical zone of pigmentary demarcation temporal to fovea. OCT macula revealed loss of ellipsoid layer corresponding to the region of abnormality visualized on fundoscopy. Serologic evaluation excluded inflammatory disorders, syphilis, tuberculosis, and TORCH infections. Cone-driven responses were undetectable on full-field ERG with inter-ocular difference in rod amplitude. Retina consultation was obtained and genetic counseling evaluation revealed consanguinity and presumptive diagnosis of autosomal recessive retinal dystrophy (genetic panel pending). Widefield color fundus, fluorescein angiography, and OCT all support a retinal etiology for her symptoms, with differential diagnoses including AZOOR, chronic chorioretinitis, and autoimmune retinopathy.

#### Conclusions:

Retinal pathology should be considered in the differential diagnosis of cases presenting with unilateral visual loss mimicking optic neuropathy, and evaluated appropriately prior to obtaining neuroimaging.

**References:** None.

**Keywords:** Retina, Visual Field, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

### Retinal Vessel Oximetry and Vessel Diameter Measurements: A Novel Metabolic Marker of Multiple Sclerosis

Marlen Lucero<sup>1</sup>, Amy L. Conger<sup>1</sup>, Darrel Conger<sup>1</sup>, Millad J. Sobhanian<sup>1</sup>, Victoria Stokes<sup>1</sup>, Teresa C. Frohman<sup>1</sup>, Owen White<sup>2</sup>, Laura J. Balcer<sup>3</sup>, Peter A. Calabresi<sup>4</sup>, Robert Rennaker<sup>5</sup>, Elliot M. Frohman<sup>1</sup>, Shin C. Beh<sup>1</sup>

<sup>1</sup>UT Southwestern Medical Center, Dallas, TX, USA, <sup>2</sup>Royal Melbourne Hospital, Parkville, Australia, <sup>3</sup>NYU Langone Medical Center, New York, NY, USA, <sup>4</sup>The Johns Hopkins University School of Medicine Baltimore, MD, USA, <sup>5</sup>The University of Texas at Dallas Richardson, TX, USA

#### Introduction:

Optic neuritis (ON), a common manifestation of multiple sclerosis (MS), often results in optic atrophy, a clinical finding characterized by optic disc pallor and retinal vessel attenuation. To date, there is no objective *in vivo* method for measuring the magnitude of pallor or vascular attenuation. We sought to characterize retinal vascular oximetry (oxygen saturation [SO<sub>2</sub>] and partial oxygen pressure [PO<sub>2</sub>]) and retinal vessel diameter in a cohort of MS patients using a novel technology – the Oxymap® T1 Retinal Oximeter.

#### Methods:

A cohort of healthy controls, and MS patients with a prior history of unilateral ON underwent assessments using spectral-domain optical coherence tomography and fundus photography. Further, using the Oxymap®, we measured the mean retinal vessel SO<sub>2</sub>, PO<sub>2</sub>, and corresponding diameter.

#### Results:

Compared to healthy controls, we found that the mean retinal vascular SO<sub>2</sub> and PO<sub>2</sub> were higher in MS patients, particularly in eyes with prior ON. Alternately, the mean retinal vessel diameter was reduced in the eyes of MS patients, when compared to those of healthy controls. The higher SO<sub>2</sub> and PO<sub>2</sub>, and reduced retinal vessel diameter corresponded to decreased retinal nerve fiber layer thickness (RNFLT).

#### Conclusions:

To our knowledge, we report the first application of objective, *in vivo*, ascertainment of retinal oxygenation and vessel diameter in MS patients, and confirm differentiation (especially ON affected eyes) from measures derived from controls. Further, we demonstrate an inverse relationship between increased SO<sub>2</sub> and PO<sub>2</sub>, and corresponding measures of RNFLT. Alternately, a direct relationship was identified between decreased retinal vessel diameter and RNFLT. We hypothesize that the pathobiological mechanisms in MS, culminate in retrograde degeneration of retinal ganglion cells, thereby reducing metabolic demand, oxygen extraction, and retinal vessel caliber. The application of retinal oximetry in MS, may facilitate the development of visual system biomarkers, germane to the identification of novel neurotherapeutic strategies in MS treatment trials.

**References:** None.

**Keywords:** Retina, Oximetry, Multiple Sclerosis, Optic Neuritis, Biomarker

**Financial Disclosures:** Teresa C. Frohman has received speaker fees for Biogen Idec, Novartis and Acorda and received compensation for participation on Scientific Advisory Board for Biogen Idec, Novartis and Questcor in the past year. Dr. Laura J. Balcer received personal compensation from Biogen Idec and consulting for Biogen Idec, Vaccinex and Genzyme. She is on a clinical trial advisory board for Biogen-Idec. Dr. Peter A. Calabresi received consulting honorarium from Abbott, Vaccinex, and Vertex. Grant support from Novartis, MedImmune and Biogen. Robert Rennaker is the owner of Vulintus LLC. Dr. Elliot M. Frohman has participated in the speakers' bureau for Teva Neurosciences, Acorda, and Novartis and has received consulting fees from TEVA Neurosciences, and Acorda. Dr. Shin C. Beh is the recipient of the 2012 Biogen Idec Clinical MS Fellowship award.

**Grant Support:** None.

## Poster 227

### Acute Zonal Occult Outer Retinopathy Presenting As Retrobulbar Optic Neuritis

Sungeun E. Kyung<sup>1</sup>, Jiwoong Park, Moohwan Chang

*Dankook university /ophthalmology, Cheonan, South Korea*

#### Introduction:

Acute zonal occult outer retinopathy (AZOOR) is an inflammatory retinopathy in the category of white dot syndromes characterized by acute loss of one or more zones of outer retinal function associated with photopsia, minimal fundusoscopic changes and abnormal electroretinography findings. The authors report a case of AZOOR in a patient presenting as retrobulbar optic neuritis.

#### Methods:

A 33 year-old male without underlying disease was referred with retrobulbar optic neuritis in the right eye. He presented with blurring in the right eye of 2 weeks duration. His visual acuity was 20/20 in left eye and 20/100 in the right eye with no ophthalmoscopic and fluorescein angiographic abnormalities. The cecocentral scotoma was found in the right eye. The amplitudes of pattern visual evoked potential (PVEP) was reduced in the right eye.

#### Results:

The amplitudes of the multifocal electroretinographics (ERGs) were reduced in the area of the cecocentral scotoma. Irregularities in the inner segment/outer segment (IS/OS) line of the photoreceptors were observed over the superior fovea by optical coherence tomography (OCT). The diagnosis of the AZOOR was made. The patient was followed with steroid treatment. The cecocentral scotoma disappeared in 3 months after the onset. At 6 months, the multifocal ERGs in the area corresponding to the cecocentral scotoma and irregularities in the IS/OS line on OCT were improved. His visual acuity was 20/20 in both eyes.

#### Conclusions:

The clinical distinction between a retinal versus an optic nerve problem may be difficult in case of subtle retinopathies. The multifocal ERGs and OCT images are helpful in this regards. The abnormalities of the inner/outer segment (IS/OS) junction and reduced amplitude corresponding to the scotoma area can be detected in patient with suspected AZOOR.

#### References:

1. Quillen DA, Davis JB, Gottlieb JL, Blodi BA, Callanan DG, Chang TS, et al. The white dot syndromes. American Journal of Ophthalmology. 2004;137(3):538-50.
2. Fujiwara T, Imamura Y, Giovannozzi VJ, Spaide RF: Fundus autofluorescence and optical coherence tomographic findings in acute zonal occult outer retinopathy. Retina 2010; 30:1206–1216.
3. Chai Y, Yamazaki H, Fujinami K, Tsunoda K, Yamamoto S: Case of acute zonal occult outer retinopathy with abnormal pattern visual evoked potentials. Clin Ophthalmol 2011; 5:1235–1241.

**Keywords:** Retina, Optic Neuropathy, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 228

### Long-term Follow-up of Laser Pointer Induced Macularopathy

Chuan-bin Sun<sup>1</sup>, Ting-ting Liu<sup>2</sup>, Ke Yao<sup>1</sup>

<sup>1</sup>Eye Center, Second Affiliated Hospital of Zhejiang University School of Medicine, Hangzhou, China, <sup>2</sup>Shierming Eye Hospital, Jinan, China

#### Introduction:

To evaluate the clinical characteristics and natural course of laser pointer induced macularopathy by long-term follow-up .

#### Methods:

14 cases (18 eyes) with laser pointer macularopathy were followed up for at least 6 months. Fundus photography, BCVA, visual field and OCT were examined at each follow-up, fundus fluorescein angiography (FFA) was tested in 10 patients at presentation.

#### Results:

At presentation, all patients with laser pointer macularopathy complained of sharp decline in BCVA accompanied by central scotoma immediately after laser exposure; fundus examination revealed yellow-white foveal lesions in 12 cases (16 eyes) and slightly elevated subfoveal hemorrhage in other 2 cases (2 eyes), which corresponded to a hyperreflexion between outer neuroretina and RPE layer or hyporeflection within neuroretina respectively by OCT, as well as late fluorescein staining or persistently blocked fluorescence respectively by FFA; Visual field test revealed central scotoma in all eyes. During follow-up, significant visual improvement was achieved within 2 weeks in most eyes except the eye with subfoveal hemorrhage, no matter oral steroid was used or not. At one month follow-up, most injured eyes showed BCVA of 0.8 or above, fundus examination showed foveal retinal depigmentation or mostly absorbed subfoveal hemorrhage, visual field test revealed relative or dense scotoma in all injured eyes. OCT shows the foveal pigment epithelium and cones IS/OS discontinuity. At 3 month and later follow-up, all patients showed normal BCVA and mild foveal depigmentation, although 2 cases (3 eyes) still complained of mild scotoma, which was proved by visual field test. OCT shows normal IS/ OS junction and mild RPE atrophy in most cases, while discontinued IS/ OS junction and RPE layer accompanied by cystic hyporeflection in outer neuroretina in eyes with scotoma complaint.

#### Conclusions:

High powered laser pointer can cause macular damage with different severity. Although visual prognosis is good in most patients, persistent scotoma might occur in some cases.

**References:** None.

**Keywords:** Macular Injury, Laser Pointer, Optical Coherence Tomography, Visual Field

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Use Of Wide-Field Retinography For Periphebitis Detection In Patients With Multiple Sclerosis.

Ruben Torres-Torres<sup>1,2</sup>, Elena Fraga-Pumar<sup>1,2</sup>, Pablo Villoslada<sup>1,2</sup>, Bernardo F Sanchez-Dalmau<sup>1,2</sup>

<sup>1</sup>*Institut Clinic d'Oftalmologia (ICOF), Hospital Clinic de Barcelona, Barcelona, Spain,* <sup>2</sup>*Center of Neuroimmunology.Institute of Biomedical Research August Pi Sunyer, Hospital Clinic, Barcelona, Spain,* <sup>3</sup>,

### Introduction:

Retinal periphebitis (RP) is characterized by perivascular exudation, hemorrhage, and retinal venous sheathing. The relationship between RP and multiple sclerosis (MS) is known and has a variable incidence among series (5-39%). RP has been related to a more active disease (1). Retinal inflammation parallels the inflammatory processes in the central nervous system and his presence of RP might favor the election of a more effective drug, even assuming a greater risk of side effects. The objective of this study was evaluate the role of wide-field retinography to detect periphebitis in patients without ocular symptoms.

### Methods:

A prospective cohort including 118 patients with remittent-recurrent multiple sclerosis (aged 18–55 years), underwent a ophthalmic examination, between May/2013 to May/2014. We collected demographic and MS-related variables, measured high- and low-contrast monocular visual acuity using ETDRS (Early Treatment Diabetic Retinopathy Study), Color vision was tested using Hardy-Rand-Ritter pseudoisochromatic plates, and ophthalmic fundus was registered using ultra-widefield scanning laser ophthalmoscopy (Optomap). Images were evaluated by and trained ophthalmologist.

### Results:

Six patients (5,08%) without ocular symptoms showed retinal venous sheathing, perivascular exudation and or hemorrhage compatible with periphebitis.

### Conclusions:

The incidence asymptomatic periphebitis detected with wide field retinography in our serie was similar to previous experience with slit lamp fundus ophthalmoscopy and pupil dilatation with tropicamide 1%. Ultra-widefield scanning laser ophthalmoscopy appears to be a useful tool for assessing easily the presence of phlebitis in patients with multiple sclerosis.

### References:

1. Ortiz-Pérez S, Martínez-Lapiscina EH, Gabilondo I, Fraga-Pumar E, Martínez-Heras E, Saiz A, Sanchez-Dalmau B, Villoslada P. Retinal periphebitis is associated with multiple sclerosis severity. *Neurology*. 2013 Sep 3;81(10):877-81.

**Keywords:** Demyelinating Disease, Diagnostic Tests, Retinography, Periphebitis, Multiple Sclerosis

**Financial Disclosures:** P. Villoslada has received consultancy fees from Roche, Novartis, MedImmune, TFS, Heidelberg Engineering, Digna, Biotech,Neurotech Pharma. And shareholder of Bionure.

**Grant Support:** Grant from the Instituto de Salud Carlos III, Spain. FIS Program PS09/00259) and RETICS program RD07/02060/01

**An Atypical Presentation of Giant Cell Arteritis**

Laurel N Vuong<sup>1</sup>, Thomas R Hedges III, Kendra Klein, Nora Laver, Frank McCabe

*Tufts-England Eye Center, Boston, MA, USA*

**Introduction:**

We observed an atypical presentation of giant cell arteritis that initially made the diagnosis difficult.

**Methods:**

A 71-year-old man began experiencing recurrent transient vision loss in the right eye in April 2014. Each episode lasted for 10-15 minutes and could sometimes be triggered by Valsalva maneuver. The episodes occurred 5-8 times daily, but gradually decreased in frequency and transitioned to episodes of "silver blurriness." He also experienced two episodes of "overall weakness" that lasted 10-15 minutes. At the end of May, he began to experience similar transient vision loss in the left eye. Fundus fluorescein angiography showed significant delayed retinal arterial, venous, and choroidal filling along with poor peripheral perfusion and microaneurysms bilaterally. ESR was 47mm/hr, brain MRI and head and neck MRA were unremarkable, and there was 25% blockage of the right carotid artery seen on carotid ultrasound. During our neuro-ophthalmologic evaluation in June, best corrected visual acuities were 20/40-1 OD and 20/25-2 OS, intraocular pressures were normal bilaterally, and there was no iris neovascularization. His funduscopic examination was unchanged. Automated visual field testing showed scattered non-specific changes OD and a paracentral scotoma OS. Five days later, he awoke with a persistent central scotoma in the left eye associated with headache, jaw pain, and bilateral temple tenderness.

**Results:**

Color Doppler of the orbits revealed no flow through the left posterior ciliary arteries with normal flow through the left central retinal artery. A temporal artery biopsy was consistent with giant cell arteritis. He was treated with three days of intravenous Solumedrol, followed by an oral Prednisone taper with no improvement in vision.

**Conclusions:**

Giant cell arteritis can have atypical presentations and should still be highly considered in elderly patients with recurrent transient vision loss. Immediate treatment with steroids is important to prevent vision loss in the fellow eye.

**References:**

1. Hutchinson, J. On a peculiar form of thrombotic arteritis of the aged which is sometimes productive of gangrene. Arch Surg. 1980;1:323-329.
2. Lipton RB, Solomon S, Wertenbaker C. Gradual loss and recovery of vision in temporal arteritis. Arch Intern Med. 1985;145:2252-2253.
3. Galor A and Lee MS. Slowly progressive vision loss in giant cell arteritis. Arch Ophthalmol. 2006;124:416-418.
4. Hedges TR. Ophthalmoscopic findings in internal carotid occlusion. Johns Hopkins Med J. 1962; 111:89-97.
5. Kearns TP, Younge BR, Piegras DG. Resolution of venous stasis retinopathy after carotid artery bypass surgery. Mayo Clin Proc. 1980;55:342-346.

**Keywords:** Vascular Disorders, Retina, Orbit/Ocular Pathology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Discordance Rates in Healing/Healed Arteritis in Temporal Artery Biopsies: The Role for Bilateral Biopsies

Sangsu Han<sup>1</sup>, Mustafa Kapasi<sup>1</sup>, Vivek Patel<sup>2</sup>, James Farmer<sup>3</sup>, Paula Blanco<sup>3</sup>, Danah Albreiki<sup>1</sup>

<sup>1</sup>Department of Ophthalmology, University of Ottawa, Ottawa, ON, Canada, <sup>2</sup>Department of Ophthalmology, University of Southern California, Los Angeles, CA, USA, <sup>3</sup>Department of Pathology and Laboratory Medicine, University of Ottawa, Ottawa, ON, Canada

### Introduction:

Temporal artery biopsy (TAB) is the gold standard diagnostic modality for giant cell arteritis (GCA). Since bilateral positivity for active GCA is 90+%, unilateral TAB is considered standard of care for diagnosing GCA. (1,2) However, GCA can present with active, healing/healed and no injury in a segmental uneven distribution along the extracranial carotid artery tree. Healing/healed arterial injuries are important to identify since they are usually treated as active GCA. We studied the discordance rate for healing/healed arterial injury in bilateral TAB to verify whether unilateral biopsies would render as high diagnostic yield as they do in active GCA.

### Methods:

We retrospectively studied TAB performed by two neuro-ophthalmologists at our institution during the last 4 years. We classified them into 3 categories: negative, healing/healed arterial injury, and active GCA. We defined healing/healed arterial injury as fibromyxoid intimal change, fragmentation and/or loss of the internal elastic lamina and medial scarring, with or without neovascularization. Focal areas of persistent chronic inflammation may remain. Statistical analysis was used to compare diagnostic sensitivity between both groups.

### Results:

Of the 257 TAB studied, 166 were negative, 56 were positive for active GCA and 35 showed healing/healed arterial injury. Fifty-two of the active GCA cases (52/56) and 34 of the healing/healed arterial injury cases (34/35) had bilateral TAB performed. Bilateral positivity was observed in 46/52 (88%) of active GCA cases, while 6/52 (12%) cases showed unilateral positivity. Healing/healed arterial injury was identified bilaterally in 11/34 (32%) cases, while unilateral lesions were observed in 23/34 (68%) cases. Thirty-four of the 35 cases (97%) with healing/healed arterial injury had high clinical suspicion and were treated as active GCA. Diagnostic sensitivity of unilateral TAB is significantly ( $p < 0.01$ ) lower for healing/healed arterial injury compared to active GCA.

### Conclusions:

Bilateral TAB should be standard of care to detect the characteristic histopathological features of healing/healed arterial injury.

### References:

1. Boyev LR, Miller NR, Green WR. Efficacy of unilateral versus bilateral temporal artery biopsies for the diagnosis of giant cell arteritis. *Am J Ophthalmol.* 1999; 128(2):211-215.
2. Melson MR, Weyand CM, Newman NJ, Biousse V. The diagnosis of giant cell arteritis. *Rev Neurol Dis.* 2007; 4(3):128-142

**Keywords:** Giant Cell Arteritis, Temporal Artery Biopsy

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## **Bilateral Intracranial Optic Nerve and Chiasmal Involvement in IgG-4 Related Disease**

Raed Behbehani<sup>1</sup>, Hmoud Al-Nomas <sup>2</sup>, Adeeba Al-herz<sup>3</sup>, Kenneth Katchi<sup>4</sup>

<sup>1</sup>Al-Bahar Ophthalmology Center, Kuwait, Kuwait, <sup>2</sup>Zain ENT Hospital, Kuwait, Kuwait, <sup>3</sup>Amiri Hospital / Department of Rheumatology, Kuwait, Kuwait, <sup>4</sup>Al-Sabah Hospital /Department of Pathology Kuwait, Kuwait

### **Introduction:**

IgG-4 related disease (IgG4-RD) is a disease that can affect many organs and is characterised by lymphoplasmocytic infiltration of IgG4-positive cells, obliterative phlebitis and storiform fibrosis. Orbital and ocular adnexal involvement in IgG4-RD has been reported with reports of optic nerve involvement. Systemic steroids are the primary treatment used in IgG4-RD. However, the visual outcome is not frequently reported. We present a case of severe loss of vision due to biopsy-proven IgG4-RD bilateral intracranial optic nerve and chiasmal involvement with pachymeningitis.

### **Methods:**

Case report and literature review

### **Results:**

The patient was treated with IV Rituximab which led to moderate improvement of visual function.

### **Conclusions:**

IgG4-RD can present as recurrent pachymeningitis and involvement the intracranial optic nerves and optic chiasm. Treatment with Rituximab may have role in the treatment of lesions of the visual pathway in IgG4-RD.

### **References:**

1. Stone, J.H., Y. Zen, and V. Deshpande, IgG4-related disease. N Engl J Med, 2012. 366(6): p. 539-51.
2. Umehara, H., et al., [IgG4-related disease -a new clinical entity established by all Japan IgG4 team in 21st century-]. Arerugi, 2013. 62(12): p. 1591-7.
3. Umehara, H., et al., IgG4-related disease and its pathogenesis- cross-talk between innate and acquired immunity. Int Immunol, 2014.
4. Pasquali, T., et al., Orbital inflammation in IgG4-related sclerosing disease. Orbit, 2011. 30(5): p. 258-60.

**Keywords:** IgG4-Related Disease, Pachymeningitis, Rituximab, Optic Neuropathy, Chiasmal Syndrome

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Neuromyelitis Optica Preceded Be Seizure Disorder

Iris Ben Bassat Mizrahi<sup>1</sup>, Bruria Ben Zeev<sup>2, 3</sup>

<sup>1</sup>*Goldschleger Eye Institute, Sheba Medical Center, Ramat Gan, Israel*, <sup>2</sup>*Pediatric Neurology unit, Safra pediatric hospital, Sheba Medical Center, Ramat Gan, Israel*, <sup>3</sup>*Sackler Medical School Tel Aviv University, Tel Aviv, Israel*

### Introduction:

We present a case of a girl with Sturge Weber and NMO in which the seizure activity and the NMO attacks concur.

### Methods:

Case report and review of literature

### Results:

An 11-year-old female with past medical history significant for IUGR at birth, cleft palate as part of Pierre Robin sequence, and Sturge Weber syndrome diagnosed at the age of three due to seizures. She was otherwise neurologically intact until the age of 11 when she had her first seizure in 8 years. Following the seizure she started complaining of tingling in left arm and hand that quickly progressed to weakness, and was diagnosed with transverse myelitis. Weakness gradually improved after three days of IV methylprednisolone treatment. NMO –IgG were positive and she was started on azathioprine. A month after initiation of treatment she experienced a couple of focal seizures and one episode of generalized seizure with urinary incontinence, after which she complained of vision loss in the left eye and was diagnosed with optic neuritis. She was treated with rituximab followed by IVIG, but 3 month later she came in with another episode of optic neuritis in the same eye that was preceded by a generalized seizure. Given the rituximab failure she was treated with IV steroids again, PLEX for 5 consecutive days, mycophenolate mofetil and levetiracetam.

### Conclusions:

This case report demonstrates a temporal relationship between the seizure activity and NMO attacks. There is no known pathogenic link among these two condition and we believe that the disruption of the blood-brain barrier (BBB) could explain this concurrent appearance. It is unclear if BBB disruption by the seizure promotes the NMO attack or vice versa.

### References:

1. Suzuki N, Takahashi T, Aoki M, Konohana S, Okumura T et al, Neuromyelitis optica precede by hyperCKemia episode. *Neurology* 74:1543-1545;2010
2. Vincent T, Saikali P, Cayrol R, Roth AD, Bar-Or A, et al, Functional consequences of Neuromyelitis Optica -IgG astrocyte interactions on blood-brain barrier permeability and granulocyte recruitment. *J Immunol* 181: 5730-5737; 2008
3. Li YJ, Wang ZH, Zhang B, Wang MJ, Shi ST, et al, Disruption of blood-brain barrier after generalized tonic-clonic seizure correlates with cerebrospinal fluid MMP-9 levels. *J of Neuroinflammation* 10:80-90; 2013
4. Marchi N, Angelov L, Masaryk T, Fazio V, Granata T et al, Seizure-Promoting effect of blood-brain barrier disruption. *Epilepsia* 48(4):732-742, 2007.

**Keywords:** Neuromyelitis Optica, Seizures, Blood-Brain Barrier

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 234

### Optic Nerve Meningioma Masquerading as Neurosarcoidosis

Ryan C. Burton<sup>1</sup>, Michael S. Vaphiades<sup>1,2,3</sup>, Matthew G. Vicinanzo<sup>4</sup>

<sup>1</sup>University of Alabama, Department of Ophthalmology, Birmingham, AL, USA, <sup>2</sup>University of Alabama at Birmingham, Department of Neurology, Birmingham, AL, USA, <sup>3</sup>University of Alabama at Birmingham, Department of Neurosurgery, Birmingham, AL, USA, <sup>4</sup>Alabama Ophthalmology Associates Birmingham, AL, USA

#### Introduction:

Meningiomas are typically benign tumors often affecting middle-aged women. The orbit is a common location for meningiomas, with many of these involving the optic nerve<sup>1</sup>. These optic nerve sheath meningiomas may have a variety of presentations, including optic atrophy, papilledema, or shunt vessels<sup>2</sup>. Rarely, neurosarcoidosis can mimic an optic nerve sheath meningioma due to their similar appearance on radioimaging<sup>3</sup>.

#### Methods:

A case report.

#### Results:

A 25-year-old healthy woman with a two-year history of facial lesions, previously diagnosed as cystic acne, presented with normal visual acuity and color vision. She had been diagnosed with right optic nerve edema on a routine eye exam. Magnetic resonance imaging (MRI) revealed a right intraconal enhancing mass. The patient had two skin biopsies that revealed non-caseating granulomatous inflammation felt to represent sarcoidosis. Given this finding, the optic nerve lesion was attributed to neurosarcoidosis and the patient was treated with intravenous and oral prednisone. Six months later, she presented with a substantial decline in visual acuity to hand motions vision in the right eye. She was also unable to identify any color plates with the right eye and had worsened optic nerve swelling. A repeat MRI showed a slightly larger enhancing lesion. Due to a decline in vision and lack of response to systemic therapy, an optic nerve biopsy and partial tumor excision was performed. Pathologic examination revealed an optic nerve meningioma. The patient was referred to radiation oncology for further therapy.

#### Conclusions:

In this case, biopsy-proven cutaneous sarcoidosis with an optic nerve lesion prompted a diagnosis of neurosarcoidosis. The true diagnosis was not confirmed until months later when the patient's vision significantly declined, prompting an optic nerve biopsy that provided the correct diagnosis of optic nerve meningioma.

#### References:

1. Vukovic Arar Z, Vatauvuk Z, Miskic B, et al. Optic nerve sheath meningioma: a case report with 15-year follow up. *Semin Ophthalmol.* 29, 52-5, 2014.
2. Saeed P, Rootman J, Nugent RA, et al. Optic nerve sheath meningiomas. *Ophthalmology.* 110, 2019-30, 2013.
3. Jennings JW, Rojiani AM, Brem SS, Murtagh FR. Necrotizing neurosarcoidosis masquerading as a left optic nerve meningioma: case report. *Am J Neuroradiol.* 23, 660-2, 2002.

**Keywords:** Neuroimaging, Tumors, Orbit, Orbit/Ocular pathology

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This work was supported in part by an unrestricted grant from the Research to Prevent Blindness, Inc. N.Y., N.Y.

**Comparison of the Clinical Characteristics of Patients with Active Arteritis and Healed Arteritis, the Two Histopathological Patterns Considered Positive in Giant Cell Arteritis**

Michael W. Salter<sup>1</sup>, Joseph G. Chacko, Arthur J. Stanley, Harry H. Brown

UAMS, Little Rock, AR, USA

**Introduction:**

Histopathologically, there are two patterns considered diagnostic of giant cell arteritis (GCA): those with inflammation of the vessel wall (active arteritis) and those with post-inflammatory alterations (healed arteritis). The aim of our study was to determine the clinical characteristics and outcomes for patients with these two types of positive temporal artery biopsy.

**Methods:**

An IRB-approved retrospective review was conducted of all patients with a positive temporal artery biopsy between 2004 and 2013. Twenty-two patients with biopsy-proven GCA were found. All biopsy specimens had been examined and interpreted by a single ocular pathologist. Eleven patients had active arteritis and eleven patients had healed arteritis. We compared presenting symptoms, ischemic ocular events, inflammatory markers (erythrocyte sedimentation rate [ESR], C-reactive protein [CRP], and platelet count), relapses, and dosage requirements of steroids between the two groups.

**Results:**

Seven of the 11 patients with active arteritis had an initial ocular ischemic event compared to 3 of the 11 patients with healed arteritis. There was no statistical difference in initial ESR between the two groups, but CRP and platelet counts on initial presentation were statistically higher in the active group ( $p = 0.0002$  and  $p < 0.0001$  respectively). Patients with active arteritis on biopsy required higher doses of steroids over a 2-year follow-up compared to the healed group: on average, 11 mg/day to 1 mg/day at 1 year ( $p = 0.0008$ ), and 7 mg/day to 0.5 mg/day at 2 years ( $p = 0.0208$ ), respectively. During the follow-up period, 2 of the 11 patients in the active group demonstrated a recurrent ischemic ocular event either to the same or fellow eye while in the healed group there were no recurrent ischemic ocular events.

**Conclusions:**

Patients with a healed arteritis pattern on histopathological examination of temporal artery biopsy appear to have a better prognosis and may require less aggressive treatment than those with active inflammation.

**References:**

1. Aiello, Trautmann, McPhee, et al. Visual prognosis in giant cell arteritis. *Ophthalmology*; 100: 550-555; 1993.
2. Murchison, Gilbert, Biky, et al. Validity of the American College of Rheumatology Criteria for the Diagnosis of Giant Cell Arteritis. *American Journal of Ophthalmology* 154: 617-619; 2012.
3. Borg, Haanen, Seldenrijk, et al. Relationship between histological subtypes and clinical characteristics at presentation and outcome in biopsy-proven temporal arteritis. *Clin Rheumatol*; 26:559-532; 2007.
4. Lee, Padera, Noss, et al. Clinical Course and Management of a Consecutive Series of Patients with "Healed Temporal Arteritis". *J Rheum*; 39:295-302; 2012.
5. Foroozan, Danesh-Meyer, Savino, et al. Thrombocytosis in patients with biopsy-proven giant cell arteritis. *Ophthalmology*; 109:1267-71; 2007.

**Keywords:** Optic Neuropathy, Neuro-Ophth And Systemic Disease

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** This work was supported in part by Research to Prevent Blindness and the Pat & Willard Walker Eye Research Center.

**Autologous Hematopoietic Stem Cell Transplantation in Neuromyelitis Optica - An Update**

Fiona Costello<sup>1</sup>, Jodie Burton, Luanne Metz, Peter Duggan, Jan Storek

*University of Calgary, Calgary, AB, Canada*

**Introduction:**

Neuromyelitis Optica (NMO) is an immune-mediated astrocytopathy characterized by frequent demyelinating/necrotizing attacks of optic nerves and spinal cord. Treatments are highly toxic and incompletely effective. Fifty percent of patients are blind in one eye or require a mobility aide within 5 years. The immunological features of NMO, coupled with its severity make it an ideal candidate for a trial of autologous non-ablative hematopoietic stem cell transplantation (AHSCT) with the goal of disease remission and freedom from immunosuppressive medications. The objective of this study is to determine if NMO patients, despite maintenance therapy, experience a reduction in relapses and disability without ongoing immunosuppressants after AHSCT. We hypothesize a greater than or equal to 50% reduction in the proportion of patients relapsing 3 years post-transplant.

**Methods:**

Patients 18-65 with greater than or equal to 1 relapse in 12 months, or 2 (or more) relapses in 24 months, despite immunotherapy and EDSS less than 6.5 are eligible. Patients undergo non-ablative stem cell mobilization and infusion using cyclophosphamide, rituximab and ATG. Outcomes include EDSS, NMO-IgG titers, OCT and MRI q6-12 months for 5 years. Ten subjects provide 80% power for our primary outcome.

**Results:**

Three NMO patients have undergone transplantation thus far. A 28F was transplanted in May 2011 with pre-transplant annualized relapse rate (ARR) of 5 and EDSS 4.5, now 0 and 2.0 at month 41 respectively. A 36F was transplanted in April 2012 with pre-transplant ARR of 5, now 1 and 3.5 at month 30 respectively, who started on mycophenolate mofetil at her request. A 39M was transplanted in January 2014 with pre-transplant ARR of 1.3 and EDSS 3.5 who remains stable at 10 months. Despite clinical improvement in all patients, the latter two patients remain NMO-IgG seropositive.

**Conclusions:**

Thus far, this AHSCT regimen is well tolerated with marked clinical improvement. Additional patients and longer follow-up will determine if these benefits persist.

**References:** None.

**Keywords:** Neuromyelitis Optica, Optic Neuritis, Autologous Hematopoietic Stem Cell Transplantation, Clinical Recovery

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Poster 237****Social Media and Susac Syndrome**

Robert A Egan<sup>1</sup>

*Oregon Neurology, Tualatin, OR, USA*

**Introduction:**

The advent of social media has revolutionized societal communication. Patients also use this medium to better educate themselves. Facebook is one of the leading social media forums and is utilized by sufferers of Susac Syndrome (SS) and caregivers. The impact of social media on impressions of disease states is unknown. The current study attempts to answer several questions regarding patient usage of Facebook.

**Methods:**

Users of the Susac Syndrome Facebook Group (SSFG) were submitted to a 44 item questionnaire. Several questions were directed at perceptions of disease state and interactions with fellow users via a linear 5-point scale.

**Results:**

On 10/30/2014, the SSFG contained 479 members and 37 subjects (7.7%) returned questionnaires. Twenty seven (73%) were female and mean age of symptom onset was 32.3 years (16-54). The first symptom was visual in 13 (35%) and auditory in 8 (22%). Fourteen (38%) were initially diagnosed with multiple sclerosis and 30 (81%) were seen by two or more physicians prior to their allegedly correct diagnosis. Regarding feeling listened to by other users of the SSFG, 30 (81%) listed a 5 on the linear scale indicating strong agreement. Support by other users scored a 5 in 25 (68%) and 23 (62%) listed a 5 in relation to presentation of useful information by other SSFG users. However only nine (24%) felt that their treatment was fully effective.

**Conclusions:**

Most users of the SSFG had very favorable experiences. Very few agreed that the disease was effectively treated however. A limitation of this study is the definitive diagnosis of SS sufferers could not be confirmed. Future study should be directed at the impact of the SSFG on parameters such as vision, cognition, or hearing or if subsequent users received an earlier correct diagnosis reducing disease morbidity.

**References:** None.

**Keywords:** Susac Syndrome, Social Media, Vascular Disorders, Neuro-Ophth & Systemic Disease

**Financial Disclosures:** The author had no disclosures.

**Grant Support:** None.

## Horner's Syndrome as Initial Manifestation of a Malignant Peripheral Nerve Sheath Tumor

Gerard L. Hershewe<sup>2</sup>, Micaela M. Koci<sup>1</sup>, Javier A. Rodriguez<sup>1</sup>, Shayne M. Ahwah<sup>1</sup>, Josh C. Gratwohl<sup>1</sup>, Mia Cozad<sup>1</sup>

<sup>1</sup>University of Nevada, Reno, Reno, NV, USA, <sup>2</sup>University of Nevada School of Medicine, Reno, NV, USA

### Introduction:

Horner's Syndrome may be the rare presenting manifestation of an occult malignancy. In a previous study, 13% of 450 patients presenting with Horner's Syndrome had tumors, but only 3% had undetected malignancies. This case illustrates a patient who presented with Horner's Syndrome several months prior to his neuroimaging studies and diagnosis.

### Methods:

Case Report.

### Results:

We report a 56-year-old male who has a history of Neurofibromatosis Type I diagnosed at age 15. Within the past year, the patient began to notice a droopy lid OD of several months duration. Subsequently he began to have difficulty swallowing both liquids and solids, associated with 100-pound weight loss over a period of 6 months. His physical examination showed bilateral Lisch Nodules, bilateral optic nerve hypoplasia, multiple cutaneous neurofibromas, palpable neck mass and café au-lait spots. The cocaine test was strongly positive confirming the diagnosis of Horner's Syndrome. The degree of anisocoria following the administration of cocaine was greater than 2.2 mm. Neuroimaging studies demonstrated a large right parapharyngeal space soft tissue tumor, measuring up to 6 cm, stretching and displacing the carotid sheath. The patient underwent surgical resection of the tumor and pathology was consistent with a malignant peripheral nerve sheath tumor. It is noteworthy that these peripheral nerve sheath tumors often arise from pre-existing neurofibromas, and up to 50% of these tumors occur in patients with Neurofibromatosis Type I.

### Conclusions:

A sarcoma is specifically defined as a malignant peripheral nerve sheath tumor when it arises from a preexisting benign nerve sheath tumor (neurofibroma). The patient underwent subtotal resection of the tumor, however he developed pulmonary metastasis, which portends a poor prognosis. This case illustrates that patients suspected of having Horner's Syndrome require prompt attention, specific pharmacologic testing with regards to neuroanatomical localization and appropriate neuroimaging studies and investigations.

### References:

1. Geller, DS, Gebhardt, M, Malignant Peripheral Nerve Sheath Tumors. Mayo Clinic, 2013.
2. Maloney, WF, Younge, BR, Moyer, NJ, Evaluation of the causes and accuracy of pharmacological localization in Horner's syndrome. American Journal of Ophthalmology, 90(3): 394-402, 1980.

**Keywords:** Horner's Syndrome, Neurofibromatosis, Malignant Peripheral Nerve Sheath Tumor, Neuroimaging

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## **Birefringence of Retinal Nerve Fiber Layer and Retinal Blood Flow Velocity in Multiple Sclerosis**

Hong Jiang<sup>1,2</sup>, Jin Yuan<sup>1,4</sup>, Silvia Delgado<sup>2</sup>, Wentao Yan<sup>3</sup>, Delia Cabrera DeBuc<sup>1</sup>, Byron Lam<sup>1</sup>, Jianhua Wang<sup>1</sup>

<sup>1</sup>Bascom Palmer Eye Institute/University of Miami, Miami, FL, USA, <sup>2</sup>Department of Neurology/University of Miami, Miami, FL, USA, <sup>3</sup>School of Ophthalmology and optometry/Wenzhou Medical College, Wenzhou, China, <sup>4</sup>Zhongshan Ophthalmic Center/Sun Yat-sen University Guangzhou, China

### **Introduction:**

Cerebral hypoperfusion in the normal-appearing white matter of MS patients are reported, indicating the role of vascular dysfunction in MS pathophysiology. The retina has been used to study the pathophysiology of MS. Retinal nerve fiber layer (RNFL) thinning is established as an ocular biomarker of neurodegeneration. The goal of this project is to determine the relationship between the birefringence of RNFL (microtubule integrity), retinal blood flow velocity and clinical MS manifestation.

### **Methods:**

Custom built polarization sensitive optical coherence tomography (PS-OCT) was used to measure the averaged birefringence and thickness of the peripapillary RNFL. A retinal function imager (RFI, Optical Imaging Ltd, Rehovot, Israel) was used to study the velocity of blood flow of retinal arterioles and venules. A clinical OCT (Zeiss Cirrus) was used to measure the peripapillary RNFL thickness and macular ganglion cell layer (GCL) thickness. Seven MS patients were clinically assessed and recruited (averaged age  $48.4 \pm 14.0$  yrs old, 6 females and 1 male).

### **Results:**

PS-OCT derived birefringence of the peripapillary RNFL is related to RNFL thickness measured by Cirrus OCT ( $r = 0.65$ ,  $P < 0.05$ ) and PS-OCT ( $r = 0.43$ ,  $P < 0.05$ ). The birefringence is also related to blood flow velocities in arterioles ( $r = 0.40$ ,  $P < 0.05$ ) and venules ( $r = 0.68$ ,  $P < 0.05$ ). The birefringence is not related to EDSS scores, but related with disease duration ( $r = -0.86$ ,  $P < 0.05$ ). In addition, the birefringence is related to GCL thickness ( $r = 0.66$ ,  $P < 0.05$ ).

### **Conclusions:**

This is the first study using PS-OCT to study the RNFL birefringence and its relationship with retinal microvascular function. The strong correlation between these two measurements indicates the coexistence of RNFL microtubule dysfunction and retinal microvascular dysfunction. Further studies with a large sample of MS patients are needed to further elucidate the findings.

### **References:** None.

**Keywords:** Demyelinating Disease, Neuro-Ophth & Systemic Disease (MS), Retina, Polarization Sensitive OCT & Birefringence, Retinal Microvascular Function

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Supported by a grant from National Multiple Sclerosis Society, R01EY020607, NIH R01EY020607S, NIH Center Grant P30 EY014801 and RPB.

## Poster 240

### Botox For Chronic Intractable Headache Among Veterans At VA Medical Center, Long Beach, California

Grace W. Kao<sup>1</sup>, Wanda Hoffart, Steven Schreiber

*Veterans Affairs Healthcare System, Neurology Clinic, Long Beach, CA, USA*

#### Introduction:

Veterans with chronic migraine often associate with comorbidity including traumatic brain injury (TBI), PTSD, control medication addition ( RxA) and occipital headache with cervical spondylosis(C-DJD). Botox (Onabotulinumtoxin A) has not been well reported the efficacy for veterans with chronic migraine and multiple comorbidity. We review the effect of Botox in this group of veterans at the Neurological Clinic, VA Long Beach CA to determine the impact of these comorbidities on the efficacy of Botox.

#### Methods:

The veterans with chronic migraine (more than 4 hours daily and 15 days monthly) who failed 3 out of 4 prophylactic medications (TCA, beta-blocker, topiramate, divalproex) without contraindication to Botox were recruited. The comorbidities were reviewed. Botox injection was performed according to 2010 Allergan Inc. protocol. The efficacy was graded by percentage of reduce in intensity and days of headache along the use frequency of acute medications at end of 3 months.

#### Results:

16 patients ( 9 F, 7 M; age 29-67, average 43 y) with chronic migraine for average of 16.1 years ( 2- 41 years) received treatments ( 1 to 8 times) from 9/2012 to 10/2014 with follow-up of 3 to 25 months. Totally 70 treatments were performed. Nine (56 %), 8 (50%), 5 (31%) and 7 (44%) patients had PTSD, RxA, TBI and C-DJD respectively. Four ( 25% ), 7 (43.7%), 1 ( 6.2%), and 2 (12.5 %) patients had 1, 2, 3 and 4 comorbidities respectively. Twelve patients (75%) reported satisfactory response with severe headache ( > 7 on scale) reduced to less than 8 days monthly; 6 of them had severe headache less than 3 days monthly. Among the 70 treatments, 53 (75.7% ) reduced severe headache to less than 8 days, and 38 (54%) after the 3rd treatment reduced to less than 3 days monthly.

#### Conclusions:

The comorbidities do not impact the Botox effect on treating the chronic migraine among the veterans. The small sample size cannot determine the individual comorbidity against Botox treatment. The favorable response among PTSD or control-medicine addicted veterans will be better determined by long term and larger sample-sized study.

#### References:

1. Loder, E, Biondi, D, Use of Botulinum Toxins for Chronic headaches: A focused Review. Clinical J. of Pain, 2002, PS169-176.
2. Carlson KF, Taylor BC, et al, Headache diagnoses among Iraq and Afghanistan war veterans enrolled in VA: a gender comparison. Headache. 2013;53(10):1573-82.

**Keywords:** Migraine, Headache, Botox

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 241

### Thin-Film Optical Notch Filter Spectacle Coatings for the Treatment of Migraine and Photophobia

Bradley J Katz<sup>1</sup>, Ryan N Hoggan<sup>1</sup>, Amith Subhash<sup>1</sup>, Steve Blair<sup>2</sup>, Kathleen B Digre<sup>1</sup>, Susan K Baggaley<sup>3</sup>, Jamison Gordon<sup>1</sup>, K C Brennan<sup>3</sup>, Judith E A Warner<sup>1</sup>, Alison V Crum<sup>1</sup>

<sup>1</sup>University of Utah/John A Moran Eye Center, Salt Lake City, UT, USA, <sup>2</sup>University of Utah/Electrical and Computer Engineering, Salt Lake City, UT, USA, <sup>3</sup>University of Utah/Neurology, Salt Lake City, UT, USA

#### Introduction:

Previous evidence suggests that optical treatments hold some promise for treating migraine and photophobia. We designed an optical notch filter, centered at 480 nm to reduce the direct stimulation of intrinsically photosensitive retinal ganglion cells. We used thin film technology to integrate the filter into spectacle lenses that could be worn by migraine patients.

#### Methods:

We designed a prospective, randomized, double-masked crossover study. Our primary endpoint was the 6-item Headache Impact Test (HIT-6). We developed two thin-film optical notch filters: the therapeutic filter blocked visible light at 480 nm, while a 620 nm filter was designed to act as a sham filter. Each participant was asked to wear a set of lenses with one of the filters for two weeks; after a 2-week washout period during which no study lenses were worn, the participant wore lenses with the other filter for two weeks

#### Results:

We enrolled 48 subjects with chronic migraine and 37 completed all study procedures. Wearing either the 480 nm or the 620 nm spectacle lenses resulted in clinically and statistically significant reductions in HIT-6 score. However, there was no significant difference when comparing the overall effect of the 480 nm lenses and the 620 nm lenses.

#### Conclusions:

Although the 620 nm filter was designed as a sham intervention, basic science research published since the institution of the trial has indicated that melanopsin, the photopigment found in intrinsically photosensitive retinal ganglion cells, is bistable. This property of the melanopsin molecule may explain the unexpected efficacy of the 620 nm filter. The effect of the two filters does not appear to be a placebo effect and we conclude that spectacle lenses outfitted with a thin-film optical notch filter may be a useful adjunct in the treatment of chronic migraine.

#### References:

None.

**Keywords:** Neuro-Ophth & Systemic Disease, Migraine, Photophobia, Light Sensitivity, Intrinsically Photosensitive Retinal Ganglion Cells

**Financial Disclosures:** Drs. Katz and Blair have equity interests in and hold management positions in Axon Optics, a limited liability corporation that markets eyewear for the treatment of migraine and photophobia. Drs. Katz, Blair, Digre and Warner are inventors on a patent pending for the thin-film coatings described in this research and stand to receive royalties on any commercial sales of products based on these coatings.

**Grant Support:** This investigation was supported in part by: Axon Optics, LLC, Salt Lake City, UT NIH T35HL007744 (RNH, AS) The University of Utah Study Design and Biostatistics Center, with funding in part from the National Center for Research Resources and the National Center for Advancing Translational Sciences, National Institutes of Health, through Grant 8UL1TR000105 (formerly UL1RR025764) and an unrestricted grant from Research to Prevent Blindness, Inc., New York, New York, USA, to the Department of Ophthalmology and Visual Sciences, University of Utah, Salt Lake City, Utah. JG was supported by the University of Utah, Office of Undergraduate Research, Undergraduate Research Opportunities Program (UROP). RNH and AS were supported by the Medical Student Research Program at the University of Utah School of Medicine, funded by NIH/NHLBI Training Grant # T35 HL007744 (PI: Jerry Kaplan, PhD) and University of Utah institutional support.

## Poster 242

### Monitoring Alzheimer's Disease with Retina Examination

Umur A. Kayabasi<sup>1</sup>, Robert C. Sergott<sup>2</sup>

<sup>1</sup>World Eye Hospital, Istanbul, Turkey, <sup>2</sup>Wills Eye Hospital, Philadelphia, PA, USA

#### Introduction:

We aimed to monitor patients with Alzheimer's Disease ( AD ) with retina examination using optical coherent tomography ( OCT) and fundus autofluorescein ( FAF ) .

#### Methods:

30 patients with mild cognitive impairment ( MCI ) were examined for 6 months. At the 3rd and 6th months, OCT and FAF tests were repeated. Curcumin ( Turmeric Phytosome with Meriva ) was also given 80 mg bid for 3 days. We tried to detect new changes on OCT and FAF. Any change in curcumin stained lesions ( increase in number and size ) and appearance of lesions on FAF were taken into account. No other tests like PET- CT were ordered. We also followed up 10 healthy age- matched controls at the same intervals.

#### Results:

In 8 patients progression in neurodegeneration was detected in the follow up. In 5 patients, appearance of the lesions on FAF changed. Hyperfluorescent images became hypofluorescent which demonstrated atrophy of RPE . In 3 patients new curcumin stained beta amyloid plaques were detected which was consistent with increased amyloid burden. In 22 patients images showed stable lesions. No defects were found in the control group. The patients in whom progression was found were sent for neurologic examination. Either drug dosages were changed or new drugs were started.

#### Conclusions:

We believe that OCT and FAF examinations with curcumin are very useful and trustable in the follow up of AD patients.

**References:** None.

**Keywords:** Monitoring, Alzheimer's Disease, OCT, FAF, Retina

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 243

### Why is He Losing His Sight? He is Losing His Photoreceptor!

Ainat Klein<sup>1</sup>, Efrat Fleissig, Anat Kesler

*Tel-Aviv Medical Center, Ophthalmology Department, Neuro-ophthalmology unit, Tel Aviv, Israel*

#### **Introduction:**

Retinitis pigmentosa has a wide spectrum of clinical and retinal presentation and can lead to progressive visual field loss. Papilledema can also lead to visual field defects and decreased vision. We describe here an unusual case of combined optic neuropathy and retinal degeneration leading to vision loss.

#### **Methods:**

Case report presentation.

#### **Results:**

26 years old male with history of nonspecific myopathy (biopsy proven) and chronic mild renal failure (biopsy showed focal segmental glomerulonephritis), was referred to the ophthalmology clinic due to difficulties in his vision especially at night. On exam visual acuity was 20/25 in both eyes, with mildly constricted visual fields. Fundus showed normal optic disc color and arteriolar narrowing. Eelectroretinography was compatible with combined rod and cone diffuse dystrophy. 2 years later he reported progressive decrease vision, headache and tinnitus. He had a vision of 20/30 and tunnel vision of central 10 degrees. The retina had only few pigmented spicules and he had chronic optic disc edema. Investigation revealed increased intracranial hypertension with elevated protein. Treatment with topiramate was initiated. High Resolution Optical coherence tomography (HRT-OCT) showed diffuse loss of photoreceptors with only less than 1  $\mu\text{m}^2$  of subfoveal photoreceptors area. We presume that the main reason for the vision loss in this patient was the progressive retinal degeneration and not optic neuropathy. He is currently treated by oral topiramate with good compliance.

#### **Conclusions:**

We describe a rare case of combined syndrome of myopathy, nephropathy, Retinitis pigmentosa and elevated intracranial pressure, who experienced progressive visual field loss.

**References:** None.

**Keywords:** Increased Intracranial Hypertension, Papilledema, Retinal Degenration, HRT-OCT

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Extensive Macular Serous Retinal Detachment As The Initial Presentation Of *Bartonella Henselae* Induced Neuroretinitis**

Darrell Lewis<sup>1</sup>, Aditya Mishra<sup>2</sup>, Vladimir Kozousek<sup>3</sup>

<sup>1</sup>Dalhousie University/Ophthalmology Resident, Halifax, NS, Canada, <sup>2</sup>Dalhousie University/Neuro-Ophthalmology, Halifax, NS, Canada, <sup>3</sup>Dalhousie University/Medical Retina, Halifax, NS, Canada

**Introduction:**

The clinical appearance of *Bartonella henselae* associated neuroretinitis is thought to develop primarily from prelaminae optic nerve head inflammation leading to disk edema and serous retinal detachment can occur. A macular star classically develops, however many cases without star formation have been reported. We report a case of extensive serous macular detachment without clinically apparent optic disk changes or macular star appearance.

**Results:**

A 28 year old female presented with a two day history of decreased vision, mild pain in the left eye. She had been scratched by a cat, although not recently. Visual acuity measured 6/9 OD and 6/120 OS with a small left RAPD. On dilated fundoscopic examination the optic nerves did not appear elevated and their margins were sharply defined. A serous macular retinal detachment (~6 500 µm in diameter) was noted in the left eye (right eye within normal limits). Fluorescein angiography showed progressive fluorescence at the optic nerve head consistent with leakage and mild pooling within the macula. There was no evidence of RPE damage, focal leakage, or choroidal neovascularization. Optical coherence tomography confirmed a neurosensory retinal detachment. CT and MRI scans were found to be normal. CRP, syphilis serology, ANA, ACE level and a chest x-ray were all negative or within normal levels. Bartonella serology revealed an IgG titre of 1:64 indicating prior exposure, although a time course could not be established. Her clinical findings spontaneously resolved over one month. Stellate macular exudates did not develop. Subsequent fluorescein angiography and indocyanine green angiography did not reveal any abnormalities.

**Conclusions:**

Our case is an atypical presentation of Bartonella neuroretinitis. There are only two other case reports of serous neurosensory retinal detachment without clinically apparent optic nerve swelling. Our case highlights the importance of performing fluorescein angiography in this setting and screening for Bartonella in patients with neurosensory detachment accompanied by angiographic leakage at the optic nerve head.

**References:**

1. Zacchei, Newman, Sternberg, Serous Retinal Detachment of the Macula Associated With Cat Scratch Disease, American Journal of Ophthalmology, Vol 120, 796-797, 1995
2. Asensio-Sanchez, Rodriguez-Delgado, Garcia-Herrero, Cabo-Vaquera, Serous Macular Detachment as an Atypical Sign in Cat Scratch Disease, Archivos de la Sociedad Española de Oftalmología, Vol 81, 717-720, 2006

**Keywords:** Neuro-Ophth & Infectious Disease, Optic Neuropathy, Retina

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Poster 245

### Visual Outcomes in Giant Cell Arteritis in Patients with Polymyalgia Rheumatica on Prior Corticosteroids

Nailyn Rasool<sup>1</sup>, Philip Skidd<sup>2</sup>, Joseph F. Rizzo<sup>1</sup>

<sup>1</sup>Neuro-Ophthalmology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston, MA, USA, <sup>2</sup>Neuro-Ophthalmology, Fletcher Allen, Vermont College of Medicine, Burlington, VT, USA

#### Introduction:

Blindness caused by giant cell arteritis (GCA) is usually severe. Some patients with GCA have an earlier diagnosis of polymyalgia rheumatica (PMR), in which case they receive relatively low dose corticosteroids to alleviate discomfort.<sup>1</sup> We hypothesized that GCA patients who were receiving corticosteroids prior to visual loss would have less severe visual loss than patients who had not received corticosteroids in advance.

#### Methods:

A retrospective chart review was performed of all patients with GCA who presented to the Massachusetts Eye and Ear Infirmary between the years of 2005-2013 and who underwent detailed Neuro-Ophthalmic examination. Visual fields were performed by automated (i.e. Humphrey) technique, unless there was severe visual loss, in which case patients underwent Goldmann perimetry or confrontation testing. Visual fields were graded as being normal or showing partial or "diffuse" (i.e. defects in all four quadrants) visual loss. Presently, 34 charts of patients with visual loss and adequate clinical information have been reviewed. Eleven of the 34 patients had PMR and visual loss secondary to GCA. Five of these patients had been on oral steroids prior to developing visual loss.

#### Results:

Three of the five PMR patients who were on corticosteroids before developing visual loss had a visual acuity of 20/25 or better; whereas, 3 out of 29 patients not on prior corticosteroids had a visual acuity  $\geq 20/25$  ( $p = 0.006$ ). For visual field testing, two of the five PMR patients on corticosteroids before visual loss had diffuse field loss versus 16 of 29 patients not on corticosteroids ( $p < 0.001$ ).

#### Conclusions:

Patients with PMR who were on steroids prior to loss of vision from GCA had better visual acuity and visual field results in comparison to patients not on prior steroids. Our preliminary results demonstrate that even low dose corticosteroids may lessen the severity of blindness caused by GCA.

#### References:

1. Kermani TA & Warrington KJ. Polymyalgia rheumatica Lancet. 381(9860): 63-72. 2013

**Keywords:** Vascular Disorders, Optic Neuropathy, Giant Cell Arteritis, Arteritic Ischemic Optic Neuropathy, Polymyalgia Rheumatica

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

## Progressive Supranuclear Palsy (PSP) After Ascending Aorta Dissection Surgery

Hyosook Ahn<sup>4</sup>, Soolienah Rhiu<sup>1</sup>, Nam Ju Moon<sup>2</sup>, Ki-Han Kwon<sup>3</sup>

<sup>1</sup>Department of Ophthalmology, Hallym University College of Medicine, Dongtan Sacred Heart Hospital, Hwaseong, South Korea, <sup>2</sup>Department of Ophthalmology, Chung-Ang University College of Medicine, Seoul, South Korea, <sup>3</sup>Department of Neurology, Hallym University College of Medicine, Dongtan Sacred Heart Hospital, Hwaseong, South Korea, <sup>4</sup>Department of Ophthalmology, Asan Medical Center, University of Ulsan College of Medicine Seoul, South Korea

### Introduction:

Progressive supranuclear palsy (PSP) is a rare brain disorder that causes serious and progressive problems with control of gait and balance, along with complex eye movement and thinking problems. We present a case in which developed a neurologic syndrome including PSP after ascending aorta dissection surgery.

### Methods:

Observational case report of PSP after aortic surgery and brain ischemia.

### Results:

A 41-year-old man complained of blurred vision and difficulty in moving both eyes. Three months ago, under the diagnosis of type A aortic dissection, he received emergency ascending and total arch replacement surgery. During surgery, cardiac arrest occurred for 5 minutes and hypothermia treatment was applied following surgery. Neuro-ophthalmic examination revealed vision 20/80 in both eyes. He also showed horizontal and vertical supranuclear gaze palsy. Neurologic examination showed a wide based ataxic gait and dysphagia. Neuro-imaging revealed multiple old infarct in both basal ganglia and corpus callosum and old microhemorrhage in both cerebellum, parietal, right occipital and left frontoparietal white matter. Nerve conduction study showed no specific findings. CBC, CSF analysis, vasculitis panel, myositis antibody profile, ESR, CRP, HIV, B12, and thyroid studies were normal. Nine months later the patient still complained of gait difficulty, blurred vision, and supraduction limitation. Genetic tests and muscle biopsy results were normal.

### Conclusions:

This is an unusual case in which the neurologic syndrome cannot be explained as a postoperative cerebral syndrome. Neuronal damage may produce a syndrome of supranuclear gaze palsy, dysarthria, dysphagia and gait ataxia. Given to these findings we were able to recognize the possibility of PSP as a complication after aortic surgery. Wider recognition of this complication of cardiac surgery should lead to better understanding of its pathogenesis and appropriate management.

### References:

1. Mokri B, Ahlskog JE, Fulgham JR, Matsumoto JY. Syndrome resembling PSP after surgical repair of ascending aorta dissection or aneurysm. *Neurology*. 2004 Mar 23;62(6):971-3.
2. Bernat JL, Lukovits TG. Syndrome resembling PSP after surgical repair of ascending aorta dissection or aneurysm. *Neurology*. 2004 Sep 28;63(6):1141-2; author reply 1141-2.
3. Leigh RJ, Tomsak RL. Syndrome resembling PSP after surgical repair of ascending aorta dissection or aneurysm. *Neurology*. 2004 Sep 28;63(6):1141-2; author reply 1141-2.
4. Antonio-Santos A, Eggenberger ER. Asaccadia and ataxia after repair of ascending aortic aneurysm. *Semin Ophthalmol*. 2007 Jan-Mar;22(1):33-4.
5. Finsterer J. Mitochondrial ataxias. *Can J Neurol Sci*. 2009 Sep;36(5):543-53.
6. Habek M, Barun B, Adamec I, Mitrović Z, Ozretić D et al. Early-onset ataxia with progressive external ophthalmoplegia associated with POLG mutation: autosomal recessive mitochondrial ataxic syndrome or SANDO? *Neurologist*. 2012 Sep;18(5):287-9.

**Keywords:** Progressive Supranuclear Palsy (PSP), Ascending Aorta Dissection Surgery, Supranuclear Gaze Palsy, Ataxia, Dysphagia

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.



## Poster 247

### Delineation Of Natural History For Spinocerebellar Ataxia Type 7 (SCA-7) In Anticipation Of Rnai Therapy

Steven F. Stasheff<sup>1</sup>, Brett Jeffrey<sup>1</sup>, Wadih Zein<sup>1</sup>, Catherine Cukras<sup>1</sup>, Kenneth H. Fischbeck<sup>2</sup>, Mark Hallett<sup>2</sup>, Albert La Spada<sup>3</sup>, Brian P. Brooks<sup>1</sup>

<sup>1</sup>National Eye Institute/Ophthalmic Genetics & Visual Function Branch, NIH, Bethesda, MD, USA, <sup>2</sup>National Institute of Neurological Disorders & Stroke, NIH, Bethesda, MD, USA, <sup>3</sup>University of California-San Diego, La Jolla, CA, USA

#### Introduction:

Among hereditary spinocerebellar ataxias, type 7 (SCA-7) is notable for a prominent retinal degeneration that adds blindness to the severe neurologic disabilities suffered by those affected with this disorder. Preclinical testing of RNAi therapy for SCA-7 has encouraged movement of this therapy to clinical trials in the near future. Critical to accurate evaluation of the safety and efficacy of such treatments are clear delineation of the disease natural history and of reliable quantitative outcome measures. We report the development of a single-center longitudinal natural history study emphasizing detailed evaluation of the retinal and optic nerve phenotype, and results from initial evaluation of two siblings with SCA-7.

#### Methods:

25 patients with genetically confirmed SCA-7 are being recruited and will undergo detailed ophthalmologic and neurologic examination, Cambridge Color Testing (CCT), spectral domain optical coherence tomography (OCT), static &/or kinetic perimetry, microperimetry (MP-1), full-field electroretinography (ERG), evaluation of ataxia with both subjective and quantitative measures, video quantification of eye movement abnormalities, and brain MRI.

#### Results:

In a pilot evaluation, two siblings with moderately advanced SCA-7 demonstrated differences in severity of gait and limb ataxia, dysmetria, dysarthria, eye movement abnormalities, and pigmentary retinal degeneration. Striking were a distinctive retinotopic pattern of the retinal dystrophy and mildly anomalous optic nerve appearance, as well as distinctive features of CCT and ERG.

#### Conclusions:

Successful evaluation of potential treatments for SCA-7 will benefit from further refining our understanding of its natural history, as well as identifying those measures of retinal and optic nerve structure and function, and of eye movement control, that progress most rapidly and may be most amenable to being slowed or reversed. A particular advantage is the ability to directly observe the treated neural tissue and correlate any changes with functional ones, increasing the likelihood that lessons learned will also apply to other neurodegenerative and nucleotide repeat disorders.

**References:** None.

**Keywords:** Neuro-Ophth & Systemic Disease, Genetic Disease, Retina, Optic Neuropathy, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** None.

**Study Of The Dynamics Of Axonal Degeneration In Chemotherapy-Induced Neuropathy By In Vivo Corneal Confocal Microscopy.**

Ruben Torres-Torres<sup>1,2</sup>, Marisol Lopez-Moreno<sup>1,2</sup>, Montserrat Muñoz<sup>3</sup>, Pablo Villoslada<sup>1,2</sup>, Bernardo F Sanchez-Dalmau<sup>1,2</sup>

<sup>1</sup>*Institut Clinic d'Oftalmologia (ICOF), Hospital Clinic, Barcelona, Spain,* <sup>2</sup>*Center of Neuroimmunology. Institute of Biomedical Research August Pi Sunyer, Hospital Clinic, Barcelona, Spain,* <sup>3</sup>*Oncology Department. Hospital Clinic, Barcelona, Spain*

**Introduction:**

Peripheral neuropathy secondary to drugs is a cause of severe disability. Usually Invasive techniques as cutaneous nerve biopsy are employed for diagnosis. Corneal confocal microscopy (CCM) is a noninvasive and rapid technique to assess in vivo all structures of the cornea, including the sub-basal nerve plexus. We designed a prospective study to evaluate the usefulness of CCM for determinate peripheral neuropathy.

**Methods:**

Ten patients with a high probability of developing peripheral neuropathy secondary to drug treatment were evaluated. Specifically, patients who started treatment with taxanes, cisplatin and / or oxaliplatin. No patients with prior use of chemotherapy, previous corneal surgery or infection and /or contact lenses were included in the study. The patients were visited before treatment and two, four months, at the end of chemotherapy and 3 months after treatment. These images were subsequently analyzed by CCmetrics Software (Manchester University) analyzing the Nerve Fibre Density (CNFD), Nerve Branch Density (CNBD), Nerve Fibre Length (CNFL), Nerve Fibre Total Branch Density (CTBD), Nerve Fibre Area (CNFA) and Nerve Fibre Width (CNFW). Results were analyzed by means Generalized Estimated Equations (GEE) models with SPSS ver.20.

**Results:**

All patients showed statistically significant decrease in all measurements except in CNFW. After ending the treatment all patients showed improve in the last evaluation of the corneal sub-basal nerve plexus. This data correlates with the evolution of the clinical periphery neuropathy.

**Conclusions:**

Confocal microscopy appears to be a reliable method for assessing acute neuropathy in chemotherapy peripheral neuropathy safely and noninvasively. It is necessary to extend the number of patients to confirm these findings.

**References:**

1. Argyriou, A. a, Koltzenburg, M., Polychronopoulos, P., Papapetropoulos, S., & Kalofonos, H. P. (2008). Peripheral nerve damage associated with administration of taxanes in patients with cancer. *Critical Reviews in Oncology/Hematology*, 66(3), 218–28.
2. Cruzat, A., Pavan-Langston, D., & Hamrah, P. (2010). In vivo confocal microscopy of corneal nerves: analysis and clinical correlation. *Seminars in Ophthalmology*, 25(5-6), 171–7.
3. Dabbah, M. a, Graham, J., Petropoulos, I. N., Tavakoli, M., & Malik, R. a. (2011). Automatic analysis of diabetic peripheral neuropathy using multi-scale quantitative morphology of nerve fibres in corneal confocal microscopy imaging. *Medical Image Analysis*, 15(5), 738–747.
4. Ferrari, G., Gemignani, F., & Macaluso, C. (2010). Chemotherapy-associated peripheral sensory neuropathy assessed using in vivo corneal confocal microscopy. *Archives of Neurology*, 67(3), 364–5.

**Keywords:** Corneal Nerves, Axonal Degeneratio, Chemotherapy-Induced Neuropathy, Confocal Microscopy, Diagnostic Tests

**Financial Disclosures:** The authors had no disclosures.

**Grant Support:** Grant of the Catalanian Society of Ophthalmology 2013



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm)</b>		<i>continued</i>
23	Clinical Characteristics of Optic Neuritis Associated with Viral Infection	Sa Kang Kim
24	Methanol Causes Highly Selective Retinal Ganglion Cell Layer Loss and Inner Nuclear Layer Microcysts	Kendra A. Klein
25	Parsing the Differences Between LHON Affected: Genetic Vs Environmental Triggered Disease	Valerio Carelli
26	Changes in Macular OCT Retinal Sublayers of Patients and Carriers in the Natural History Phase of the Leber Hereditary Optic Neuropathy G11778A Gene Therapy Clinical Trial	Byron L. Lam
27	Low Grade Glioma of the Pituitary Stalk, Case Report with Review of the Literature	Su Ann Lim
28	Visualisation of Nerve Fibre Orientation in the Human Optic Chiasm Using Photomicrographic Image Analysis	Neeranjali Jain
29	Optic Neuropathy in Chronic Lymphocytic Leukemia	Amina I. Malik
30	How is Eye Fixation Affected by Optic Neuropathy? Diagnostic Value of Precise Recording of Retina Movement During an OCT Scan	Robert M. Mallery
31	A Novel OPA1 Mutation in Autosomal Dominant Optic Atrophy (ADOA)	Jordan A. Margo
32	The Vegetative Aspects of Neuroprotective Action of High Corticosteroid Doses in Compressive Traumatic Optic Neuropathies (TON)	Yulya T. Maslyak
33	Optic Atrophy in a Large Specialist Hospital	Joyce N. Mbekeani
34	Neurofibromatosis 1 with Large Suprasellar and Bilateral Optic Nerve Pilocytic Astrocytoma	Joyce N. Mbekeani
35	Optic Neuropathy in Wolfram's Syndrome Imaged with High-Definition Spectral Domain OCT	Dan Milea
36	Retinal Oximetry (Oxygen Saturation) and Peripapillary Vascular Diameters in Normal Eyes and in Non-Arteritic Ischemic Optic Neuropathy (NAION)	Ashwin Mohan
37	Is Ishihara Color Plate Testing as Reliable on iPod/iPhone and iPad as on Paper Format?	Katherine Boudreault
38	Multifactorial Optic Neuropathy - When it isn't Always Glaucoma	Deborah C. Parish
39	Cobalt-Chromium Metallosis with Normal ERG	Huy V. Nguyen
40	Nonarteritic Anterior Ischemic Optic Neuropathy (NAION): A Misnomer. A Non-Ischemic Papillopathy Caused by Vitreous Separation	Cameron F. Parsa
41	Visual and Oculomotor Outcomes in Children with Posterior Fossa Tumors	Crandall E. Peeler
42	Radiation Optic Neuropathy and Retinopathy from Low Dose (20Gy) Radiation Treatment	Crandall E. Peeler
43	Bilateral Optic Neuropathy in Superficial Intracranial Siderosis	Thong D. Pham
44	Binocular Acuity Summation (BAS) in Multiple Sclerosis (MS): Relation to Retinal Architecture and Visual System Neurophysiology	Sara S. Qureshi
45	Nutritional Optic Neuropathy after Bariatric Surgery	Yuna Rapoport
46	Lyme Disease Mimicking Giant Cell Arteritis	Nailyn Rasool
47	Structural Analyses of the Anterior Visual Pathway in Compressive Neuropathy	Won Hyung A. Ryu



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm)</b>		<i>continued</i>
48	Changes in Thickness of Retinal Segments in the Early Course of Non-Arteritic Ischaemic Optic Neuropathy	Bernardo F. Sanchez-Dalmau
49	A First Case Report of Chordoid Glioma Invading Optic Nerve	Nicolae Sanda
50	Clinical Spectrum of Optic Neuritis in Indian Children at a Tertiary Care Centre	Swati Phuljhele
51	Prognosticating Factors in Nonarteritic Ischaemic Optic Neuropathy	Swati Phuljhele
52	Sweep My Blindness Away!	Salwa Abdel Aziz
53	Progressive Non-Arteritic Optic Neuropathy as the First Presentation of Anti-Phospholipid Antibody Syndrome	Tarek A. Shazly
54	Anterior Visual Pathway(AVP) Meningiomas: A Dosimetric Comparison of IMRT to Pencil-beam Proton Therapy	Scott L. Stafford
55	A Case of Lyme Neuroretinitis- An Elusive Entity	Padmaja Sudhakar
56	50% of Non-Arteritic Anterior Ischemic Optic Neuropathy Occurs Between 40-55 Years Old	Ming-Hui Sun
57	Evaluation of Retinal Nerve Fiber Layer and Ganglion Cell Complex in a Cohort of Chinese with Optic Neuritis or Neuromyelitis Optica Spectrum Disorders Using SD-OCT	Guohong Tian
58	Transient Monocular Vision Loss Upon Awakening: A Benign Phenomenon	Marc A. Bouffard
59	Autosomal Dominant Optic Atrophy In Singapore: Multiethnic Involvement And Report Of A New Gene Mutation Causing Optic Atrophy And Deafness	Sharon L. Tow
60	Optic Nerve Head and Macular Choroidal Vascularization in Patients Affected by Normal Tension Glaucoma and Non-Arteritic Ischemic Optic Neuropathy: What Do They Share? What Is Different?	Giacinto Triolo
61	Multifocal ERG Shows Pre-Ganglion Cells Dysfunction in Dominant Optic Atrophy: Genotype-Phenotype Correlation	Maria Lucia Cascavilla
62	Pediatric Primary Optic Nerve Sheath Meningioma	Kavin Vanikieti
63	Syphilitic Optic Neuropathy : Reemerging Cases Over A 2-Year Period	Anuchit Poonyathalang
64	Anterior Ischemic Optic Neuropathy as Sole Presenting Sign of Internal Carotid Artery Occlusion	Kimberly M. Wings
65	Challenges in the Management of Apoplexy of a Growth Hormone(GH)-Secreting Pituitary Adenoma During Pregnancy	John Wong
66	The Prevalence of Mitochondrial Disease in the Adult Population – Implications for the Prevention of Maternal Transmission	Patrick Yu-Wai-Man
67	Neuropathies in Children Involving Vision Caused by a Fall From Height	Alon Zahavi
68	Immunosuppressive Therapy of Chinese Isolated Non-MS Idiopathic Optic Neuritis	XiaoJun Zhang
69	How does Exogenous ROS Effect LHON Conversion?	Jeffrey Tran
<b>Category: Cranial Nerves (Paresis, etc.)</b>		
70	Chronic, Painful Abducens Palsy May Require Angiography: Lesson Re-Learned from a Case of a White-Eyed Shunt.	Aliaa H. Abdelhakim
71	Eight and a Half Syndrome-A Rare Neuro Ophthalmologic Manifestation Due to Pontine Infarction	Sanam Anwer



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Cranial Nerves (Paresis, etc.)</b>		<i>continued</i>
72	Where the Lung Meets the Eye	Nathan W. Blessing
73	Metastasis or Metamorphosis?	Krista Kinard
74	Acute Onset Internuclear Ophthalmoplegia and Migraine Headache	Oana M. Dumitrascu
75	Traumatic Avulsion of the Oculomotor Nerve: First Definitive Documentation on High Resolution MRI	Lauren C. Ditta
76	Ocular Motor Cranial Nerve Palsies in Pituitary Apoplexy	Rabih Hage
77	Later Life Decompensation of Congenital Trochlear Palsy due to Agenesis	Ji-Soo Kim
78	Presentation of Primary Hodgkin's Lymphoma as Multiple Cranial Neuropathies	Hreem N. Patel
79	Acute Angle Closure Glaucoma in a Patient with Miller Fisher Syndrome without Pupillary Dysfunction	Won Yeol Ryu
80	Congenital Oculomotor Nerve Paresis with Isolated Cyclic Pupillary Spasms	Michael Salman
81	Abducent Nerve Palsy in a Patient with Castleman Disease!	Islam Zaydan
82	Nivolumab Plus Ipilimumab Induced Aseptic Meningitis and Bilateral Sixth Nerve Palsy in Metastatic Melanoma Treatment	Mitchell Strominger
83	Combined Third Nerve Palsy and Vertical Gaze Palsy in Midbrain Thalamic Stroke	Kara Fister
84	A Case of Abducens Nerve Palsy Followed by Cyclic Esotropia: A Case Report and Review of the Literature	Takako Sugimoto
85	Functional MRI and MRI Tractography in Progressive Supranuclear Palsy-Like Syndrome	Michael Vaphiades
86	Abducens Palsy in a Patient with Pityriasis Rubra Pilaris	Amanda L. Way
<b>Category: CSF (Intracranial Hypertension, Intracranial Hypotension, etc.)</b>		
87	Characterization of Idiopathic Intracranial Hypertension (IIH) in Qatar	Mais N. Alkawaz
88	Osmometry of Cerebrospinal Fluid from Patients with Idiopathic Intracranial Hypertension (IIH)	Steffen E. Hamann
89	Corneal Biomechanics : A Journey Through Uncharted Territories	Ashwin Mohan
90	Clinical and Prognostic Significance of CSF Closing Pressure in Pediatric Pseudotumor Cerebri Syndrome	Shannon J. Beres
91	Subretinal Fluid in Idiopathic Intracranial Hypertension: Clinical Course and Outcome	John J. Chen
92	Overdiagnosis of Idiopathic Intracranial Hypertension (IIH)	Adeniyi Fisayo
93	An Update on Terson Syndrome: Prevalence and Prognosis	Philip S. Garza
94	Aberrant Presentation of Pseudotumor Cerebri	Sawyer B. Hall
95	Jugular Vein Thrombosis as a Cause of Intracranial Hypertension	Josepha Horowitz
96	Autism Spectrum Disorder in Pediatric Pseudotumor Cerebri Syndrome	Anne K. Jensen
97	Lumbar Drain in Fulminant Intracranial Hypertension	Mays A. El-dairi
98	Papilledema with Unilateral Occlusion of Internal Jugular Vein	Abhishek Thandra
99	Papilledema Secondary to Internal Jugular Vein Thrombosis in a Dialysis Patient	Shauna W. Berry
100	Ultramicroscopic Study of the Optic Nerve Sheath in Patients with Severe Vision Loss from Idiopathic Intracranial Hypertension- Methodology	Joshua W. Evans



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: CSF (Intracranial Hypertension, Intracranial Hypotension, etc.)</b>		<i>continued</i>
101	Ultramicroscopic Study of the Optic Nerve Sheath in Patients with Severe Vision Loss from Idiopathic Intracranial Hypertension- Results	Marla Davis
102	Exploratory Study of the Relationship Between the Levonorgestrel-Releasing Intrauterine System and Idiopathic Intracranial Hypertension	Reuben M. Valenzuela
103	Coning Following Lumbar Puncture in a Patient with Idiopathic Intracranial Hypertension	Christian J. Lueck
104	Long Term Follow-Up of PTC: Pre-Pubertal Children vs. Adolescents and Adults	Assaf Hilely
105	Hemodialysis Graft-Induced Intracranial Hypertension	Devin D. Mackay
106	Incidence of Idiopathic Intracranial Hypertension (IIH) Among Users of Tetracycline Antibiotics	Samuel F. Passi
107	CSF Characteristics in Patients with High Frisén Papilledema Grades from the IIHTT	John H. Pula
108	Visual Outcomes After Treatment of Venous Sinus Stenosis with Dural Venous Sinus Stenting	Ahmara G. Ross
109	Relationship between High Opening Pressure on Lumbar Puncture and Failure of Optic Nerve Sheath Decompression to Prevent Progressive Visual Loss in Patients with Idiopathic Intracranial Hypertension	Mark E. Robinson
110	Venous Sinus Stenting for Treatment of Increased Intracranial Pressure Secondary to Venous Sinus Stenosis	Nikisha Richards
111	Pediatric Primary Pseudotumor Cerebri Syndrome (PTCS): A Detailed, Retrospective, Multicenter Analysis of Anthropometrics	Claire A. Sheldon
112	Endovascular Venous Stenting in Treatment of Primary Idiopathic Intracranial Hypertension	Rajeev Sivasankar
113	Idiopathic Intracranial Hypertension Mimicking Foster Kennedy Syndrome	Chuan-Bin Sun
114	Going for the Jugular! Paragangliomas, Papilledema, and Vision Loss	Jonathan Trobe
115	Visual Outcomes Following Optic Nerve Sheath Fenestration	Neel S. Vaidya
116	New Side Effect of Acetazolamide: Palinopsia	Peggy H. Vogt
117	Endovascular Intervention in a Chronic Case of Idiopathic Intracranial Hypertension (IIH)	Muhammad-Atif Zubairi
<b>Category: Disorders of Vision Processing</b>		
118	Optical Aberrations- A Trigger Factor for Migraine?	Rohit Shetty
119	An Interesting Field of Study	Andrew S. Camp
120	Visual Field Assessment in the "Split-Brain" Patient: Effect of the Method/Laterality of Obtaining Patient Responses on Apparent Visual Field Defect Size	J. Alexander Fraser
121	Binasal Hemianopsia	Erica Ballard
122	Voice Processing in Developmental Prosopagnosia	Ran R. Liu
123	Building a Repository for Posterior Cortical Atrophy	Jennifer J. Olds
124	Subretinal Choroidal Neovascularization Associated with Unilateral Papilledema and IIH: Case Report	Jalpa M. Patel
125	The Initiation of Smooth Pursuit Eye Movements in Anisometropic Amblyopia	Rana A. Raashid
126	The Pulfrich Phenomenon: An Objective Signature-Biomarker of MS Pathophysiology	Millad J. Sobhanian
127	Distractibility in Multiple Sclerosis: A Potential Cause of Morbidity	Derek Sears



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Disorders of Vision Processing</b>		<i>continued</i>
128	Disconjugacy of eye alignment is greater with near fixation during binocular viewing in amblyopia	Vivian Xu
<b>Category: Misc. Motility Disorders (Nystagmus, etc.)</b>		
129	An Adjustable Magnetic Prism Carrier for Strabismus Evaluation	David S. Bardenstein
130	Patient Satisfaction with Prismatic Correction of Diplopia	Shauna E. Berry
131	Duane Retraction Syndrome in Duchenne Muscular Dystrophy	Thomas M. Bosley
132	Peribulbar Botulinum Toxin as a Treatment for Symptomatic Opsoclonus in Oculopalatal Myoclonus	Katherine Duncan
133	Thalamic Stroke in a Young Patient Presenting with Sudden Onset Large Skew Deviation	Oana M. Dumitrascu
134	Ophthalmological Spectrum of Locked-In Syndrome	Martin Graber
135	Cigarette Smoking and Activities of Daily Living in Ocular Myasthenia Gravis	Sean M. Gratton
136	An Evaluation of Educational Neurological Eye Movement Disorder Videos Posted on Internet Video Sharing Sites	Simon J. Hickman
137	How Do Patients with Strabismus Locate Visual Targets?	Jonathan C. Horton
138	Superior Cerebellar Peduncle Demyelination Causing Geotropic Central Positional Nystagmus	Cristina Duque
139	Normative Database for the King-Devick Test in Adults and Adolescents	Danielle Leong
140	Marcus Gunn Jaw Winking with Electronegative Cone-Rod Dystrophy: Case Report	Su Ann Lim
141	Levodopa-Induced Ocular Dyskinesias	Bryan V. Pham
142	Clinical Features, Diagnostic Findings and Treatment of Adult-Onset Opsoclonus-Myoclonus Syndrome: A Case Series	Olga Rosenvald
143	Divergence Palsy Due to Antiepileptic Drugs	Marc A. Bouffard
144	Quantifying the Vestibulo-ocular Reflex with Video-Oculography: Nature and Frequency of Artifacts	Ali S. Saber Tehrani
<b>Category: Miscellaneous</b>		
145	Neuroophthalmic Manifestations of Intracranial Tumours in South India	Kowsalya Akkayasamy
146	Sideline Testing in Youth and Collegiate Athletes: What Does Vision Add to the Concussion Puzzle?	Laura J. Balcer
147	Assessment of Recruitment Patterns in a Neuro-Ophthalmology Registry	Kimberly D. Blankshain
148	Prediction of Eye Position During General Anesthesia Using Bispectral Index Monitoring	Seung Ah Chung
149	Demographic Profile of the Patients Presenting to a Neuro-Ophthalmology Clinic of a Tertiary Eye Care Centre	Shiva Prasad Gantala
150	Botulinum Toxin-Augmented Strabismus Surgery versus Conventional Surgery in the Treatment of Large-Angle Infantile Esotropia	Aubrey L. Gilbert
151	Heidenhain Variant of Creutzfeldt Jakob's Disease (CJD), In a Patient Who had Bovine Bioprosthetic Valve Implantation	Haneen Jabaly-Habib



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Miscellaneous</b>		<i>continued</i>
152	The Effects of Pediatric Primary Brain Tumors on Vision and Quality of Life	Supharat Jariyakosol
153	A Method for Quantifying off Chart Visual Acuities	Rustum Karanjia
154	Differential Functions Mediated by Melanopsin Assessed in Subjects with Healthy and Diseased Eyes	Aki Kawasaki
155	Familial Papillitis and Macular Cystoid Edema: A Genetic Autoimmune Disorder?	Chiara La Morgia
156	New Daily Persistent Headache Triggered by Cataract Extraction?	Julia E. Mallory
157	Ten Years of Temporal Artery Biopsies in Ontario, Canada: A Population-Based Study on Practice Patterns and the Incidence of Giant Cell Arteritis	Jonathan A. Micieli
158	A Method for Recognizing Colorblind Malingering	Andrew E. Pouw
159	Mycotic Aneurysms of Intracavernous Internal Carotid Artery	Kumudini Sharma
160	Platelet-Mediated Microvascular Ischemia Mimicking Migraine in Patients with Thrombocythemia	Melissa C. Tien
<b>Category: Nerve Fiber Layer and Retinal Testing (OCT, ERG, etc.)</b>		
161	Ganglion Cell Damage and Functional Recovery after Optic Neuritis	Alexander U. Brandt
162	To Evaluate the Retinal Nerve Fibre Layer Loss in Optic Neuritis Using Spectral Domain Optical Coherence Tomography	Kowsalya Akkayasamy
163	Retinal Ganglion Cell Injury in Early Pediatric Onset MS	Jennifer Graves
164	The Correlation of Critical Flicker Fusion Function and P100 Latency of Visual Evoked Potential with Luminance	Yanjun Chen
165	The Effects of Amblyopia on Visual Evoked Potentials	Florin Grigorian
166	Doubling Method for Papilledema and Pseudopapilledema	Bokkwan Jun
167	Correlation Between Structural and Functional Changes in Retina in Parkinson's Disease	Manpreet Kaur
168	Relationship Between Visual Acuity and Retinal Nerve Fiber Layer Thickness Measured by Spectral Domain Optical Coherence Tomography in Patients with Optic Neuropathy	Sungeun Kyung
169	Ganglion Cell Layer Thinning Detected by Optical Coherence Tomography as a Sign of Early Optic Atrophy in Pediatric Papilledema	Andrew R. Lee
170	Peripapillary Retinal Nerve Fiber Layer Thickness Corresponds to Drusen Location and Extent of Visual Field Defects in Patients with Optic Disc Drusen	Lasse Malmqvist
171	Progressive Retinal Structure Abnormalities in Multiple System Atrophy	Carlos E. Mendoza-Santiesteban
172	OCT Shows Consistent Relationships Between Macular Layers and Peripapillary RNFL in Chronic Demyelinating and Compressive Optic Neuropathies	Mark J. Morrow
173	OCT Shows Different Relationships Between Macular Layers and Peripapillary RNFL in Acute Versus Chronic Papillitis and Papilledema	Fawzi Abukhalil
174	The Photopic Negative Response in Idiopathic Intracranial Hypertension	Heather E. Moss
175	Attenuated Hunter Syndrome: A Rare Cause of Simultaneous Retinal and Optic Nerve Disease	Kannan Narayana
176	Ganglion Cell and Retinal Nerve Fiber Layer Analysis in a Case of PION	Joshua Pasol





# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Nerve Fiber Layer and Retinal Testing (OCT, ERG, etc.)</b>		<i>continued</i>
177	Perimacular Ganglion Cell Complex Thinning Detected By Spectral-Domain OCT Useful In Detecting Optic Tract Syndrome	Fannie Petit
178	To Evaluate Changes in Retinal Nerve Fiber Layer and Ganglion Cell Layer on Cirrus HD-OCT in Cases of Multiple Sclerosis (with and without Optic Neuritis) and Optic Neuritis	Ganesh Pillay
179	Examination of Visual Evoked Potential (VEP) in a Pediatric Population with Newly Diagnosed Elevated Intracranial Hypertension	Ahmara G. Ross
180	Retinal Nerve Fiber Layer Thickness in a Population-Based Study of Elderly Subjects: The Alienor Study	Marie B. Rougier
181	OCT-Derived Retinal Capillary Density is Decreased in Corresponding Areas of Retinal Neuron Loss in Optic Neuropathy	Min Wang
182	Adaptive Optics Imaging with Histopathologic Correlation in Cancer-Associated Retinopathy	Zoë R. Williams
183	Robust Optic Nerve Head Analysis Based on 3D Optical Coherence Tomography	Ella M. Kadas
<b>Category: Neuro-Imaging (MRI, CT, etc.)</b>		
184	Ocular Motility Defects with Concordance Neuroimaging	Nagham Al-Zubidi
185	Mural Enhancement of the Intracranial Internal Carotid Artery in Giant Cell Arteritis	Sidney M. Gospe III
186	Morning Glory Disk Anomaly Associated with Absence of Intracranial Internal Carotid Artery	Adriana P. Grigorian
187	Measurement of Optic Nerve Sheath Diameter by CT, MRI and Ultrasound	Klara Landau
188	Occipital Partial Status Epilepticus with Abnormal MRI Imaging	Sonalee Kulkarni
189	Neuro-Imaging Characteristics of Common Extraocular Prosthetic Devices	Samantha F. Lu
190	A Lion in the Bush	Padmaja Sudhakar
<b>Category: Orbital and Eyelid Disorders</b>		
191	Why the Delay in Diagnosis? Increased Time From Symptom Onset to Diagnosis in Blepharospasm: A Prospective, Clinic-Based Study	Kristen E. Dunbar
192	Atypical Presentation of Orbital Lymphangioma	Haydée S. Martinez
193	Prospective Assessment of Peri-Oral Weakness Following Peri-Orbital Botulinum Toxin for Blepharospasm	Alexander J. Hartmann
194	Vertical Diplopia and Ptosis from Removal of the Orbital Roof in Pterional Craniotomy	Jonathan C. Horton
195	Treatment of rivaroxaban associated orbital hemorrhage	Marilyn C. Kay
196	Stonewalled: Bilateral Sequential Vision Loss In A Peritoneal Dialysis Patient Category: Systemic Disease	Angelina Espino Barros Palau
197	Allergic Fungal Sinusitis Mimicking Thyroid Orbitopathy	Lina Nagia
198	Optic Nerve Sheath Meningioma. No Longer Impossible but Still Difficult	T. Ben Ableman
199	An Advanced Pancoast Tumor Masquerading as Congenital Hereditary Ptosis	Deborah C. Parish
200	It's All Going Dark!	Tarek A. Shazly
201	Bartonella Henselae-Associated Neuroretinitis with Orbital Intraconal and Optic Nerve Sheath Involvement	Ann Shue
202	Impact of Injection Site on Dose Escalation in the Treatment of Blepharospasm	Rachel G. Simpson



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Orbital and Eyelid Disorders</b>		<i>continued</i>
203	A Case of Nasopharyngeal Carcinoma Masquerading as Primary Orbital Tumour	Shantha Amrith
204	Visual Field and Graves Ophthalmopathy	Sabine Defoort
<b>Category: Posterior Afferent Visual Pathway (Post-Chiasmal)</b>		
205	VEPs to Lateralized Stimuli to Measure the Interhemispheric Transfer Time (IHTT)	Ilie P. Cretu
206	The Heidenhain Variant of Creutzfeldt-Jakob Disease	Kristen E. Dunbar
207	Six-Year Follow-Up of the Progression of Cortical Vision Loss in a Patient with HIV-Related Progressive Multifocal Leukoencephalopathy	Bahareh Hassanzadeh
208	Retrograde Degeneration of Retinal Ganglion Cells in Homonymous Hemianopia	Angela M. Herro
209	Lesions of the Optic Tract: A Review of 35 Cases	Kristopher M. Kowal
210	Hydrocephalus and More!	Islam Zaydan
<b>Category: Pupil</b>		
211	Ipratropium-Induced Mydriasis: A Possible Exception to the Current Anisocoria Diagnostic Pathway	Erica L. Archer
212	Automated Pupillograph as a Screening Tool in Ophthalmology Clinic	Ashwin Mohan
213	The Yield of Diagnostic Imaging in Patients with Isolated Horner's Syndrome	Johanna D. Beebe
214	Evaluation of Pupil Response as Ocular Marker for Pre-Clinical Alzheimer's Disease	Ling Bei
215	A Case of Horner's Syndrome After Clipping of the Internal Carotid-Posterior Cerebral Artery Aneurysm Associated with Subarachnoid Hemorrhage	Tamura Koichiro
216	Test-Retest Reliability of Hemifield, Central-Field and Full-Field Chromatic Pupillometry for Assessing the Function of Melanopsin-Containing Retinal Ganglion Cells	Shaobo Lei
217	Unique Presentation of Anti-GQ1b Antibody Syndrome	Tin Yan A. Liu
<b>Category: Retina</b>		
218	Novel Retinal Observations in Genetically Confirmed Kearns Sayre Syndrome	Thomas M. Bosley
219	Autoimmune Retinopathy and Optic Neuropathy in a patient with Anti-GAD and Other Retinal Antibodies	Sumayya J. Almarzouqi
220	GM2-gangliosidosis, AB variant: An Elusive Cause of Neurodegenerative Cherry Red Spots	Michael C. Brodsky
221	Malignant Optic Nerve Glioma Manifesting as Central Retinal Vein Occlusion and Papilledema	Ryan C. Burton
222	Comprehensive Postmarketing Review of Visual Defects Reported with Topiramate	Lisa Ford
223	Sequential Fundus Findings After Platelet Rich Plasma for Facial Rejuvenation	Emely Z. Karam
224	Contrast Sensitivity Visual Acuity is Degraded in REM Sleep Behavior Disorder	Matthew J. Khayata
225	Ophthalmoscopy Vs. MRI – Sometimes Less is More	Anton Kolomeyer
226	Retinal Vessel Oximetry and Vessel Diameter Measurements: A Novel Metabolic Marker of Multiple Sclerosis	Marlen Lucero
227	Acute Zonal Occult Outer Retinopathy Presenting as Retrobulbar Optic Neuritis	Sungeun E. Kyung
228	Long-term Follow-up of Laser Pointer Induced Macularopathy	Chuan-Bin Sun



# North American Neuro-Ophthalmology Society

## 41<sup>st</sup> Annual Meeting

February 21-26, 2015

Hotel del Coronado • San Diego, California

Poster #		Presenting Author
<b>Category: Retina</b>		<i>continued</i>
229	Use of Wide-field Retinography for Periphlebitis Detection in Patients with Multiple Sclerosis	Ruben Torres-Torres
230	An Atypical Presentation of Giant Cell Arteritis	Laurel N. Vuong
<b>Category: Systemic Disease</b>		
231	Discordance Rates in Healing/Healed Arteritis in Temporal Artery Biopsies: The Role for Bilateral Biopsies	Sangsu Han
232	Bilateral Intracranial Optic Nerve and Chiasmal Involvement in IgG-4 Related Disease	Raed Behbehani
233	Neuromyelitis Optica Preceded by Seizure Disorder	Iris Ben Bassat Mizrahi
234	Optic Nerve Meningioma Masquerading as Neurosarcoidosis	Ryan C. Burton
235	Comparison of the Clinical Characteristics of Patients with Active Arteritis and Healed Arteritis, the Two Histopathological Patterns Considered Positive in Giant Cell Arteritis	Michael W. Salter
236	Autologous Hematopoietic Stem Cell Transplantation in Neuromyelitis Optica - An Update	Fiona Costello
237	Social Media and Susac Syndrome	Robert A. Egan
238	Horner's Syndrome as Initial Manifestation of a Malignant Peripheral Nerve Sheath Tumor	Gerard L. Hershewé
239	Birefringence of Retinal Nerve Fiber Layer and Retinal Blood Flow Velocity in Multiple Sclerosis	Hong Jiang
240	Botox for Chronic intractable Headache among Veterans at VA Medical Center, Long Beach, California	Grace W. Kao
241	Thin-Film Optical Notch Filter Spectacle Coatings for the Treatment of Migraine and Photophobia	Bradley J. Katz
242	Monitoring Alzheimer's Disease with Retina Examination	Umur A. Kayabasi
243	Why is He Losing His Sight? He is Losing His Photoreceptor!	Ainat Klein
244	Extensive Macular Serous Retinal Detachment as the Initial Presentation of Bartonella Henselae Induced Neuroretinitis	Darrell Lewis
245	Visual Outcomes in Giant Cell Arteritis in Patients with Polymyalgia Rheumatica on Prior Corticosteroids	Nailyn Rasool
246	Progressive Supranuclear Palsy (PSP) After Ascending Aorta Dissection Surgery	Hyosook Ahn
247	Delineation of Natural History for Spinocerebellar Ataxia Type 7 (SCA-7) in Anticipation of RNAi Therapy	Steven F. Stasheff
248	Study of the Dynamics of Axonal Degeneration in Chemotherapy-Induced Neuropathy by in Vivo Corneal Confocal Microscopy	Ruben Torres-Torres