



North American Neuro-Ophthalmology Society

38th Annual Meeting

February 11–16, 2012

JW Marriott San Antonio Hill Country Resort and Spa • San Antonio, Texas

POSTER PRESENTATIONS

Tuesday, February 14, 2012 • 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.

The Tour of Posters has been replaced with Distinguished Posters. The top-rated posters this year are marked with a “” in the Syllabus and will have a ribbon on their poster board.*

Poster #	Title	Presenting Author
1*	Relation of Ouabain-Like Compounds and Intracranial Pressure in Idiopathic Intracranial Hypertension	Raed Behbehani
2*	Correlation between multifocal pattern electroretinogram and Fourier-domain OCT in eyes with temporal hemianopia from chiasmal compression	Mário Monteiro
3*	Development of Pupillary Adrenergic Supersensitivity after Pharmacologic Induction of Oculosympathetic Defect	Christopher Kirkpatrick
4*	Sleepiness and circadian timing have a differential effect on pupil responses mediated by inner retinal versus outer retinal photoreception	Aki Kawasaki
5*	Comparison of Cerebrospinal Fluid Opening Pressure in the Prone and Lateral Decubitus Positions	Anne Abel
6*	The King-Devick (K-D) Test of Rapid Eye Movements: A Bedside Correlate of Disability and Quality of Life in Multiple Sclerosis	Stephen Moster
7*	Why Mothers Say ‘Look At Me When I’m Talking To You’: The Effect of Amblyopia on Visual-Auditory Speech Perception	Rajen Desai
8*	Hypopigmented Mice as an Animal Model for Infantile Nystagmus Syndrome	Ghislaine Traber
9*	Longitudinal Study of Retinal Ganglion Cell Layer Neuronal Loss by Spectral-Domain Optical Coherence Tomography (SD-OCT) in Multiple Sclerosis	Emma Davies
10*	Saccadic adaptation in amblyopia	Rana Arham Raashid
11*	Optic nerve and melanopsin retinal ganglion cells involvement in relation to circadian dysfunction in Alzheimer disease	Chiara La Morgia
12*	The effect of strabismic amblyopia and strabismus without amblyopia on saccade latencies	Ewa Niechwiej-Szwedo
13*	Giant Cell Arteritis: Treatment of Burkholderia Pseudomallei Infection with Antibiotics	Bonnie Keung
14*	The Role of Vitamin D Status in Optic Neuritis	Fiona Costello
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16	Cranial Neuropathy and Ectatic Vessels: A Close Up of Vascular Loops	Bonnie Keung
17	Relationship between fixation stability and saccade initiation in patients with amblyopia	Ewa Niechwiej-Szedo
18	MRI Findings in Idiopathic Intracranial Hypertension (IIH) Compared with Cerebral Venous Thrombosis (CVT)	Maysa Ridha
19	Does Bilateral Transverse Cerebral Venous Sinus Stenosis (TSS) Really Exist in Patients without Increased Intracranial Pressure (ICP)?	Linda P. Kelly
20	Obstructive Sleep Apnea In Idiopathic Intracranial Hypertension: Comparison With Matched Population Data	Matthew Thurtell
21	Emergency and Inpatient Services Utilization in Idiopathic Intracranial Hypertension	Jagger Koerner



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24	A Retrospective Study Of Idiopathic Intracranial Hypertension and Metabolic Syndrome: Do They Co-Exist?	Rudrani Banik
25	JAK2V617F Mutation Associated with "Pseudotumor Cerebri"	Iris Ben-Bassat Mizrachi
26	Correlation Between Visual Parameters And Neuroimaging Features Of Papilledema	Leena Padhye
27	Evaluation of Optic Nerve Head Elevation in Idiopathic Intracranial Hypertension with 3-Dimensional Optical Coherence Tomography	Ilana Traynis
28	Identifying Trends In The Treatment And Outcomes Of Patients With Idiopathic Intracranial Hypertension (IIH)	Cindy Tsai
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77	Increasing interval between injections after prolonged onabotulinum toxin type A use for blepharospasm	Lindsey DeLott
78	Bilateral abducens nerve palsy as a manifestation of Epstein Barr virus (EBV) encephalitis in children	Jin Choi
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103	Unexplained Hydrocephalus in a Patient with Neuromyelitis Optica	Sean Gratton
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107	Diagnostic Uncertainty Due to Optic Disc Drusen	Michael Vaphiades
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109	Acute Zonal Occult Outer Retinopathy Mimicking Retrobulbar Optic Neuritis	JaeHo Jung
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111	Cross sectional survey of visual acuity and self-reported visual function in patients with amyotrophic lateral sclerosis	Heather E. Moss
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116	Driver Fitness in patients with Glaucoma and Cognitive Impairment	Peter N Rosen
117	Development of Clinical Markers to Avoid Unnecessary Temporal Artery Biopsy in Giant Cell Arteritis	Katherine Fallano
118	The Optic Nerve Head Component of the Multifocal Electroretinogram: A Novel Metric for Dissecting Pathophysiologic Mechanisms in MS	Teresa Frohman
119	Bilateral Retinal Artery Occlusion associated with Posterior Reversible Encephalopathy Syndrome	Sun-Young Oh
120	White Dots and Cerebral Spots: APMPE and Cerebral Vasculitis	Robert Mallery
121	The Efficacy of Toothpaste refraction in Children with Functional Visual Loss and its prognosis	Yeon-Hee Lee
122	Treatment of Functional Vision Loss with Duloxetine	Robert Egan
123	Bilateral endogenous endophthalmitis associated with meningitis in a healthy woman.	Minsoo Lee
124	Parry-Romberg Syndrome: Presentation of Two Patients; Review of Neuro-Ophthalmologic Manifestations	Warren Felton
125	Appearance of cavernous angiomas after brain radiation therapy.	Yanny Phillips
126	New Onset Leber's Hereditary Optic Neuropathy in a 79 year-old male	Marc Malouf
127	Amaurosis fugax and stroke in a boy due to supernumary rib in the neck	Kumudini Sharma
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129	Electrophysiological Testing to Diagnose Sudden Bilateral Blindness from Cryptococcal Meningitis	Eli Moses
130	Occult Giant Cell Arteritis: Overcoming Hurdles to Diagnosis.	Mitchel Williams
131	Creutzfeldt-Jakob disease with cortical blindness	Guohong Tian
132	Web-based Neuro-Ophthalmic Patient Education	Sashank Prasad
133	Development of Smartphone Technology to Monitor Disease Progression in Multiple Sclerosis	Sashank Prasad
134	Rodent Model of Perioperative Ischemic Optic Neuropathy	Steven Roth
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137	A slide is worth a thousand scans: Diplopia with a cavernous sinus mass lesion	Gregory Van Stavern
138	Acute vision loss in intracranial hypertension secondary to choroidal neovascularization.	Laurel N. Vuong
139	Congenital Mydriasis Associated with Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS)	Collin McClelland
140	Curved fingers and tight eyes	Sui H. Wong
141	Oculomotor palsy and contralateral hemianesthesia & hemiparesis secondary to rhombencephalitis presumably due to CNS infection with Listeria monocytogenes	Sarkis Nazarian
142	Orbital Inflammation as a Paraneoplastic Manifestation of non-CNS Lymphoma	Hangxiu Xu
143	Progressive, Painful Vision Loss in a Young Female with Systemic Sarcoid due to Meningothelial Meningioma	Michelle Liang



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145	Eosinophilic Granuloma: Evolution of Diagnosis and Treatment	Ryan Whitted
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147	Leber Hereditary Optic Neuropathy (LHON): a novel mutation?	Mariana de Virgiliis
148	Neurocysticercosis and subsequent Multiple Sclerosis	Haydée Susana Martinez

Posters 54 and 86 have been withdrawn.

Poster 1*

Relation of Ouabain-Like Compounds and Intracranial Pressure in Idiopathic Intracranial Hypertension

Raed Behbehani¹, Mark K. Borsody², Tamara N. Heaton², Chisa Yamada², Asser Mabrook¹, Beau B. Bruce³, Ehud Reich⁴, Hadas Stiebel-Kalish⁴, Katherine Carroll⁵, Lyla Kamsheh⁵, Frans Leenen⁶, Roselyn White⁶

¹*Ibn Sina Hospital, Neuroophthalmology Service, Kuwait, Kuwait*, ²*Northern Neurosciences, Inc, San Francisco, CA, USA*, ³*Emory Eye Center, Atlanta, GA, USA*, ⁴*Neuro-Ophthalmology Unit, Rabin Medical Center, Petah Tikva, Israel*, ⁵*Department of Neurology, Northwestern University, Chicago, IL, USA*, ⁶*University of Ottawa Heart Institute, Ottawa, Ontario, Canada*

Introduction:

Oubain-like compounds (OLC), present in serum and cerebrospinal fluid (CSF), are endogenous inhibitors of the Na-K ATPase pump, which is the rate limiting step in CSF formation. We ought to investigate if IIH patients have lower levels of OLC, which can potentially increase the CSF production and caused high intracranial pressure.

Methods:

In this multicenter cross-sectional study we prospectively recruited 31 subjects who met the diagnostic criteria for IIH. CSF was obtained via lumbar puncture with measurement of the opening pressure. CSF samples were analyzed for OLC by means of ELISA. The CSF OLC levels for our 31 IIH subjects was compared to that of 31 historical controls from a previous pilot study using a Wilcoxon rank test and linear regression was done to determine if there any correlation between CSF OLC and CSF opening pressure.

Results:

The mean CSF OLC for IIH patients, 0.0530 ng/mL, was significantly lower than the mean CSF OLC for controls, 0.1155 ng/mL ($P < 0.001$). Moreover, there was negative correlation between CSF OLC and CSF opening pressure (Spearman correlation coefficient = -0.463, $p < 0.0001$)

Conclusion:

We have found that IIH patients have lower levels of OLC than controls. Although low OLC levels relationship with IIH may not be causative, it may play a role in the pathophysiology of IIH.

References:

1. Borsody M, Semenov I, Carroll K, Kessler A, Dubow et al. The relation of brain ouabain-like compounds and idiopathic intracranial hypertension. *Headache* 2006; 46: 1255-60.
2. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology* 2002; 59:1492-5.

Key Words: Idiopathic intracranial hypertension, Oubain-like compounds

Financial Disclosure: The authors had no disclosures.

Poster 2*

Correlation between multifocal pattern electroretinogram and Fourier-domain OCT in eyes with temporal hemianopia from chiasmal compression

Mário Monteiro, Kenzo Hokazono, Leonardo Cunha

University of São Paulo Medical School, Division of Ophthalmology, São Paulo, São Paulo, Brazil

Introduction:

To investigate the correlation between multifocal pattern electroretinogram (mfPERG) amplitude and fourier-domain optical coherence tomography (OCT) macular and retinal nerve fiber layer (RNFL) thickness measurements in eyes with temporal hemianopia from chiasmal compression.

Methods:

Twenty-five eyes from 25 patients with permanent temporal visual field (VF) defects from chiasmal compression and 25 eyes from 25 healthy subjects underwent mfPERG using a stimulus pattern of 19 rectangles, each consisting of twelve squares, standard automated perimetry (SAP) and fourier-domain OCT macular measurements. mfPERG response was determined for the central rectangle, the complete set of 19 rectangles, for the nasal and temporal hemifields (8 rectangles each) and for each quadrant (3 rectangles) in both patients and controls. The OCT macular thickness was registered according to an overlaid OCT generated checkboard with 36 checks. Macular thickness measurements were averaged globally and separately for each quadrant and half of that area, whereas RNFL thickness was determined for the average and sectoral segments around the disc. Comparisons were made using variance analysis. Correlations between OCT and mfPERG measurements were verified with Pearson's correlation coefficients and linear regression analysis.

Results:

mfPERG amplitude and OCT measurements were significantly lower in eyes with temporal field defect than in normals. In general, a significant correlation was found between mfPERG and both RNFL and macular thickness parameters. Correlations with mfPERG were slightly stronger for the OCT-measured macular thickness measurements compared to RNFL thickness parameters, although both sets of OCT measurements were strongly correlated with mfPERG measurements.

Conclusion:

In patients with chiasmal compression, mfPERG amplitudes and OCT macular thickness were significant related. Both mfPERG and OCT quantify neuronal loss and the technologies can be complementary in understanding structure-function relationship in patients with chiasmal compression.

References:

1. Hood DC, Xu L, Thienprasiddhi P, et al. The pattern electroretinogram in glaucoma patients with confirmed visual field deficits. *Invest Ophthalmol Vis Sci* 2005;46:2411-8.
2. Cunha LP, Oyamada MK, Monteiro ML. Pattern electroretinograms for the detection of neural loss in patients with permanent temporal visual field defect from chiasmal compression. *Doc Ophthalmol* 2008;117:223-32.
3. Monteiro ML, Hokazono K, Cunha LP, Oyamada MK. Multifocal pattern electroretinography for the detection of neural loss in eyes with permanent temporal hemianopia or quadrantanopia from chiasmal compression. *Br J Ophthalmol* 2011 Mar 17 [Epub ahead of print]
4. Parisi V, Manni G, Centofanti M, Gandolfi SA, Olzi D, Bucci MG. Correlation between optical coherence tomography, pattern electroretinogram, and visual evoked potentials in open-angle glaucoma patients. *Ophthalmology* 2001;108:905-12.
5. Monteiro ML, Cunha LP, Costa-Cunha LV, Maia OO, Jr., Oyamada MK. Relationship between optical coherence tomography, pattern electroretinogram and automated perimetry in eyes with temporal hemianopia from chiasmal compression. *Invest Ophthalmol Vis Sci* 2009;50:3535-41.
6. Danesh-Meyer HV, Carroll SC, Foroozan R, et al. Relationship between retinal nerve fiber layer and visual field sensitivity as measured by optical coherence tomography in chiasmal compression. *Invest Ophthalmol Vis Sci* 2006;47:4827-35.

Key Words: multifocal pattern electroretinogram, optical coherence tomography, band atrophy, chiasmal compression

Financial Disclosure: The authors had no disclosures.

Poster 3*

Development of Pupillary Adrenergic Supersensitivity after Pharmacologic Induction of Oculosympathetic Defect

Christopher Kirkpatrick, Randy Kardon

University of Iowa Hospitals and Clinics, Iowa City, IA, USA

Introduction:

Interruption of oculosympathetic nerve activity induces the development of adrenergic supersensitivity of end-organs. Evidence suggests that this receptor upregulation takes 3-5 days after denervation, but some report as few as 36 hours. Brimonidine is a selective alpha-2 agonist and will produce a pharmacologic oculosympathetic deficit by inhibiting presynaptic sympathetic neurotransmitter release. Apraclonidine is an alpha-2 agonist with weak alpha-1 agonistic properties that will manifest if end-organ supersensitivity is present. This study seeks to investigate the time course for development of adrenergic supersensitivity by monitoring pupil size in response to apraclonidine after the induction of a pharmacologic oculosympathetic defect using brimonidine.

Methods:

In this cohort study, 10 healthy subjects were prospectively studied by administering one drop of 0.2% brimonidine to the right eye twice daily for 5 days to induce a unilateral oculosympathetic defect. The left eye was used as a control. Adrenergic supersensitivity was assessed by measuring the pupillary response of both eyes to one drop of 0.5% apraclonidine daily.

Results:

9 of 10 subjects developed an anisocoria 2 days after initiation of unilateral brimonidine that persisted throughout all study days. This anisocoria was reversed in 8 of 9 subjects by the third study day, 45 minutes after administration of one drop of apraclonidine bilaterally. 1 subject never developed an anisocoria with daily brimonidine dosing and 1 subject had a baseline dilation reaction to apraclonidine and could not be included in analysis.

Conclusion:

The results of this study suggest that detectable adrenergic supersensitivity of end-organs develops in as few as 48 hours from the onset of the functional sympathetic denervation, induced in normal subjects with brimonidine, and that apraclonidine can be utilized as a diagnostic aid for detecting oculosympathetic defects in an acute setting of 48 hours from onset.

References:

1. Freedman KA. Brown SM. Topical apraclonidine in the diagnosis of suspected Horner syndrome. *Journal of Neuro-Ophthalmology*. 25(2):83-5, 2005 Jun.
2. Kardon R. Are we ready to replace cocaine with apraclonidine in the pharmacologic diagnosis of Horner syndrome? *Journal of Neuro-Ophthalmology*. 25(2):69-70, 2005 Jun.
3. Brown SM. The utility of 0.5% apraclonidine in the diagnosis of horner syndrome. *Archives of Ophthalmology*. 123(4):578; 2005 Apr.
4. Kesler A. Shemesh G. Rothkoff L. Lazar M. Effect of brimonidine tartrate 0.2% ophthalmic solution on pupil size. *Journal of Cataract & Refractive Surgery*. 30(8):1707-10, 2004 Aug.
5. Thordsen JE. Bower KS. Warren BB. Stutzman R. Miotic effect of brimonidine tartrate 0.15% ophthalmic solution in normal eyes. *Journal of Cataract & Refractive Surgery*. 30(8):1702-6, 2004 Aug.

Key Words: apraclonidine, adrenergic supersensitivity, oculosympathetic defect

Financial Disclosure: Dr. Randy Kardon discloses financial affiliation as consultant for Novartis and steering committee for OCTiMS multicenter study.

Poster 4*

Sleepiness and circadian timing have a differential effect on pupil responses mediated by inner retinal versus outer retinal photoreception

Aki Kawasaki¹, Lorette Léon¹, Sylvain Crippa¹, Mirjam Münch²

¹Hopital Opthalmique Jules Gonin, Lausanne, Switzerland, ²Ecole Polytechnique Fédérale de Lausanne, Lausanne, Switzerland

Introduction:

Non-visual, light-dependent functions of the eye are signalled by intrinsically photosensitive retinal ganglion cells that express melanopsin photopigment. We wished to identify the pupil response parameter that best distinguished effects of sleepiness (elapsed time of wakefulness) from circadian rhythms (biological clock).

Methods:

Ten healthy adults were tested hourly in two 12-hour sessions which covered a 24-hour period. The tests were: pupil light responses to red light (635±18 nm) and blue light (463±24 nm) having equivalent irradiance (1×10^{14} photons/cm²/s), salivary melatonin levels and assessments of subjective sleepiness. The pupil response was defined from the maximal pupil contraction to light stimulation and the post-stimulus response which was measured in 3 ways: the pupil size at 6 seconds following termination of a 1-second light stimulus and following termination of a 30-second light stimulus, the area within the stimulus-response curve during the 60 seconds after the 1s stimulus and the exponential rates of re-dilation back to baseline.

Results:

Compared to red light, maximal pupillary contraction was greater to blue light and demonstrated a more prolonged contraction following light termination, i.e. greater post-stimulus response ($p < 0.05$ for all parameters). There was a significant trend of increasing contraction amplitude with elapsed time of wakefulness for both blue and red 1-second light stimuli. Post-stimulus response parameters following blue light correlated with melatonin levels. In particular, the post-stimulus pupil size after 1-second blue light stimulation showed a circadian modulation with maximal values on the decreasing limb of melatonin secretion. In contrast, post-stimulus size at 6s following 1-second red light stimulation correlated with subjective sleepiness ($p < 0.05$).

Conclusion:

An effect of sleepiness versus circadian timing on the pupil light reflex can be differentiated from evaluation of the post-stimulus pupil response to red versus blue light. The pupil light reflex might be a useful marker to assess circadian-dependent light effects in humans.

References: None

Key Words: pupil, melanopsin, intrinsically photosensitive retinal ganglion cells, circadian rhythm

Financial Disclosure: Aki Kawasaki has received financial reimbursement for panel advisory work for Bayer SpG within the last 5 years. The remaining authors had no disclosures.

Poster 5*

Comparison of Cerebrospinal Fluid Opening Pressure in the Prone and Lateral Decubitus Positions

Anne Abel, Jeffrey Brace, Alexander McKinney, Lei Zhang, Andrew Harrison, Michael Lee

University of Minnesota, Minneapolis, MN, USA

Introduction:

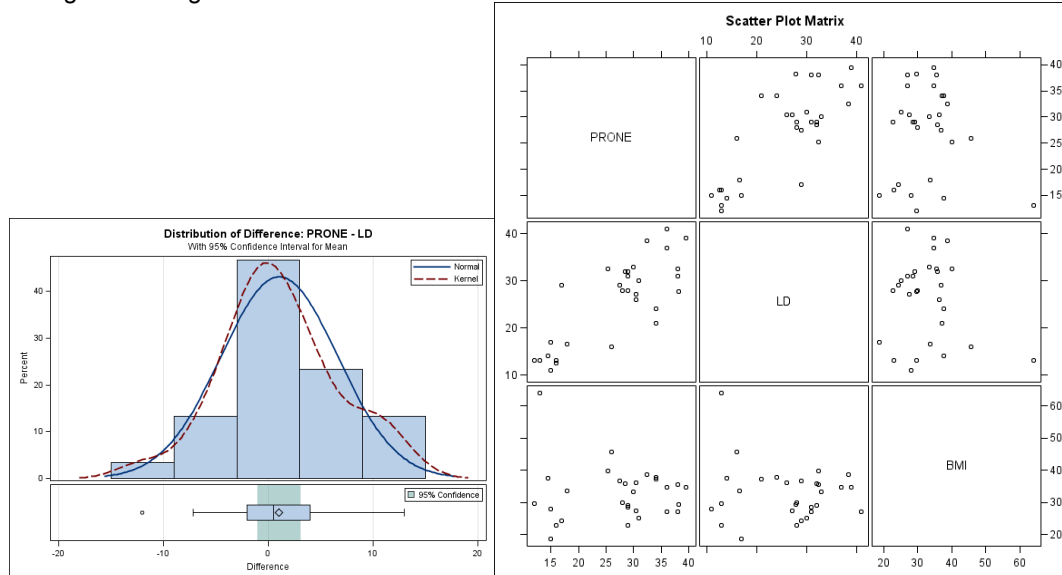
Prone is the preferred patient position for fluoroscopic guided lumbar puncture (LP). A normal range for lateral decubitus (LD) cerebrospinal fluid (CSF) opening pressure (OP) exists, but normative data for prone CSF OP does not. This study compares CSF OP values in the prone and LD positions. The effect of body mass index (BMI) on these OP measurements was also examined.

Methods:

Patients undergoing diagnostic or therapeutic fluoroscopic guided LP CSF OP were recruited. The LP needle and manometer were connected with flexible tubing, allowing OP measurement in both positions without CSF loss. Following prone fluoroscopic-guided LP, subjects were rolled to the LD position for repeat CSF OP measurement with legs extended and patient relaxed. Zero was defined at the level of the spinal canal for all OP measurements.

Results:

Thirty subjects participated. There were 26 (87%) women with a mean age of 31 years. Using the paired t-test, there was no significant difference between OP measured in the prone and LD position ($p=0.3108$, CI [-1.0286, 3.1219]). In 50% of subjects, the difference between OP in the two positions was ≤ 3 cm H₂O (Figure 1). Pearson's correlation did not show a correlation between CSF OP and BMI in either position (Figure 2). Among patients undergoing an LP for papilledema, 2 (9%) crossed the 25-cmH₂O threshold between positions, which could potentially change their diagnosis.



Conclusion:

A statistically significant difference between prone and LD CSF OP was not found. BMI does not appear to correlate with CSF OP measurement in either the LD or prone positions.

References: None

Key Words: lumbar puncture, fluoroscopy, prone, lateral decubitus, opening pressure

Financial Disclosure: The authors had no disclosures.

Poster 6*

The King-Devick (K-D) Test of Rapid Eye Movements: A Bedside Correlate of Disability and Quality of Life in Multiple Sclerosis

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Introduction:

Based on the speed of rapid number naming, the King-Devick (K-D) test captures impaired eye movements and saccades, findings that correlate with suboptimal brain function. Time scores for the K-D test are significantly higher (worse) in athletes immediately following concussion, consistent with involvement of the widely distributed visual pathways. We examined the potential role for the K-D test as a measure of eye movements in multiple sclerosis (MS), and determined the relation of scores to visual function and quality of life (QOL).

Methods:

Patients with MS and disease-free controls completed the K-D test at a single study visit. The K-D test requires <1 minute, and scores represent the total time needed to read single-digit numbers on 3 test cards. Patients were also evaluated using monocular and binocular low (2.5%, 1.25%) and high-contrast acuity, spectral-domain optical coherence tomography (OCT), NEI-VFQ-25 (QOL questionnaire), and MS Functional Composite (timed 25-foot walk, 9-hole peg test, and Paced Auditory Serial Addition Test [PASAT]).

Results:

In the MS cohort (n=40), K-D scores were significantly higher (worse) compared to disease-free controls (55.3±17.8 vs. 41.2±7.2 seconds, P=0.008, linear regression, accounting for age). Higher K-D scores in MS were associated with worse scores for vision-specific quality of life (P=0.002 for NEI-VFQ-25 composite, P=0.001 for 10-Item Neuro-Ophthalmic Supplement), binocular low- and high-contrast acuity (P=0.03), timed 25-foot walk (P<0.001), 9-hole peg test (P≤0.001), and 2-second PASAT (P=0.04, accounting for age). While correlations of OCT RNFL thickness did not reach significance (P=0.10) monocular low-contrast acuity (2.5%) correlated with K-D scores (P=0.04, GEE models) in the MS cohort.

Conclusion:

The K-D test, a <1 minute bedside assessment of rapid number naming, correlates well with binocular visual function, disability, and vision-specific quality of life in MS. The K-D test should be considered for inclusion in MS trials as a rapid efferent visual function test.

References:

1. Heitger MH, Jones RD, Macleod AD, et al. Impaired eye movements in post-concussion syndrome indicate suboptimal brain function beyond the influence of depression, malingering, or intellectual ability. *Brain* 132:2850-2570, 2009.
2. Galetta KM, Brandes LE, Maki K, et al. The King-Devick test and sports-related concussion: study of a rapid visual screening tool in a collegiate cohort. *J Neurol Sci* 309:34-39, 2011.
3. Galetta KM, Barrett J, Allen M, et al. The King-Devick test as a determinant of head trauma and concussion in boxers and MMA fighters. *Neurology* 76:1456-1462, 2011.

Key Words: multiple sclerosis, eye movements, visual function, King-Devick test

Financial Disclosure: Dr. S. Galetta has received honoraria from Biogen-Idec, Teva, and Novartis. Dr. Calabresi has received personal compensation for consulting and serving on scientific advisory boards from Biogen Idec, Teva, Vaccinex, Abbott, and Novartis; Dr. Calabresi has also received research funding from Biogen Idec, Teva, EMD Serono, Vertex, Genentech, Abbott, and Bayer. Dr. Frohman has received speaker fees from Biogen, Teva, Novartis, Acorda and Bayer. He has received consulting fees from Biogen, Teva, Acorda, Novartis and Abbott. Dr. Balcer has received honoraria from Biogen-Idec, Novartis, Bayer and Vaccinex.

Poster 7*

Why Mothers Say ‘Look At Me When I’m Talking To You’: The Effect of Amblyopia on Visual-Auditory Speech Perception

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Introduction:

Speech perception is multimodal, intertwining auditory signals with lip reading. With inaccurate visual input, speech is heard incorrectly, as demonstrated from an illusion called the McGurk effect. Here, a video is presented of a person silently mouthing the sound “ka.” The auditory output, however, is the sound “pa”. When the signals are presented simultaneously, 70% of normal individuals report hearing “ta.” However, children with amblyopia may have missed a critical window period of such audio-visual integration in the brain. Our study purpose is to determine if speech perception is influenced by amblyopia.

Methods:

Our stimuli present the face of a person on a 13” monitor, 50 cm away, at 60 dB, with the output as: visual “pa” with audio “pa” (i.e. an unedited video of the person saying “pa” normally), visual “ka” with audio “ka”, and visual “ka” with audio “pa.” Ten trials of each stimulus were presented in random order. Inclusion criteria for amblyopes were: 1) age at least 4 years; 2) amblyopia in only one eye; 3) stereopsis worse than 50 seconds; and 4) at least 20/25 acuity in the better-seeing eye at the time of testing. Inclusion criteria for control patients were: 1) age over 4 years; 2) 20/20 acuity in both eyes; and 3) 40 seconds of stereopsis. Test subjects viewed the stimulus binocularly, and were asked to choose what sound was heard: “pa”, “ka”, or “ta.”

Results:

Of the 50 trials among 5 control subjects, the fusion “ta” was heard 33 (66%) times. In contrast, of the 110 trials among 11 amblyopic subjects, “ta” was heard only 21 (19%) times; two-tailed Fisher’s exact test $p < 0.0001$.

Conclusion:

Children with a history of amblyopia, even if they are at least 20/25 in their better seeing eye and are tested binocularly, have impaired visual-audio fusion in speech perception.

References:

1. Schorr EA, Fox NA, van Wassenhove V, Knudsen EI. Auditory-visual fusion in speech perception in children with cochlear implants. *Proc Natl Acad Sci U S A*. 2005;102:18748-50
2. van Atteveldt N, Roebroek A, et al. Interaction of speech and script in human auditory cortex: insights from neuro-imaging and effective connectivity. *Hear Res*. 2009; 258:152-64.

Key Words: Amblyopia, Speech, Hearing, sensory integration

Financial Disclosure: The authors had no disclosures.

Poster 8*

Hypopigmented Mice as an Animal Model for Infantile Nystagmus Syndrome

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Introduction:

Individuals with oculocutaneous albinism are predisposed to visual system abnormalities affecting the retina and the retinofugal projections leading to reduced visual acuity and infantile nystagmus syndrome (INS) in a majority of the patients¹⁻⁴. Based on recent results in zebrafish⁵⁻⁸ and in humans (unpublished results), the INS-specific ocular motor instabilities may be due to the misprojection of retinal ganglion cell (RGC) axons in the optic chiasm. Since albino mice show a similar projection error⁹, we screened wildtype mice of varying pigmentation for ocular motor abnormalities.

Methods:

Two albino strains (Cri:CD1(ICR), BALB/cAnNTac), the grey DBA/1JbomTac, the brown 129S6/SvEvTac and the black C57BL/6NTac strains were screened using infrared-oculography. Varying visual stimuli (black background, white background, stationary pattern, rotating pattern) were displayed to the full or anterior visual field of the restrained mouse.

Results:

Reversal of the optokinetic reflex in albino mice has been reported before¹⁰. In our study, we also found spontaneous nystagmus, specifically jerks and oscillations, in hypopigmented mice during stimulation with a stationary background pattern, while in pigmented mice ocular positions were near to stable. In all three hypopigmented strains, the described ocular motor instabilities seemed to increase (more spontaneous oscillations and reversal of the OKR) with stimulation of the temporal retina, where the misprojecting axons originate from.

Conclusion:

Our study suggests that albino mice might be considered as an animal model for INS. The pigmentation of the retinal pigment epithelium has been shown to influence the retinal anatomy¹¹ and the projection of retinal ganglion cell axons^{12, 13}. Based on recent results in zebrafish⁵⁻⁸ we hypothesize that the spontaneous nystagmus and the reversal of the OKR may be due to the projection error. Further studies will clarify whether this hypothesis holds true.

References:

1. Gronskov K, Ek J, Brondum-Nielsen K. Oculocutaneous albinism. *Orphanet J Rare Dis*. 2007; 2: 43.
2. Kelly JP, Weiss AH. Topographical retinal function in oculocutaneous albinism. *Am J Ophthalmol*. 2006; 141: 1156-1158.
3. Collewijn H, Apkarian P, Spekreijse H. The oculomotor behaviour of human albinos. *Brain*. 1985; 108: 1-28.
4. Dorey SE, Neveu MM, Burton LC, Sloper JJ, Holder GE. The clinical features of albinism and their correlation with visual evoked potentials. *Br J Ophthalmol*. 2003; 87:767-772.
5. Huang YY, Neuhauss SC. The optokinetic response in zebrafish and its applications. *Front Biosci*. 2008; 13: 1899-1916.
6. Huang YY, Rinner O, Hedinger P, Liu SC, Neuhauss SC. Oculomotor instabilities in zebrafish mutant belladonna: a behavioral model for congenital nystagmus caused by axonal misrouting. *J Neurosci*. 2006; 26: 9873-9880.
7. Huang YY, Tschopp M, Neuhauss SC. Illusionary self-motion perception in zebrafish. *PLoS One*. 2009; 4: e6550.
8. Huang YY, Tschopp M, Straumann D, Neuhauss SC. Vestibular deficits do not underlie looping behavior in achiasmatic fish. *Commun Integr Biol*. 2010; 3: 379-381.

9. Balkema GW, Drager UC. Origins of uncrossed retinofugal projections in normal and hypopigmented mice. *Vis Neurosci*. 1990; 4: 595-604.
10. Mangini NJ, Vanable JWJ, Williams MA, Pinto LH. The optokinetic nystagmus and ocular pigmentation of hypopigmented mouse mutants. *J Comp Neurol*. 1985; 241:191-209.
11. Donatien P, Jeffery G. Correlation between rod photoreceptor numbers and levels of ocular pigmentation. *Invest Ophthalmol Vis Sci*. 2002; 43: 1198-1203.
12. Marcus RC, Wang LC, Mason CA. Retinal axon divergence in the optic chiasm: midline cells are unaffected by the albino mutation. *Development*. 1996; 122: 859-868.
13. Gimenez E, Lavado A, Giraldo P, Cozar P, Jeffery G, Montoliu L. A transgenic mouse model with inducible Tyrosinase gene expression using the tetracycline (Tet-on) system allows regulated rescue of abnormal chiasmatic projections found in albinism. *Pigment Cell Res*. 2004; 17:363-370.

Key Words: infantile nystagmus syndrom, ocular motor instability, hypopigmented mice, albinism, infrared oculography

Financial Disclosure: The authors had no disclosures.

Poster 9*

Longitudinal Study of Retinal Ganglion Cell Layer Neuronal Loss by Spectral-Domain Optical Coherence Tomography (SD-OCT) in Multiple Sclerosis

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Introduction:

Neuronal degeneration is an important correlate of disability in MS. We used SD-OCT with computerized segmentation to determine how ganglion cell neuronal loss over time relates to optic neuritis (ON) history, retinal nerve fiber layer (RNFL) thinning, vision, and quality of life (QOL) in MS.

Methods:

SD-OCT images were captured at 6-month intervals. Segmentation was performed automatically using a customized software developed at the University of Pittsburgh. Thicknesses of ganglion cell/inner plexiform layer (GCL+IPL), RNFL, outer plexiform/inner nuclear layer (OPL+INL), and outer nuclear/photoreceptor layer (ONL+PRL) were measured in MS and control eyes. High-contrast visual acuity (VA), low-contrast acuity (2.5%, 1.25%), and NEI-VFQ-25 QOL scores were determined.

Results:

Among 117 MS eyes (60 patients) with ≥ 6 months follow-up, average GCL+IPL thickness decreased by 3.0 microns during a mean follow-up of 12 months ($P < 0.001$ vs. controls, GEE models accounting for age and within-patient, inter-eye correlations). GCL+IPL thinning increased over time, with average losses of 2.0 microns at 12-18 months ($P = 0.02$), 4.6 microns at 18-24 months ($P = 0.02$), and 5.8 microns at > 24 months ($P < 0.001$ vs. 6-12-month follow-up interval). Similar patterns were noted for macular RNFL, with average losses of 2.8 microns at 12-18 months and 5.4 microns by > 24 months. Associations of GCL+IPL thinning with increasing follow-up remained significant in models accounting for RNFL thinning. GCL+IPL thinning was significant at > 24 months for eyes with ($P = 0.03$) and without ON history ($P < 0.001$). GCL+IPL thinning was associated with worse scores for low-contrast acuity ($P = 0.03$) and NEI-VFQ-25 ($P = 0.02$).

Conclusion:

GCL+IPL loss occurs over time in MS eyes with and without history of ON, and is significant even when accounting for RNFL thinning. Loss of GCL+IPL is associated with reduced visual function and QOL, providing further evidence of the importance of neuronal loss in MS and supporting a role for GCL+IPL measurement in MS trials of neuroprotection and repair.

References:

1. Talman LS, Bisker ER, Sackel DJ, et al. Longitudinal study of vision and retinal nerve fiber layer thickness in multiple sclerosis. *Ann Neurol* 67:749-760, 2010.
2. Davies EC, Galetta KM, Sackel DJ, et al. Retinal ganglion cell layer volumetric assessment by spectral-domain optical coherence tomography in multiple sclerosis: application of a high-precision manual estimation technique. *J Neuro-Ophthalmol* 31:260-264, 2011.
3. Saidha S, Syc SB, Durbin MK, et al. Visual dysfunction in multiple sclerosis correlates better with optical coherence tomography derived estimates of macular ganglion cell thickness than peripapillary retinal nerve fiber layer thickness. *Mult Scler* 2011 [epub August 24].

Key Words: multiple sclerosis, optical coherence tomography (OCT), ganglion cell layer, neuroprotection

Financial Disclosure: Dr. Galetta has received honoraria from Biogen-Idec, Teva, and Novartis. Dr. Calabresi has received personal compensation for consulting and serving on scientific advisory boards from Biogen Idec, Teva, Vaccinex, Abbott, and Novartis; Dr. Calabresi has also received research funding from Biogen Idec, Teva, EMD Serono, Vertex, Genentech, Abbott, and Bayer. Dr. Frohman has received speaker fees from Biogen, Teva, Novartis, Acorda and Bayer. He has received consulting fees from Biogen, Teva, Acorda, Novartis and Abbott. Dr. Balcer has received honoraria from Biogen-Idec, Novartis, Bayer and Vaccinex.

Poster 10*

Saccadic adaptation in amblyopia

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Introduction:

Saccadic adaptation is an important motor learning mechanism that maintains the accuracy of goal-directed saccades. It is generally believed to be driven by visual error signal at the end of saccades. Amblyopia is characterized by impairment of spatiotemporal visual processing. The purpose of this study is to investigate the effects of spatiotemporal visual deficits in amblyopia on saccadic adaptation.

Methods:

Study participants included 10 visually normal adults and 7 adult patients with amblyopia (5 anisometric, 1 strabismic, 1 mixed). Participants were instructed to follow a visual target on a screen while their eye movements were recorded. Saccadic adaptation was induced experimentally using a double-step target paradigm, in which the visual target (at 18° eccentricity) was displaced toward initial fixation (by 4°) during the saccade. The experiments included a single-step pre-test, a double-step adaptation test, and a single-step post-test, performed during binocular viewing and monocular viewing with the amblyopic and fellow eye in three different sessions. Saccadic latencies and percentage change in saccadic gain were measured.

Results:

Patients with amblyopia adapted their saccadic gain to a lesser extent than normal subjects during amblyopic eye viewing (patients=54±4.46%, normal=70.4±3.73%; p=0.013). Furthermore, patients with anisometric amblyopia exhibited decreased saccadic gain adaptation than normal participants during both amblyopic eye viewing (patients=42.8%, normal=69.1%; p=0.001) and binocular viewing (patients=51.8%, normal=71.7%; p=0.012). Initiation of saccades was also delayed in patients during amblyopic eye viewing in both pre-adaptation and adaptation phase.

Conclusion:

Saccadic gain adaptation was less robust in patients during amblyopic eye viewing. The effect was more pronounced in patients with anisometric amblyopia who exhibited impaired adaptation during both amblyopic eye and binocular viewing. Our findings suggest that processing of visual error information is imprecise in amblyopia, and support the hypothesis that the error signal that drives saccadic adaptation is visual in nature.

References:

1. Barrett, B.T., Bradley, A., & McGraw, P. V. (2004). Understanding the neural basis of Amblyopia. *The Neuroscientist*, 10(2): 106-117.
2. Pelisson, D., Alahyane, N., Panouilleres, M & Tilikete, C. (2010). Sensorimotor adaptation of saccadic eye movements. *Neuroscience & Behavioral Reviews*, 34: 1103-1120.

Key Words: Amblyopia, Physiological Adaptation, Saccades, Vision, Spatial Precision

Financial Disclosure: The authors had no disclosures.

Poster 11*

Optic nerve and melanopsin retinal ganglion cells involvement in relation to circadian dysfunction in Alzheimer disease

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Introduction:

Optic nerve involvement and circadian rhythm disturbances are reported in Alzheimer disease (AD) (1,2). We aimed at characterizing optic nerve involvement with particular reference to melanopsin retinal ganglion cells (mRGCs) in relation to the occurrence of rest-activity rhythm dysfunction in AD.

Methods:

Ophthalmologic evaluation and retinal nerve fiber layer thickness measurements by optical coherence tomography (Stratus OCT 3000, Zeiss) were performed in 15 AD (age 72.3 ± 10.8) and 74 controls (age 69.1 ± 8.1). Actigraphic monitoring for 7 days was performed in 14 AD patients (age 72 ± 9.03) and 10 controls (age 65.8 ± 7.54). Non-parametric methods were applied to assess interdaily stability (IS), intradaily variability (IV) and relative amplitude (RA) of rest-activity rhythm. We also performed immunohistochemical and morphometric analysis of mRGCs in 15 AD (age 82.1 ± 12.5 ; 1) and control (age 78.6 ± 15.3) retinas. Axonal count on optic nerve cross-sections was also performed in 7 AD and 7 controls.

Results:

OCT evaluation demonstrated reduced average ($p=0.01$), superior ($p=0.006$) and nasal ($p=0.01$) RNFL thickness in AD patients. Average ($p=0.002$), superior ($p=0.03$) and nasal ($p=0.02$) RNFL thickness significantly correlate with age in AD patients. Actigraphic monitoring demonstrated increased IV ($p=0.026$), reduced sleep minutes ($p=0.036$) and sleep efficiency ($p=0.002$) in AD patients. No significant correlation was detected between OCT measurements and actigraphic data. Similar loss of mRGCs and axons was observed in AD retinas ($p=0.005$) and optic nerve cross-sections ($p=0.048$). Thus, mRGCs/RGCs ratio was similar in AD (1.48%) and controls (1.38%).

Conclusion:

We demonstrated optic nerve pathology, and loss of mRGCs in AD. The reduction of mRGCs may contribute to circadian rhythm dysfunction in AD.

References:

1. Guo L, Duggan J, Cordeiro MF. Alzheimer's disease and retinal neurodegeneration. *Curr Alzheimer Res.* 2010;7:3-14.
2. Wu YH, Swaab DF. Disturbance and strategies for reactivation of the circadian rhythm system in aging and Alzheimer's disease. *Sleep Med.* 2007 Sep;8(6):623-36.

Key Words: Alzheimer, optic nerve, melanopsin, retinal ganglion cells, circadian rhythms

Financial Disclosure: The authors had no disclosures.

Poster 12*

The effect of strabismic amblyopia and strabismus without amblyopia on saccade latencies

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Introduction:

Impairment of spatiotemporal visual processing is the hallmark of amblyopia, but its effects on eye movements during visuomotor tasks have rarely been studied systematically. Here, we investigate saccadic eye movements in patients with strabismic amblyopia, and compare their behaviours with patients who have strabismus without amblyopia and with visually-normal control subjects.

Methods:

Fourteen patients with strabismic amblyopia (5 with mild acuity deficits and stereopsis [MILD+], 5 with mild acuity deficits and negative stereopsis [MILD-], 4 with severe acuity deficits and negative stereopsis [SEVERE]), 11 patients with strabismus (7 had negative stereopsis), and 14 visually-normal subjects were recruited. Each trial began with a central fixation cross presented on a computer monitor. After a delay, a target appeared at $\pm 5^\circ$ or $\pm 10^\circ$ of fixation. Subjects were instructed to execute saccades to targets during binocular and monocular viewing.

Results:

A significant interaction between group and viewing condition was found for saccade latency ($p < 0.0001$). Patients with strabismic amblyopia who had residual stereopsis had longer saccade latencies during binocular viewing (192 ± 42 ms) as compared to patients with negative stereopsis (MILD-: 165 ± 31 ms; SEVERE-: 165 ± 41 ms) and control subjects (170 ± 29 ms). The presence of stereopsis did not affect saccade latencies in patients with strabismus without amblyopia during binocular viewing (stereo: 171 ± 23 ms; no stereo: 180 ± 23 ms). As expected, all patients with amblyopia had longer saccade latencies when viewing with the amblyopic eye compared to binocular or fellow eye viewing.

Conclusion:

Our data show that residual binocularity impacts saccade initiation during binocular viewing in patients with strabismic amblyopia, but not in patients with strabismus without amblyopia. Patients with strabismic amblyopia and negative stereopsis had shorter latencies than those with positive stereopsis, suggesting that the amblyopic eye might have been under stronger suppression, such that input from that amblyopic eye is less likely to be integrated and interfere with processing of input from the fellow eye during binocular viewing.

References: None

Key Words: eye movements, amblyopia, strabismus, binocular vision

Financial Disclosure: The authors had no disclosures.

Poster 13*

Giant Cell Arteritis: Treatment of *Burkholderia Pseudomallei* Infection with Antibiotics.

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Introduction:

At NANOS 2011 we presented data establishing an association between a strain of *Burkholderia pseudomallei* and giant cell arteritis (GCA). This strain is sensitive to minocycline and doxycycline *in vitro*. We are treating a cohort of GCA patients with a course of minocycline or doxycycline in addition to steroids to determine if antibiotic treatment is an effective adjuvant in the treatment of this disease.

Methods:

Five patients with biopsy-proven GCA were enrolled. One patient, who was biopsy negative but fit the clinical and laboratory features of GCA, was also enrolled. Subjects underwent monthly evaluations to monitor for signs and symptoms of relapse. ESR, CRP, and platelet counts, as well as an ELISA for *B. pseudomallei* LPS were evaluated at various intervals.

Results:

Ages of the 6 subjects ranged from 69-88 years (mean 80 years; 3 male; 3 female). All subjects experienced at least one neuro-ophthalmologic manifestation. Three subjects were newly diagnosed with GCA and 3 subjects were relapsing at the time of enrolment. All subjects had positive ELISA tests for *B. pseudomallei*. Five subjects were treated with oral minocycline for one month; one subject was treated with oral doxycycline for 2 months. All subjects followed a prednisone taper. All subjects had declining serum levels of *B. pseudomallei* LPS. One subject experienced an acute visual change associated with a flame haemorrhage attributed to concomitant warfarin use. No other adverse events were recorded, no subjects have suffered a relapse, and all subjects have continued to taper off steroids.

Conclusion:

Minocycline and doxycycline appear to be effective in reducing the burden of *B. pseudomallei* infection. Although these data appear promising, a prospective randomized trial of steroids+placebo vs. steroids+antibiotics will be required to determine if antibiotic treatment is an effective adjuvant in the treatment of GCA.

References: None

Key Words: Giant Cell Arteritis, *Burkholderia Pseudomallei*

Financial Disclosure: The authors had no disclosures.

Poster 14*

The Role of Vitamin D Status in Optic Neuritis

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Introduction:

Optic neuritis (ON) is a common manifestation of multiple sclerosis (MS), and provides a single-lesion model to assess both inflammatory and degenerative activity in the central nervous system using optical coherence tomography (OCT) to measure retinal nerve fiber layer thickness (RNFLT). Vitamin D insufficiency is an accepted risk factor in MS development, and vitamin D sufficiency appears to be an ameliorator of inflammatory activity. We hypothesize that vitamin D sufficiency will be associated with higher RNFLT values, indicated better axonal integrity in the anterior visual pathway. The primary objective of this study is to evaluate the relationship between vitamin D status and ON recovery, using RNFLT at 6 months as the main outcome measure.

Methods:

In this prospective pilot cohort study, patients will undergo OCT to assess mean and temporal quadrant RNFLT, macular volume (MV) and other visual outcomes at baseline, months 3 and 6. Vitamin D status (25(OH)D) will be evaluated at baseline, months 3 and 6, with sufficiency defined as 25(OH)D => 80 nmol/L.

Results:

The first 17 patients enrolled (14 women, 3 men) had a mean baseline 25(OH)D of 84 nmol/L. Vitamin D insufficiency was associated with greater overall RNFLT (144 vs 89 μm , $p=0.030$), increased temporal quadrant RNFLT (78 vs 58 μm , $p=0.063$) and higher MV (10.4 vs 9.6 mm^3 , $p=0.0014$). Vitamin D insufficient patients had higher RNFLT values in their unaffected eye (101 vs 78 μm , $p=0.0006$). Data from month 3 thus far supports baseline trends.

Conclusion:

Early analysis suggests that vitamin D insufficiency is associated with a greater degree of optic nerve edema at ON onset. Additional patients and data at 3 and 6 months will determine if this trend continues, and if vitamin D status ultimately impacts the degree of axonal loss as evidenced by RNFLT measures.

References: None

Key Words: Vitamin D, Optic Neuritis, Multiple Sclerosis, Neuroprotective, Central Nervous System Inflammation

Financial Disclosure: Dr. Jodie Burton received advisory board compensation from Novartis and Biogen Idec. Dr. Fiona Costello received advisory board compensation from Novartis and Biogen Idec. The remaining author had no disclosures.

Poster 15

The impact of gender on recovery from acute optic neuritis

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Introduction:

Optic neuritis (ON) is an inflammatory injury of the optic nerve, which represents the first clinical manifestation of multiple sclerosis (MS) in 20% of patients. Most ON patients are young women, albeit the basis of the female predilection remains unclear. Gender effects are significant in MS, with men having worse clinical outcomes than women. Our primary objective was to explore the impact of gender in ON, by determining if retinal nerve fiber layer (RNFL) values differed between men and women after an acute ON event.

Methods:

In this prospective cohort study, 39 men and 105 women with acute ON underwent repeat ophthalmic and optical coherence tomography (OCT) testing over a 6-month period. The main outcome measures were RNFL measurements in ON eyes for male and female patients 6-months after ON.

Results:

Men were older (mean age = 39 years) than women (35) ($p = 0.05$) in this study; and more men (62%) than women (41%) had a diagnosis of MS ($p = 0.02$). Because age and MS diagnosis were two significant covariates, both variables were controlled for in subsequent multiple regression analyses, along with disease duration, steroid therapy, and disease modifying therapy use. Six-months after ON, mean RNFL values were lower in men ($74\mu\text{m}$) than women ($91\mu\text{m}$) ($p < 0.001$). Retinal nerve fiber layer measurement below $75\mu\text{m}$, were associated with clinically and statistically significant reductions in visual recovery after ON.

Conclusion:

Men have significantly lower RNFL values than women after acute ON, which may be viewed as a single-lesion model of MS. Our findings raise intriguing questions about the potential impact of gender in MS, which may be explored in future studies.

References: None

Key Words: optic neuritis, Gender, multiple sclerosis, retinal nerve fiber layer, optical coherence tomography

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Poster 16

Cranial Neuropathy and Ectatic Vessels: A Close Up of Vascular Loops

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Introduction:

With the advent of high resolution MRI with constructive interference in steady state (CISS) we are now able to appreciate vasculature in contact with cranial nerves with higher resolution than before.

Methods:

We reviewed all cases in the neuro-radiology database that had cranial neuropathy and imaging. Entry criteria included: cranial neuropathy, CISS imaging, and vascular distortion by an artery near the nerve root entry zone (REZ). Anyone with aneurysmal compression was excluded. We also reviewed the literature for similar cases with adequate imaging. We analyzed the medical history for risk factors for microvascular disease including advanced age, hypertension, hypercholesterolemia, and tobacco use.

Results:

Three cases met our criteria. We found seven cases in the literature. The mean age was 58 years. Eight of the ten patients had hypertension and seven had hypertension as the sole vascular risk factor. One patient was female. Eight cases were associated with vertebral basilar dolichoectasia, two with an anterior inferior cerebellar artery, and one with a large posterior communicating artery. Three patients had recurrence of cranial neuropathy. One patient had eleven episodes of abducens palsy of the same nerve. One patient developed diplopia after a bicycle race, and one patient developed two recurrences, each after febrile illness. Sixty-three percent had resolution of symptoms.

Conclusion:

Most reports of dolichoectasia and cranial neuropathy have been single case reports. We present 3 new cases describing cranial neuropathies from vessels using CISS. Combining these cases with the literature, we found the previously described male predominance. Hypertension was the only vascular risk factor. Patients with nerve palsies associated with vascular distortion at the REZ have similar rates of symptom resolution as patients with purely ischemic peripheral nerve palsies. However, these cases suggest that the number of recurrences may be higher. A proposed mechanism may be an acute demyelinating injury at the vulnerable REZ.

References:

1. Durand JR, Samples JR. Dolichoectasia and cranial nerve palsies. A case report. *J Clin Neuroophthalmol.* 1989 Dec; 9(4): 249-53
2. Zhu Y, Thulborn K, Curnyn K, Goodwin J. Sixth cranial nerve palsy caused by compression from a dolichoectatic vertebral artery. *J Neuroophthalmol.* 2005 Jun; 25(2): 134-5
3. Sandvand KA, Ringstad G, Kerty E. Periodic abducens nerve palsy in adults caused by neurovascular compression. *J Neurol Neurosurg Psychiatry.* 2008 Jan; 79(1): 100-2. Epub 2007 Aug 23
4. Kato H, Nakajima M, Ohnaka Y, Ishihara K, Kawamura M. Recurrent abducens nerve palsy associated with neurovascular compression. *J Neurol Sci.* 2010 Aug 15; 295(1-2): 135-6
5. Sanders SK, Kawasaki A, Purvin VA. Long-term prognosis in patients with vasculopathic sixth nerve palsy. *Am J Ophthalmol.* 2002 Jul; 134(1): 81-4

Key Words: Dolichoectasia, Cranial neuropathy, Hypertension, Constructive Interference in Steady State

Financial Disclosure: The authors had no disclosures.

Poster 17

Relationship between fixation stability and saccade initiation in patients with amblyopia.

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Introduction:

Saccades have longer and more variable latency in patients with amblyopia¹. Patients with amblyopia often have unsteady fixation which may interfere with saccade initiation. In this study, we investigated the relationship between fixation stability and saccade initiation in patients with amblyopia.

Methods:

Thirteen patients with amblyopia were recruited and participated in 2 experimental sessions. In the first session, fixation stability was examined. Patients fixated on a stimulus for 15 sec during binocular and monocular viewing (fellow & amblyopic eye) while eye position was recorded with a video-based eye-tracker. Fixation stability was quantified with a bivariate contour ellipse area (BCEA), based on the standard deviations of the horizontal and vertical eye positions during fixation. Saccadic eye movements were examined in 11 of 13 patients in the second session. Each trial began with a central fixation cross presented on a computer monitor. After a variable delay, a target appeared randomly at $\pm 10^\circ$ of fixation. Patients were instructed to execute saccades to the targets.

Results:

Mean BCEA was twice as large for the amblyopic than fellow eyes when viewing was binocular (0.36 deg^2 vs. 0.17 deg^2 , $p < 0.001$) or monocular (0.62 deg^2 vs. 0.30 deg^2 , $p < 0.01$). Saccade latencies (210 ± 34 ms) during amblyopic eye viewing were longer ($p < 0.05$) than when viewing binocularly (180 ± 29 ms) or with the fellow eye (177 ± 29 ms). Although the relationships between fixation stability and saccade latency did not reach statistical significance, there were trends showing that: 1) patients with better fixation stability in the amblyopic eye have shorter saccade latency ($r = 0.29$) during amblyopic eye viewing; 2) better fixation stability is associated with lower variability of saccade latencies during amblyopic ($r = 0.33$) and fellow eye viewing ($r = 0.38$).

Conclusion:

Fixation stability is impaired in amblyopia. Preliminary data suggested that fixation instability is associated with longer and more variable saccade latencies.

References:

1. Niechwiej-Szwedo E, Goltz H, Chandrakumar M, Hirji Z, Wong A. Effects of anisometric amblyopia on visuomotor behavior, I: saccadic eye movements, IOVS, 51(12),6348-54, 2010.

Key Words: fixation stability, oculomotor control, amblyopia

Financial Disclosure: The authors had no disclosures.

Poster 18

MRI Findings in Idiopathic Intracranial Hypertension (IIH) Compared with Cerebral Venous Thrombosis (CVT)

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Introduction:

Numerous publications have emphasized that IIH patients often have MRI findings suggestive of raised ICP in addition to TSS on vascular imaging. We compared MRI findings suggestive of elevated ICP in IIH patients with transverse sinus stenosis (TSS) versus CVT patients.

Methods:

Among 240 patients who underwent standardized contrast-enhanced brain MRI and contrast-enhanced MRV at our institution between 9/2009 and 9/2011, 60 with abnormal imaging findings on MRV were included: 29 patients with definite IIH, and 31 with definite CVT. Medical records were reviewed, and imaging studies were prospectively evaluated by the same neuroradiologist to assess for presence or absence of TSS, site of CVT if present, orbital findings of posterior globe flattening, optic nerve sheath dilation/tortuosity, and the size/appearance of the sella turcica.

Results:

29 IIH patients (28 women, 19 African-Americans, mean-age 30.6, mean-BMI 35.2) had bilateral (28) or unilateral (1) TSS. 31 CVT patients (19 women, 13 African-Americans, mean-age 45.6, mean-BMI 28.6) had thrombosis of the sagittal (3), sigmoid (3), cavernous (1), unilateral transverse (7), or multiple (16) sinuses or cortical veins (1). 14 CVT patients presented with raised ICP, 15 had focal symptoms, 2 were asymptomatic. IIH patients had larger sella turcicas than CVT patients ($p < 0.03$). Empty or partially-empty sellas were more common in IIH (3/29 and 24/29) than in CVT patients (1/31 and 19/31) ($p < 0.001$). Flattening of the globes and dilation/tortuosity of the optic nerve sheaths were more common in IIH (20/29 and 18/29) than in CVT patients (13/31 and 5/31) ($p < 0.04$).

Conclusion:

Although abnormal imaging findings involving the globes, optic nerves, and sella turcica are more common in IIH patients with TSS than in CVT patients, these findings are not specific for IIH and are found in patients with raised ICP from other causes such as CVT, along with other signs suggestive of raised ICP.

References:

1. Yuh WT, Zhu M, Taoka T, et al. MR imaging of pituitary morphology in idiopathic intracranial hypertension. *J Magn Reson Imaging* 2000; 12: 808-813
2. Degnan AJ, Levy LM. Pseudotumor cerebri: brief review of clinical syndrome and imaging findings. *AJNR Am J Neuroradiol* June 16, 2011. Epub ahead of print.
3. Rohr AC, Riedel C, Fruehauf MC, et al. MR imaging findings in patients with secondary intracranial hypertension. *AJNR Am J Neuroradiol* 2011; 32: 1021-1029.

Key Words: idiopathic intracranial hypertension, cerebral vein thrombosis, magnetic resonance imaging

Financial Disclosure: The authors had no disclosures.

Poster 19

Does Bilateral Transverse Cerebral Venous Sinus Stenosis (TSS) Really Exist In Patients Without Increased Intracranial Pressure (ICP)?

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Introduction:

TSS is common among patients with elevated ICP, particularly those with idiopathic intracranial hypertension. Although it is believed that TSS exists in the general population, and is not specific to elevated ICP, the prevalence of TSS among individuals with normal ICP is unclear. The goal of this study was to identify patients with asymptomatic TSS and normal ICP and describe their characteristics.

Methods:

Among 240 adult patients who underwent standardized brain MRI/MRV with contrast at our institution between 09/2009 and 09/2011, 50 had TSS. Medical records were reviewed for symptoms of increased ICP, evidence of papilledema, CSF constituents and opening pressure (OP) and reason for brain imaging. We identified 5/50 (10%) well-documented patients without papilledema and CSF-OP ≤ 25 cmH₂O, and 2/50 without measured ICP, but who had no papilledema or other symptoms of increased ICP. Imaging was re-interpreted by a neuroradiologist to assess for signs suggestive of elevated ICP and to characterize the stenoses further.

Results:

All patients were women (5 African-American, 2 Caucasian, mean age: 41, mean BMI: 37.1). CSF contents were normal but OP were at the upper limit of normal (22, 23, 24, 24, 25cm H₂O). Indications for MRI/MRV included pituitary abnormality (1), migraine (4), and anomalous-appearing optic nerves (2). All had bilateral TSS. Six had a short TSS and empty sella; one had a long TSS but no empty sella; one had flattening of the globes; two had dilated optic nerve sheaths. None had bony changes.

Conclusion:

Asymptomatic bilateral TSS does exist in patients with ICP ≤ 25 cmH₂O, but is likely uncommon. Interestingly, CSF-OP was at the upper limit of normal in our patients, who also had other radiologic signs suggestive (but not specific) of chronically-raised ICP. Findings of bilateral TSS on brain imaging should probably prompt further investigations for raised ICP.

References:

1. Farb RI, Vanek I, Scott JN, Mikulis DJ, Willinsky RA et al. Idiopathic intracranial hypertension: the prevalence and morphology of sinovenous stenosis. *Neurology* 2003; 60: 1418-1424.
2. Bono F, Lupo MR, Lavano A, Mangone L, Fera F, et al. Cerebral MR venography of transverse sinuses in subjects with normal CSF pressure. *Neurology* 2003; 61: 1267-1270.
3. Degnan AJ, Levy LM. Pseudotumor cerebri: brief review of clinical syndrome and imaging findings [published online ahead of print June 16, 2011]. *American Journal of Neuroradiology*. www.ajnr.org. Accessed October 17, 2011.

Key Words: Cerebral venous sinus stenosis, Intracranial pressure, Neuroimaging

Financial Disclosure: The authors had no disclosures.

Poster 20

Obstructive Sleep Apnea In Idiopathic Intracranial Hypertension: Comparison With Matched Population Data

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Introduction:

Patients with idiopathic intracranial hypertension (IIH) frequently have coexisting obstructive sleep apnea (OSA). However, it is unclear if the prevalence and severity of OSA is greater in IIH patients than would be expected given their other risk factors for OSA (e.g., obesity).

Methods:

We included patients with a new diagnosis of IIH who satisfied modified Dandy criteria and underwent overnight polysomnography. We collected demographic data, anthropometric data, and the apnea-hypopnea index (AHI) from polysomnography. We also calculated the expected AHI for our patients, based on their age, sex, race, BMI, and menopausal status, using a model derived from 1741 randomly-sampled members of the general population with overnight in-lab polysomnography scored using similar criteria. We compared the obtained AHI values to those predicted by the model using a paired t-test. Our study had 80% power to detect a 10 unit change in AHI at $\alpha=0.05$.

Results:

We included 24 IIH patients, of which 20 were female (83.3%), 13 were Caucasian (54.2%), and 11 were African-American (45.8%). Patient age ranged from 16-54 yrs (median = 32 yrs; mean \pm SD = 32 \pm 10 yrs). Body mass index ranged from 27.3-45.9 (median = 39.5; mean \pm SD = 38.1 \pm 5.3). AHI from the sleep studies ranged from 0-63.3 (median = 3.3; mean \pm SD = 9.4 \pm 15.8). Predicted AHI ranged from 2.1-14.5 (median = 5.4; mean \pm SD = 5.9 \pm 3.0). The AHIs from the sleep studies were not significantly different from those predicted by the population-derived model ($p=0.14$).

Conclusion:

The prevalence and severity of OSA in IIH patients is no more than would be expected for their age, sex, race, and BMI. It remains unclear if the presence or treatment of coexisting OSA influences the clinical course of IIH.

References:

1. Bixler EO, Vgontzas AN, Ten Have T, Tyson K, Kales A. Effects of age on sleep apnea in men: I. Prevalence and severity. *Am J Respir Crit Care Med* 1998; 157: 144-148.
2. Bixler EO, Vgontzas AN, Lin HM, Ten Have T, Rein J, Vela-Bueno A, Kales A. Prevalence of sleep-disordered breathing in women: effects of gender. *Am J Respir Crit Care Med* 2001; 163: 608-613.
3. Lee AG, Golnik K, Kardon R, Wall M, Eggenberger E, Yedavally S. Sleep apnea and intracranial hypertension in men. *Ophthalmology* 2002; 109: 482-485.
4. Marcus DM, Lynn J, Miller JJ, Chaudhary O, Thomas D, Chaudhary B. Sleep disorders: a risk factor for pseudotumor cerebri? *J Neuroophthalmol* 2001; 21: 121-123.
5. Thurtell MJ, Bruce BB, Rye DB, Newman NJ, Biousse V. The Berlin questionnaire screens for obstructive sleep apnea in idiopathic intracranial hypertension. *J Neuroophthalmol*; doi:10.1097/WNO.0b013e31821a4d54
6. Wall M, Purvin V. Idiopathic intracranial hypertension in men and the relationship to sleep apnea. *Neurology* 2009; 72: 300-301.

Key Words: Idiopathic Intracranial Hypertension, Obstructive Sleep Apnea

Financial Disclosure: The authors had no disclosures.

Poster 21

Emergency and Inpatient Services Utilization in Idiopathic Intracranial Hypertension

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Introduction:

Patients with IIH may seek care in the emergency department (ED) and sometimes require hospitalization. The magnitude of ED and inpatient use is uncertain; nationwide data collection regarding hospital-based visits uses the ICD-9 code for IIH (348.2) which has never been validated.

Methods:

We reviewed ED and inpatient encounters from patient (ages ≥ 18) charts coded with a diagnosis of IIH, optic nerve sheath fenestration, or lumboperitoneal shunting. We recorded patient demographics, reason for the encounter, diagnostic testing and procedures performed, and hospital charges. We calculated the positive predictive value (PPV) of the ICD-9 code for IIH.

Results:

137 visits from 51 patients were identified. The majority of encounters occurred in the ED (68%), and 40% of those converted into inpatient admissions. 94% of patients were seen in the ED at least once. The use of ED services increased from <5 /year from 2001-2003 to >10 /year in 2007-2010. Encounters resulted from recurrent IIH symptoms (43%), surgical complications (26%) and initial presentation (12%). The PPV of the ICD-9 code was 50%. Patients averaged nearly 3 visits each over the 10 year study period.

Conclusion:

The use of the ED by patients for IIH for IIH-related issues increased over the 10-year study period. This may be attributable to the increasing rates of obesity in the population, heightened awareness of the disorder by medical professionals, relative ease of ED access compared to outpatient care, insurance coverage, and increasing use of shunt procedures, which often require hospitalization (1). Consistent with previous reports, surgical complications were common, accounting for 26% of hospital admissions from the ED. The 348.2 diagnosis code may not reliably capture all IIH-related services (2).

References:

Curry WT Jr, Butler WE, Barker FG 2nd. Rapidly rising incidence of cerebrospinal fluid shunting procedures for idiopathic intracranial hypertension in the United States, 1988-2002. *Neurosurgery* 57(1):97-108, 2005. McGirt MJ, Woodworth G, Thomas G, Et al Cerebrospinal Fluid Shunt Placement for Pseudotumor Cerebri-Associated Intractable Headache: Predictors of Treatment Response and an Analysis of Long-Term Outcomes. *J. Neurosurg* 101(4): 627-632, 2004

Key Words: idiopathic intracranial hypertension, emergency department, medical coding

Financial Disclosure: Deborah Friedman, MD, MPH (disclosure not related to abstract content) - Grant support from the National Eye Institute, Merck, Pfizer, MAP Pharmaceuticals, Amplatz, Quark Pharmaceuticals. Speaker for Allergan. Advisory board of MAP Pharmaceuticals and Merck. The remaining author has no disclosures.

Poster 22

Comparison of Optic Nerve Head Parameters using Optical Coherence Tomography to Evaluate Papilledema in Idiopathic Intracranial Hypertension

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Introduction:

Optical coherence tomography (OCT) has begun to be utilized as an adjunctive, quantitative tool in the evaluation of patients with idiopathic intracranial hypertension (IIH) to help monitor response to treatment. Focus has primarily been upon the anatomic relationship of retinal nerve fiber layer (RNFL) thickness and its correlation with the clinical appearance of optic nerve edema.¹ We sought to explore whether alternative optic nerve parameters could show a greater correlation with subjective clinical changes at the optic nerve head to determine the sensitivity of OCT to treatment failure that may prompt an alteration in management.

Methods:

OCT data obtained using spectral domain OCT (Heidelberg Spectralis) for patients with the diagnosis of IIH over a two year study period were retrospectively reviewed. Optic nerve parameters including maximal optic nerve height, optic nerve total thickness (measured at diameters 1, 2, and 3 mm), and RNFL thickness were recorded.

Results:

Both eyes of 41 patients were included. As analyzed by Spearman rank correlation, there was a strong correlation between each of the parameters studied in relation to mean RNFL thickness. Versus maximal optic nerve height, the Spearman rank correlation was 0.647 ($P = <0.0001$), versus central total optic nerve thickness, the correlation was 0.629 ($P = <0.0001$), versus inner circle total optic nerve thickness, the correlation was 0.789 ($P = <0.0001$), versus outer circle optic nerve thickness, the correlation was 0.828 ($P = <0.0001$).

Conclusion:

With spectral domain OCT, parameters of optic nerve imaging, including maximal optic nerve height and total optic nerve head thickness measurements, show a strong correlation with traditional evaluation of RNFL thickness. These alternative measures might provide greater sensitivity of the response to treatment in patients with IIH as they may better correspond with the clinical optic nerve head appearance. Additional studies are needed to determine which parameter is most reflective of the functional impact of optic nerve edema on vision in patients with IIH.

References:

1. Scott CJ, Kardon RH, Lee AG, Frisen L, Wall M. Diagnosis and grading of papilledema in patients with raised intracranial pressure using optical coherence tomography vs clinical expert assessment using a clinical staging scale. *Arch. Ophthalmol.* 128, 705-711, 2010.

Key Words: Idiopathic intracranial hypertension, Optical coherence tomography, Papilledema

Financial Disclosure: The authors had no disclosures.

Poster 23

Familial Pseudotumor Cerebri - Case Series

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Introduction:

Pseudotumor cerebri (PTC) is a disorder of unknown etiology. Few reports have described its occurrence amongst families.

Methods:

The medical records of all patients diagnosed with PTC in our institution were reviewed to identify familial cases and study their phenotype.

Results:

Eleven patients from 5 different families were found within a cohort of 440 PTC patients. The relation was mother and daughters in 2 families, brothers and sisters in 2 families and in one family it was an aunt and two first degree cousins. The symptoms, course of disease and risk factors were similar between family relatives of all 5 families, except the age of diagnosis which was different in 1 family. All adult patients were mildly obese (BMI 25-35) except in one family in which all members had morbid obesity.

Conclusion:

Although familial PTC is very uncommon, its occurrence within families may justify evaluation of asymptomatic first degree relatives of a PTC patient. Further evaluation of these cases may yield useful information about the pathogenesis and the genetic bases of this disorder.

References:

1. Corbett, James J MD, Familial Idiopathic Intracranial Hypertension, Journal of Neuro-Ophthalmology, Dec2008, Volume 28(4), pages 337-347
2. Gardner K, Cox T, Digre KB, Idiopathic intracranial hypertension associated with tetracycline use in fraternal twins: case reports and review., Neurology, 1995 Jan;45(1), pages 6-10
3. Kesler A, Goldhammer Y, Gadoth N. Do men with pseudomotor cerebri share the same characteristics as women? A retrospective review of 141 cases, J Neuroophthalmol 2001, volume1, pages15-7

Key Words: Pseudotumor Cerebri, Familial, Idiopathic Intracranial Pressure

Financial Disclosure: The authors had no disclosures.

Poster 24

A Retrospective Study Of Idiopathic Intracranial Hypertension and Metabolic Syndrome: Do They Co-Exist?

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Introduction:

To determine whether there is an association between Idiopathic Intracranial Hypertension (IIH) and Metabolic Syndrome (MS).

Methods:

A retrospective review was performed of all patients with a diagnosis of IIH seen between July 2010 to October 2011. The primary outcome measure was presence or absence of MS as defined by the American Heart Association and the National Heart, Lung and Blood Institute.

Results:

30 female patients were included, 22 (73%) of which met the criteria for MS. Mean age at diagnosis of the MS group was 39.4 years versus 28.5 years in the non MS group ($p=.0030$). A significant difference was found between triglyceride level, fasting blood glucose, hemoglobin A1c, systolic blood pressure, and diastolic blood pressure ($p= 0.018, 0.0036, 0.021, 0.0006, 0.0043$, respectively) of the MS and non MS patients. Body mass index, waist circumference, opening pressure, and high density lipoprotein level were not significantly different between the two groups.

Conclusion:

A high percentage of patients with IIH also have MS, suggesting an association. Patients with IIH and MS had higher age at diagnosis, triglyceride levels, fasting blood glucose, hemoglobin A1c, and systolic and diastolic blood pressure compared to patients without MS. Whether MS is a co-morbidity of IIH or plays a role in the pathogenesis of IIH remains to be investigated.

References: None

Key Words: idiopathic intracranial hypertension, pseudotumor cerebri, metabolic syndrome

Financial Disclosure: The authors had no disclosures.

Poster 25

JAK2^{V617F} Mutation Associated with “Pseudotumor Cerebri”

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Introduction:

Myeloproliferative disorders are known to be associated with the high risk of major venous thrombosis. The International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT) data showed that overt MPDs were responsible for 2.8% of cerebral venous thrombosis (CVT) cases. The JAK2^{V617F} mutation was found in more than 90% of Polycythemia Vera and 50% of Essential Thrombocytemia patients. We describe two cases of CVT with papilledema, one with overt, the other with latent MPD, diagnosed by presence of the JAK2^{V617F}. The few cases reported in the literature have diverse prognosis but none elaborate neuro-ophthalmic or neuro-radiologic findings.

Methods:

Case reports.

Results:

Case 1: An 11-year-old girl known to have thrombocytosis with JAK2^{V617F} mutation. A routine eye examination indicated bilateral disc edema. CT was normal; intracranial pressure was 50cm H₂O on lumbar puncture. She was treated with acetazolamide and steroids but continued to experience headaches and vision deterioration. Follow-up CT venography diagnosed an extensive CVT. She underwent optic nerve sheath fenestration and VP shunt due to the visual deterioration.

Case 2: A 22-year-old male complained of headaches and double vision two weeks after a motor vehicle accident. He was found to have optic disc edema, CVT on CT venography and elevated intracranial pressure of 50cm H₂O. Hemoglobin levels and platelet count were elevated, with positive JAK2^{V617F} mutation. Despite maximal medical treatment, his vision deteriorated. Hence, he underwent optic nerve sheath fenestration and LP shunt.

Conclusion:

These are the first case reports elaborating on neuro-ophthalmic findings and course of the disease among young patients with a JAK2^{V617F} mutation and CVT induced papilledema together with severe visual compromise. Despite early medical treatment, visual prognosis was grave in both cases with worsening thrombosis on imaging, probably due to hemo-concentration caused by the acetazolamide treatment.

References: None

Key Words: Pseudotumor cerebri, Papilledema, Cerebral venous thrombosis, Myeloproliferative disorders, Jak2 mutation

Financial Disclosure: The authors had no disclosures.

Poster 26

Correlation Between Visual Parameters And Neuroimaging Features Of Papilledema

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Introduction:

Papilledema refers to optic disc swelling resulting from high intracranial pressure. The precise mechanism by which papilledema occurs remains uncertain. Although orbital neuroimaging features associated with papilledema are well-described, it is unclear whether these findings correlate with visual function. Idiopathic Intracranial Hypertension (IIH) is a condition in which the intracranial pressure is elevated with no obvious cause, causing papilledema and visual loss. The utility of papilledema neuroimaging findings as a surrogate marker for visual loss, or a predictor of visual loss, is understudied. This retrospective cross-sectional review aims to correlate parameters of visual function with orbital magnetic resonance imaging (MRI) findings.

Methods:

Patients meeting criteria for IIH who had received orbital imaging within 4 weeks of examination were included. Visual parameters of papilledema grade, visual field mean deviation, visual acuity, and color vision were correlated with specific neuroimaging features, including optic nerve sheath thickness and globe configuration, among others. All MRI scans were reviewed by a neuroradiologist blinded to clinical status. Kendall's Tau Rank correlations and t-tests were generated with SAS (v9.2).

Results:

Thirty five patients were included (34 female, 1 male), with mean age 28.9 (\pm 8.2) years. No significant relationships were found between the main visual parameters of papilledema grade and visual field mean deviation, and MRI findings. Further analysis comparing severe (papilledema grades 4-5) with non-severe (papilledema grades 0-3) patients found a modest difference in optic nerve thickness.

Conclusion:

We found no significant correlation between visual parameters and imaging features of papilledema, aside from a modest relationship between severity of papilledema and nerve thickness. MRI features may not be a valid surrogate marker of visual loss in patients with IIH in particular, and papilledema in general. Although our sample size was low, we believe the low correlations between the main visual parameters are valid, and non-artifactual.

References:

1. Brodsky MC, Vaphiades M. Magnetic resonance imaging in pseudotumor cerebri. *Ophthalmology* 1998;105:1686-1693.
2. Mashima Y, Oshitari K, Imamura Y, Momoshima S, Shiga H, Oguchi Y. High-resolution magnetic resonance imaging of the intraorbital optic nerve and subarachnoid space in patients with papilledema and optic atrophy. *Arch Ophthalmol* 1996;114:1197-1203.
3. Agid R, Farb RI, Willinsky RA, Mikulis DJ, Tomlinson G. Idiopathic intracranial hypertension: the validity of cross-sectional neuroimaging signs. *Neuroradiology* 2006;48:521-527.

Key Words: Papilledema, Idiopathic Intracranial Hypertension, Neuroimaging, Visual loss

Financial Disclosure: The authors had no disclosures.

Poster 27

Evaluation of Optic Nerve Head Elevation in Idiopathic Intracranial Hypertension with 3-Dimensional Optical Coherence Tomography

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Introduction:

Optical coherence tomography (OCT) is useful in the evaluation of papilledema and resulting changes in the retinal nerve fiber layer. However, the relationship between optic nerve head (ONH) elevation or volume measured with 3-Dimensional OCT and visual field recovery is unknown. In this study, we compared the reliability of two spectral-domain OCTs in assessing ONH elevation and volume, and correlated their findings to visual field recovery in six patients with papilledema due to Idiopathic Intracranial Hypertension (IIH).

Methods:

Six patients with IIH underwent automated perimetry at diagnosis and after standard of care treatment. Mean deviation scores were calculated for the superior and inferior hemifields. Volumetric OCT cubes of the ONH and peripapillary retina were obtained with either the Cirrus (Carl Zeiss Meditech) or Spectralis (Heidelberg) OCT. Superior and inferior ONH elevation and/or volume were manually measured by two independent raters.

Results:

Inter-rater reliability between ONH elevation and volume on both spectral-domain OCTs was obtained with the Pearson r coefficient. For the Cirrus OCT, $r = 0.828$ and for the Spectralis OCT, $r = 0.991$. Using data from the most reliable OCT, we compared peak ONH elevation, mean ONH elevation and ONH volume with visual field recovery in the corresponding hemifield using linear regression analysis. In both superior and inferior hemifields, an inverse relationship was observed between each ONH measurement and the percentage of visual field recovery, with R^2 values for the superior hemifield of 0.59, 0.51, 0.51, respectively, and for the inferior hemifield of 0.42, 0.47, 0.47, respectively.

Conclusion:

In IIH patients, ONH elevation and volume on presentation correlate with the degree of visual field recovery after treatment. OCT-determined ONH elevation and volume measurements are reliable and may have value in predicting recovery of vision in IIH and in guiding medical management in the future.

References:

1. Heidary G., Rizzo J.F., Use of Optical Coherence Tomography to Evaluate Papilledema and Pseudopapilledema, *Semin Ophthalmol*, 25:198-205, 2010
2. Rebolleda G., Munoz-Negrete F.J., Follow-up of Mild Papilledema in Idiopathic Intracranial Hypertension with Optical Coherence Tomography, *Invest Ophthalmol Vis Sci*, 50:5197-5200, 2009
3. Sanchez-Tocino H., Bringas R., Iglesias D., et al, Utility of Optic Coherence Tomography in the Follow-up of Idiopathic Intracranial Hypertension in Childhood, *Arch Soc Esp Oftalmol*, 81:383-390, 2006
4. Skau M., Sander B., Milea D., et al, Disease Activity in Idiopathic Intracranial Hypertension: a 3-Month Follow-up Study, *J Neurol*, 258:227-283, 2011

Key Words: Idiopathic intracranial hypertension, Papilledema, Visual field recovery, Optical Coherence Tomography, Optic nerve head volume

Financial Disclosure: Marc Dinkin - on an advisory board for Acorda pharmaceuticals. Nathan Radcliffe -Consultant/Speaker for Allergan, Inc., Alcon, Merck & Co., Ophthalmic Imaging Systems, Carl Zeiss Meditec, Iridex. The remaining authors had no disclosures.

Poster 28

Identifying Trends In The Treatment And Outcomes Of Patients With Idiopathic Intracranial Hypertension (IIH)

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Introduction:

Idiopathic Intracranial Hypertension (IIH) is characterized by increased cerebrospinal fluid (CSF) pressure in the absence of common causes of intracranial hypertension. Although management strategies of IIH are well-documented^{1,2}, there is currently no published, comprehensive research that conclusively outlines the best treatment options for IIH. This study aimed to collect information from the first and largest patient registry for intracranial hypertension and identify criteria that physicians are using to make treatment decisions.

Methods:

Inclusion criteria were a confirmed diagnosis of IIH using Dandy Criteria and follow-up time of 5-10 years. Seven indicators were selected to classify patients as either grade I, II, or III in severity at presentation. These include: logMAR measures of visual acuity, visual field, papilledema (Frisen grade), BMI, lumbar puncture opening pressure, systemic hypertension, and "other comorbidities" in the relevant medical history. Patients were assigned a severity grade and followed through time to identify outcomes, which included average change in visual acuity/field, dosage requirements of acetazolamide, percentage receiving shunt or ONSF surgery, and average time to shunt/ONSF. The training (neurology v. ophthalmology v. neuro-ophthalmology) of the primary physician was identified to determine how this affected treatment and outcomes.

Results:

IRB approval for this study has been received and access to the patient files and physician surveys is anticipated in early November. Analysis will be conducted throughout the following month.

Conclusion:

A previous survey sent to physicians by the authors of this study indicated that there is significant variation in the treatment of IIH based on physician training. These results will most likely be confirmed in a more objective way using this national patient registry. It is also anticipated that there will be variations in the care of IIH patients based on the perceived severity of disease at presentation. It is possible that visual outcomes are affected by these variations in treatment.

References:

1. Dhungana, Sharrack, Woodroffe, Review Article: Idiopathic Intracranial Hypertension, *Acta Neurologica Scandinavica*, 121, 71-82, 2010
2. Digre, Idiopathic Intracranial Hypertension, *Current Treatment Options in Neurology*, 1, 74-81, 1999

Key Words: Idiopathic Intracranial Hypertension (IIH), severity, treatment variation, physician training, outcomes

Financial Disclosure: The authors had no disclosures.

Poster 29

Aetiology and clinical features of intracranial hypertension mainly presented as papilledema.

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Introduction:

Intracranial hypertension mainly presented as papilledema and related visual disturbance is easily diagnosed as idiopathic intracranial hypertension (IIH). We conducted this study to investigate the causes and clinical features of intracranial hypertension patients mainly presented as papilledema (IHPP) in China.

Methods:

Medical records of patients with IHPP were reviewed. Clinical features and imaging data were retrospectively analysed and etiological diagnosis were determined according to the relevant diagnosis criteria

Results:

In totally 174 cases included, 127(73.0%) met the diagnostic criteria of idiopathic intracranial hypertension (IIH). Twenty cases (11.5%) met a strict criteria of cerebral venous sinus thrombosis (CVST). Other causes included benign intracranial mass for 7 cases, dural arteriovenous fistula (DAVF) for 6 cases, surgery related complication for 6 cases and other causes of single case including castleman disease, malignant lymphoma, meningeal carcinomatosis, churg-strauss syndrome, cysticercosis, Arnold-Chiari malformation. Two cases were classified as uncertain intracranial infection. Female cases (96/127, 75.6%) and overweight (73.4%) were common in IIH group while male predominance (13/20, 65%) was seen in CVST group. Isolated lateral sinus thrombolysis was found in 13 of 20 (65%) CVST patients.

Conclusion:

IIH is the most common cause in this group of IHPP patients. Overweighed female was common in IIH group. CVST was the second commonest cause, of which isolated lateral sinus thrombosis is most common. Even rarely, some malignant entity can be the cause of intracranial hypertension mainly presented as papilledema.

References:

1. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*, 2002,59(10):1492-5.
2. Bousser MG. Cerebral venous thrombosis: diagnosis and management. *J Neurol*, 2000,247(4):252-8.
3. Bousser MG, Ferro JM. Cerebral venous thrombosis: an update. *Lancet Neurol*, 2007,6(2):162-70.

Key Words: intracranial hypertension, idiopathic, papilledema, cerebral venous sinus thrombosis, cause

Financial Disclosure: The authors had no disclosures.

Poster 30

Pseudotumor cerebri syndrome associated with giant arachnoid granulation.

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Introduction:

We report a rare case of pseudotumor cerebri (PTC) associated with a giant arachnoid granulation (GAG) in the transverse venous sinus.

Methods:

Single case report describing presentation, work-up and management.

Results:

A 26 year old healthy male complained of horizontal diplopia for 1 month. Visual acuity was 20/25 OU. EOM revealed bilateral sixth nerve palsies. Funduscopy showed bilateral optic disc edema suspicious for papilledema and HVF testing revealed enlarged blind spots OU. CT revealed a hypodense filling defect at the origin of the right transverse venous sinus, and MRI/MRV revealed a 3.5 cm filling defect, isointense to cerebrospinal fluid.



The diagnosis of GAG was made. Lumbar puncture revealed opening pressure of 56 cm H₂O with normal CSF composition. The patient was treated with acetazolamide. Despite titrating doses of acetazolamide, the patient's diplopia persisted and he developed severe headaches. Fundoscopy revealed worsening papilledema with peripapillary retinal striae. Because of the patient's refractory symptoms and papilledema, surgical intervention via an endovascular approach is planned with the goal of either draining or stenting the GAG. The outcome of the intervention is pending.

Conclusion:

Giant arachnoid granulations are large growths of arachnoid membrane into the dural sinus, which serve to filter CSF across the lining of the arachnoid cells to drain into the cerebral venous system. Though usually asymptomatic, GAGs have been reported in association with increased intracranial pressure and PTC in only a handful of cases. Whether GAGs are a cause or consequence of PTC remains to be determined, as is the ideal management of these rare cases.

References:

1. Choi, HJ, Cho, CW, Kim, YS, Cha, JH, Giant arachnoid granulation misdiagnosed as transverse sinus thrombosis, *Journal of Korean Neurosurgical Society*, 43, 48-50, 2008.
2. Peters, SA, Frombach, E, Heyer, CM, Giant arachnoid granulation: differential diagnosis of acute headache, *Australian Radiology*, 51, B18-B20, 2007.
3. Arjona, A, Delgado, F, Fernandez-Romero, E, Intracranial hypertension secondary to giant arachnoid granulation, *Journal of neurology, neurosurgery, and psychiatry*, 74, 418, 2003.
4. Zheng, Z, Zhou, M, Zhao, B, Zhou, D, He, L, Pseudotumor cerebri syndrome and giant arachnoid granulation: treatment with venous sinus stenting, *Journal of Vascular Interventional Radiology*, 21, 927-929, 2010.

Key Words: Giant Arachnoid Granulation, Papilledema, Neuroradiology

Financial Disclosure: The authors had no disclosures.

Poster 31

Relapsing Dorsal Midbrain Syndrome Following Interventions for Hydrocephalus in Aqueductal Stenosis

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Introduction:

Dorsal midbrain syndrome (DMS) is a recognized clinical manifestation of increased intracranial pressure (ICP) associated with ventricular enlargement, especially in shunt malfunction. The mechanism by which DMS occurs in this setting is unsettled. (1-3) It cannot simply be the result of high ICP because most patients with high ICP do not get DMS. We report a patient with triventriculomegaly attributed to aqueductal narrowing by a tectal mass who twice developed and resolved DMS promptly after undergoing interventions that altered the size of the posterior third ventricle and proximal aqueduct but probably did not markedly alter ICP. We offer this case as evidence that deformation of these structures causes DMS.

Methods:

This is a single case report of the neuro-ophthalmic and imaging documentation of a 9-year-old girl who underwent serial procedures for hydrocephalus and subdural fluid accumulations.

Results:

The patient was observed to develop recurrent DMS as neurosurgical procedures enlarged the size of the posterior third ventricle and proximal aqueduct but did not alter intracranial pressure. The DMS resolved promptly when procedures caused these structures to return to more normal size.

Conclusion:

Two previous publications (4, 5) have documented reversal of dorsal midbrain syndrome by ventriculocisternostomy or third ventriculostomy. In those cases, the procedures reduced intracranial pressure. Our report is the first to show cycles of development and regression of DMS following procedures that probably *did not alter intracranial pressure*, but did alter the configuration of the posterior third ventricle and proximal aqueduct. It provides evidence that deformation of these structures causes DMS in the setting of hydrocephalus.

References:

1. Baloh RW, Furman JM, Yee RD. Dorsal midbrain syndrome: clinical and oculographic findings. *Neurology* 1985;35:54-60.
2. Chattha AS, DeLong GR. Sylvian aqueduct syndrome as a sign of acute obstructive hydrocephalus in children. *J Neurol Neurosurg Psychiatry* 1975;38:288-96.
3. Chou SY, Digre KB. Neuro-Ophthalmic complications of raised intracranial pressure, hydrocephalus, and shunt malfunction. *Neurosurg Clin N Am* 1999;10:587-608.
4. Lerner MA, Kosary IZ, Cohen BE. Parinaud's syndrome in aqueduct stenosis: its mechanism and ventriculographic features. *Br J Radiol* 1969;42:310-2.
5. Cinalli G, Sainte-Rose C, Simon I, et al. Sylvian aqueduct syndrome and global rostral midbrain dysfunction associated with shunt malfunction. *J Neurosurg* 1999;90:227-36.

Key Words: Aqueductal stenosis, Intracranial hypertension, Dorsal Midbrain Syndrome, Third ventricle, Hydrocephalus

Financial Disclosure: The authors had no disclosures.

Poster 32

Microvascular Cranial Nerve Palsies: Risk Factor Analysis

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Introduction:

An isolated cranial nerve palsy is commonly attributed to microvascular ischemia when occurring in the setting of vascular disease. Common vasculopathic risk factors include hypertension, diabetes, dyslipidemia, and advanced age. This project sought to study the prevalence of poor control of vasculopathic risk factors in patients who have been diagnosed with a cranial nerve palsy of a microvascular etiology.

Methods:

A retrospective chart review of all patients who were diagnosed with a palsy of cranial nerve III, IV, and VI whose symptoms resolved and events are attributed specifically to a microvascular event. Data that was obtained and compared were blood pressure, blood sugar, HgbA1C, Cholesterol Panel, Triglycerides.

Results:

To date twenty-one of the forty patients enrolled have complete data, palsies identified: CN3 (7 patients) CN4 (2 patients), CN6 (12 patients). The demographics of the group were as follows: 57% Male, 43% Female; Average Age 61.4±10 years; Race 57% Hispanic, 14% Black, 19% White, 10% Other. Comorbidities: 76% Hypertension 76%, Diabetes 71.4%, Dyslipidemia 52%. Ninety percent of the patients studied had at least one uncontrolled vasculopathic risk factor as defined by National Institute of Health standards: Uncontrolled hypertension 42%, uncontrolled diabetes 38%, uncontrolled lipid profile 52.8%, more than one uncontrolled risk factor 47.6%.

Conclusion:

Our preliminary results strongly suggest that the overwhelming majority of these patients who undergo an ischemic cranial nerve palsy, have at least one poorly controlled risk factor. Management of these risk factors should be a priority. Perhaps these results can further stratify which patients we can safely identify cranial nerve palsies as ischemic versus those that need immediate imaging.

References: None

Key Words: cranial nerve palsy, vasculopathy, diplopia, diabetes, hypertension

Financial Disclosure: The authors had no disclosures.

Poster 33

Photographic assessment of optic disc and retinal vasculature in obstructive sleep apnea (OSA) patients.

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Introduction:

Obstructive sleep apnea (OSA) alters cerebral vascular reactivity, possibly mediated via hypercapnea. While OSA has been associated with anterior ischemic optic neuropathy, glaucoma, and raised intracranial pressure (ICP), the potentially detrimental effects upon optic nerve and retinal microvasculature health have not been comprehensively addressed. We evaluated optic nerve and retinal vasculature appearance prospectively in patients undergoing routine, diagnostic polysomnography (dPSG).

Methods:

Non-mydratic fundus photography of patients undergoing overnight polysomnography was performed. Demographics and BMI were documented. A prospective questionnaire exploring medical/ocular history, increased ICP symptoms, headache, and sleepiness (including HIT6 & Berlin questionnaire) was administered. Fundus photographs were assessed for abnormalities. Fractal dimension and lacunarity (ImageJ/FracLac) of the retinal vascular tree were determined. Apnea-hypopnea index (AHI) and hypoxic burden were the primary polysomnography measures. OSA was defined as AHI>15.

Results:

Of 180 patients undergoing dPSG, fundus photographs were performed on 155 patients (exclusions: 20 refusals, 3 unable to give consent, 2 known hydrocephalus). Demographics: mean age 58 years, 84M 71F, 59% Caucasian, 35% African-American. 23 patients had severe OSA (AHI>20, hypoxic burden>10%), and 63 moderate/mild OSA. The remaining 69 patients with other sleep diagnoses became the control population. 239 photographs were taken from the 155 patients before, during and after a night of sleep. There was no difference between OSA and non-OSA patients with respect to age, sex, ethnicity and headache phenotype ($p<0.05$). OSA patients had a significantly greater BMI (34 versus 30; $p=0.02$). No patients had symptoms of increased ICP. No evidence of disc edema or vascular changes were observed in any patients (95%CI:0-4.5%). There was no association between vascular fractal measures and indices of OSA.

Conclusion:

There was no evidence of retinal vascular changes or optic disc edema to suggest subclinical increased ICP in our OSA population, even in those with severe OSA. Routine clinical fundoscopic screening of OSA patients appears unwarranted.

References:

1. Urbano F, Roux F, Schindler J, Mohsenin V. Impaired cerebral autoregulation in obstructive sleep apnea. *J Appl Physiol* 2008;105:1852-7.
2. Palombi K, Renard E, Levy P, et al. Non-arteritic anterior ischaemic optic neuropathy is nearly systematically associated with obstructive sleep apnoea. *Br J Ophthalmol* 2006;90:879-82.
3. Mojon DS, Hess CW, Goldblum D, et al. Normal-tension glaucoma is associated with sleep apnea syndrome. *Ophthalmologica* 2002;216:180-4.
4. Lim MJ and Lin JP. The effects of carbon dioxide on measuring cerebral spinal fluid pressure. *Childs Nerv Syst*, 2009. 25(7): 783-4
5. Liew G et al. Fractal analysis of retinal microvasculature and coronary heart disease mortality. *European heart journal*, 2011. 32(4):422-9.

Key Words: optic disc edema, retinal vasculature, fundus photography, obstructive sleep apnea, idiopathic intracranial hypertension

Financial Disclosure: Dr Donald Bliwise: Consultant, Ferring Pharmaceuticals. The remaining authors had no disclosures.

Poster 34

Retinal Nerve Fibre Layer Thickness in Paediatric Demyelinating Disease: The Canadian Paediatric Disease Study Group

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Introduction:

To compare the average retinal nerve fibre layer(RNFL) thicknesses of paediatric patients with demyelinating disease to age matched normal controls.

Methods:

RNFL at SD-OCT(Cirrus, Carl Zeiss) scanning 6/12 following a demyelinating episode. 35 paediatric patients (in the CPDDS) were compared to 20 age matched controls.

Eyes categorized as;

First episode of optic neuritis(ON) as a clinically isolated syndrome(CIS)

Recurrent ON

ON in the context of multiple sclerosis(MS)

Fellow eyes of ON patients(CIS or MS)

Asymptomatic eyes of CPDDS patients without clinical/radiological evidence of ON (MS/non-ophthalmic CIS)

Results:

RNFL significantly lower in eyes following ON CIS(88.5um) than normal controls(105um).

Recurrent ON: progressive deductive effect(64.4um =>2 episodes).

ON with MS: significantly greater reduction(72.9um).

Fellow eyes: RNFL (96.5um)

MS/CIS patients without clinical or radiological evidence of ON: RNFL(94.8um).

Conclusion:

Average RNFL thickness at 6 month SD-OCT scanning is significantly reduced following ON. Recurrent ON compounds RNFL loss. Fellow eyes of ON and eyes of MS/CIS without clinically or radiologically evident ON have reduced RNFL compared to normal controls.

References: None

Key Words: Pediatric, Demyelination, Retinal Nerve Fibre Layer, Cirrus OCT, National Study

Financial Disclosure: The authors had no disclosures.

Poster 35

Retinal Nerve Fibre Layer Thickness Thresholds for Visual Function in Pediatric Demyelinating Disease: Further evidence of the RNFL Tipping Point

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Introduction:

RNFL thresholds for visual function have been demonstrated in adult neuropathy ¹⁻².

Methods:

Average RNFL thickness at Cirrus,SD-OCT in 35 CPDDS patients and 20 normal controls were analysed in relation to the following visual function metrics;

High Contrast Visual Acuity(using PEDIG Amblyopia Visual Acuity Testing Protocol)

Low Contrast Sensitivity(Pelli-Robson)

Colour Vision(HRR)

Visual Field(Humphreys)

A 'Broken Stick' statistical model was applied to estimate the RNFL thickness 'tipping point' where visual function metrics become associated with structural measures

Results:

Threshold RNFL thickness for visual field function was found at 77um. Threshold RNFL thickness for low contrast sensitivity and colour vision appear to be higher. HCVA does not show a significant correlation.

Conclusion:

'Broken stick' analysis of RNFL thresholds for visual function metrics in pediatric demyelinating disease indicate that a 'tipping point' may exist. For visual fields this appears to be similar to that found in the adult population and in other neuropathies. However low contrast testing and colour testing indicate higher thresholds exist for different elements of vision.

References:

1. Costello F, Hodge W, Pan YI, Eggenberger E, Freedman MS. Using retinal architecture to help characterize multiple sclerosis patients. *Can J Ophthalmol*. 2010 Oct;45(5):520-6
2. Wollstein G., et al Retinal Nerve Fibre Layer and visual function loss in glaucoma: the tipping point. *BJO Online First* (April 8, 2011) 10.1136/bjo.2010.196907

Key Words: Demyelination, Pediatric, Tipping Point, RNFL Cirrus OCT, Function

Financial Disclosure: F Costello: had board compensation from Biogen Idec and Novartis. The remaining authors had no disclosures.

Poster 36

Idiopathic Dural and Optic Nerve Sheath Calcification – A Rare and Unusual cause of Papilledema

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Introduction:

Duro-optic calcification is a very rare condition characterized by calcification of optic nerve sheaths and duramater. Only 5 cases have been reported worldwide with various clinical manifestations. We report a patient with duro-optic calcification and papilledema.

Methods:

Case report

Results:

A 53 year-old woman presented with 3-month history of headaches, pulse synchronous tinnitus, horizontal diplopia and transient visual obscurations. Past medical history included use of lithium for mood disorder and meningitis at age 15. Examination revealed visual acuity of 20/25 OD and 20/30 OS, normal intraocular pressures, color vision, equal and reactive pupils with no afferent pupillary defect. Ocular motility demonstrated limited abduction OS. Automated perimetry (Humphrey) revealed an enlarged blind spot with generalized depression and constriction of visual fields in both eyes. Fundoscopy demonstrated severe bilateral papilledema with peripapillary hemorrhages and tortuosity of blood vessels. Optical coherence tomography showed marked thickening of the retinal nerve fiber layer in both eyes. B-scan ultrasonography showed bilateral buried optic disc drusen. CT head revealed patchy calcification of optic nerve sheaths anteriorly and the duramater, predominantly involving the falx cerebri, superior sagittal sinus, straight sinus and vein of Galen. CT Venogram did not reveal cerebral venous sinus thrombosis or stenosis. Lumbar puncture revealed opening pressure of 25 cm of water with normal cerebrospinal constituents. Work up for inflammatory causes, vasculitis, connective tissue disorders, infections, metabolic and malignancy were negative. The exact cause of increased intracranial symptoms remains unknown. She was started on acetazolamide 500 mg twice a day but developed intolerable side effects. The intracranial symptoms and papilledema showed minimal improvement and she continued to have chronic daily headaches. She awaits a neurosurgical consultation for potential shunting procedure.

Conclusion:

Our report highlights a rare condition of idiopathic duro-optic calcification as a potential cause of increased intracranial pressure and papilledema.

References:

1. Phadke RV, Agarwal P, Sharma K, et al. Idiopathic duro-optic calcification - a new entity? Clin Radiol 1996; 51:359–61.
2. Nicholson BP, Lystad LD, Singh AD. Idiopathic Dural Optic Nerve Sheath Calcification. Br J Ophthalmol 2011;95:290

Key Words: Duro-optic calcification, Intracranial hypertension, Headache, Papilledema

Financial Disclosure: The authors had no disclosures.

Poster 37

Benign chiasmal glioma of adulthood

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Introduction:

Optic pathway gliomas in adults are usually malignant and represent a distinct clinical entity from the more common childhood benign optic glioma. Clinically, they are characterized by rapid loss of vision leading to blindness and ultimately death. We report two middle-aged adults who presented with chiasmal gliomas and subsequently followed a benign course. Neither patient had a family history or findings of neurofibromatosis.

Methods:

Case reports

Results:

Case 1: A 53-year-old woman presented in 2007 with a one-year history of seeing flashing lights and difficulty reading. She was failing to see letters at the ends of words. Visual acuity was 20/70 OD and 20/25 OS. Color vision was impaired OD > OS. There was no relative afferent pupillary defect. Automated perimetry (Humphrey) showed depression of central and temporal field iOD and temporal hemianopic depression OS. There was temporal pallor of both optic discs OD > OS. Slit lamp exam showed no Lisch nodules. MRI in 2007 showed a non-enhancing globular chiasmal mass measuring 2x3x2 cm, with contiguous enlargement of right intracranial optic nerve. Craniotomy confirmed an intrinsic lesion of chiasm and optic nerve. No biopsy was taken. She underwent stereotactic radiotherapy. At follow-up 3.5 years later, visual function was slightly worse but the glioma was smaller on MRI.

Case 2: A 38-year-old man presented in 2008 with bitemporal field defects, found incidentally on routine optometric examination. Visual acuity was 20/15 OD and 20/25 OS, with mildly impaired color vision OS. Kinetic visual fields (Goldmann) showed superior bitemporal depression. There was equivocal temporal pallor of both optic discs. Slit lamp exam showed no Lisch nodules. MRI showed a non-enhancing globular chiasmal mass measuring 3x3x2 cm, with areas of cystic change. At last follow-up 3 years later, visual function and MR appearance of the lesion were stable.

Conclusion:

Chiasmal gliomas presenting in adulthood may be benign.

References:

1. Hoyt WF, Meshel LG, Lessell S, Schatz NJ, Suckling N. Malignant optic glioma of adulthood. *Brain* 1973;96:121-32.
2. Rudd A, Rees JE, Kennedy P, Weller RO, Blackwood W. Malignant optic gliomas in adults. *J Clin Neuroophthalmol* 1985;5:23843.

Key Words: Chiasmal glioma, Adult, Benign

Financial Disclosure: The authors had no disclosures.

Poster 38

Papilledema from Pycnodysostosis

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Introduction:

Pycnodysostosis is an autosomal recessive skeletal disorder caused by a mutation in the gene for cathepsin K, a lysosomal proteinase required for bone remodeling by osteoclasts. It is usually associated with delayed closure of the cranial sutures. Craniosynostosis is a rare manifestation of this condition. We report a child with probable papilledema from pycnodysostosis.

Methods:

A 6-year-old boy was referred to the pediatric neuro-ophthalmologic clinic because of optic disc swelling. In addition to clinical examination, his evaluation included a skeletal survey, head CT, head MRI, MRV, and spine MRI.

Results:

The child came to medical attention because of delayed development and small stature. He walked at 18 months and had slow speech acquisition. His height, weight, and head circumference were 3d percentile. No visual symptoms were reported. Visual acuity was 20/20 OU. Pupils, eye movements, alignment, and stereopsis were normal. Cycloplegic refraction was +3.50 OU. There was mild, bilateral optic disc edema. On palpation the lambdoid suture was gaping. Head imaging showed diastasis of the lambdoid suture, but synostosis of the coronal and metopic sutures. The skull was abnormally dense. The optic canals were adequate in caliber. MR venogram showed a right dominant transverse and sigmoid sinus, but was otherwise unremarkable. The brain was normal. A skeletal survey showed typical findings of pycnodysostosis.

Conclusion:

Pycnodysostosis affects skeletal growth by interfering with osteoclast function. This child had diastasis of the lambdoid suture but premature fusion of the coronal and metopic sutures. Raised intracranial pressure from impaired skull growth was the most likely explanation for his optic disc swelling. Lumbar puncture was deferred because the child was asymptomatic and had normal visual function. We conclude that papilledema can occur as a rare finding in pycnodysostosis if craniosynostosis is present.

References:

1. Bertola D, Amaral C, Kim C, Albano L, Agüena M, et al. Craniosynostosis in pycnodysostosis: broadening the spectrum of the cranial flat bone abnormalities. Am J Med Genet A. 152A(10):2599-2603, 2010
2. Mujawar Q, Naganoor R, Patil H, Thobbi AN, Ukkali S, et al. Pycnodysostosis with unusual findings: a case report. Cases J. 23;2:6544, 2009
3. Osimani S, Husson I, Passemard S, Elmaleh M, Perrin L, et al. Craniosynostosis: A rare complication of pycnodysostosis. Eur J Med Genet. 53(2):89-92, 2010

Key Words: Pycnodysostosis, craniosynostosis, Papilledema, cathepsin K

Financial Disclosure: The authors had no disclosures.

Poster 39

Giant cell arteritis in the frontal branch of the superficial temporal artery with complete sparing of the parietal branch

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Introduction:

The superficial temporal artery splits into frontal and parietal branches. The frontal branch is usually chosen for biopsy, but there is a small risk of facial nerve injury. It is unknown which branch is more likely to demonstrate granulomatous inflammation in temporal arteritis. No prior report has described the occurrence of vasculitis confined to the frontal branch of the superficial temporal artery, without involvement of the parietal branch.

Methods:

A 69-year-old woman had neck stiffness, headache, and a sed rate of 55mm/hr. A biopsy was performed of the parietal branch of the left superficial temporal artery. Two days later, a biopsy was done of the frontal branch of the left superficial artery. Both specimens were processed for histopathology using a hematoxylin and eosin and an elastic Gieson stain.

Results:

108 sections were cut at 36 different levels through a 3.0cm segment of the parietal branch of the superficial temporal artery. None showed evidence of arteritis. In contrast, every section from the frontal branch of the superficial temporal artery showed extensive lymphocytic and granulomatous inflammation with multinucleated giant cells.

Conclusion:

The existence of “skip area” in the superficial temporal artery has been long debated. Here, we document the ultimate skip area: a parietal branch completely free of inflammation in a patient with extensive arteritis of the frontal branch. This finding raises the possibility that positive biopsy rates might differ for the frontal versus parietal branches. Surgeons should note which branch is biopsied, so data can be gathered to address this issue. After negative biopsy, one might consider harvesting the other branch on the same side, rather than performing a biopsy on the other side.

References:

1. Yoon MK, Horton JC, McCulley TJ. Facial nerve injury: a complication of superficial temporal artery biopsy. *Am. J. Ophthalmol.* 152:251-255, 2011.
2. Breuer GS, Neshet G, Neshet R. Rate of discordant findings in bilateral temporal artery biopsy to diagnose giant cell arteritis. *Rheumatol.* 36(4):794-796, 2009.
3. Bley TA, Wieben O, Uhl M, Vaith P, Schmidt D et. al. Assessment of the cranial involvement pattern of giant cell arteritis with 3T magnetic resonance imaging. *Arthritis Rheum.* 52:2470-2477, 2005.
4. Poller DN, van Wyk Q, Jeffrey MJ. The importance of skip lesions in temporal arteritis. *J Clin Pathol.* 53:137-139, 2000.

Key Words: Giant cell arteritis, Temporal artery biopsy, Skip lesions

Financial Disclosure: The authors had no disclosures.

Poster 40

Don't think it as pseudopapilledema.

Soh-youn Suh¹, Jin Choi², Seong-Joon Kim¹

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Introduction:

Idiopathic intracranial hypertension (IIH) is a condition of raised intracranial pressure (ICP) in the absence of space occupying lesions. Headache is the most common symptom at presentation. ICP is usually measured by lumbar puncture and cerebrospinal fluid pressure (CSF) above 250 mmH₂O is one of the diagnostic criteria in IIH. Recently we've met some patients who complained of headaches and showing disc swelling without increased ICP. Here we want to introduce a few cases of IIH patients whose ICP were within normal range but had their symptoms relieved by taking acetazolamide.

Methods:

A 39-year-old woman was presented with ocular pain accompanied by headache. Her optic discs and surrounded retinal nerve fiber layers (RNFL) were swollen. A second patient was a 52-year old woman who presented with impaired color perception and headache. She had diffuse disc elevation and her goldmann perimetry showed enlarged blind spots bilaterally. Another patient was a 15-year-old girl who had headache and tinnitus and she also had disc swelling. In each patient, the ICP was checked as 170 mmH₂O, 130 mmH₂O and 200 mmH₂O. The brain MRI was normal in three patients. We prescribed acetazolamide and made frequent follow-ups, since they had definite disc swelling with IIH related symptoms.

Results:

Symptoms have resolved in three patients after they started to take acetazolamide. Repeated lumbar puncture was done, however, the rechecked CSF pressure was still within the normal range in each patient. The drug had been tapered in all patients after their symptoms have been resolved.

Conclusion:

It is generally known that increased ICP measured by lumbar puncture is the most important sign in diagnosing IIH. However, even the CSF pressure is within the normal range, we should be alert when the patient has papilledema with related symptoms, since the untreated papilledema may cause progressive and irreversible visual loss.

References: None

Key Words: papilledema, Idiopathic intracranial hypertension(IIH), headache, intracranial pressure(ICP), acetazolamide

Financial Disclosure: The authors had no disclosures.

Poster 41

Is it Toxic or Cancer-associated?

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Introduction:

A 19-year-old girl was referred to our ophthalmologic department for yellowish vision and acute visual loss. She was an acute leukoid leukemia patient and had been on voriconazole treatment for invasive aspergillosis since 2 months earlier. Here, we want to present a case of a patient that was difficult to make a differential diagnosis since she had some characteristics of both voriconazole-induced toxic retinopathy and cancer-associated retinopathy (CAR).

Methods:

Her visual acuity was hand motion bilaterally and slit lamp test revealed normal anterior segment. On fundoscopic examination, her optic discs looked normal; however, there were non-specific retinal pigment epithelial (RPE) changes. Goldmann perimetry showed only peripheral islands of vision and the standard electroretinography (ERG) was barely recordable. Fluorescein angiography showed mild RPE atrophy over the whole retina and the macular inner/outer segments (IS/OS) junction of the photoreceptors seemed to be disrupted on spectral domain optical coherence tomography (SD-OCT). From the serum sample, anti-recoverin antibodies were not detected.

Results:

Immediately after she complained of her visual symptoms, the treatment regimen for her invasive aspergillosis was changed from voriconazole into other anti-fungal agent. 11 days after her initial visit, her visual acuity improved to 20/30 OD, 20/25 OS naturally. Even though the whole visual field was still constricted, central visual field has improved. However, there was no significant change on follow-up ERG.

Conclusion:

We could not make final diagnosis of this patient since there were some confusing aspects. Voriconazole related visual adverse events are known to be transient and fully reversible in most of the cases. Even though her visual acuity improved after discontinuation of voriconazole, the ERG after her visual improvement was still non-recordable. On the other hand, as her vision improved without any treatment and anti-recoverin antibody was not detected in her serum, we thought that it was less likely to be CAR.

References: None

Key Words: acute visual loss, differential diagnosis, voriconazole, toxic retinopathy, cancer-associated retinopathy

Financial Disclosure: The authors had no disclosures.

Poster 42

Nerve fiber loss in children with craniopharyngioma is in correlation with visual fields defects

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Introduction:

Children with craniopharyngioma are usually diagnosed after the development of optic neuropathy. The tumor directly compresses the optic nerve, leading to visual field defects. The common objective measurement is the automated perimetry test, inappropriate for young children or those with poor visual acuity (VA). Recently, the use of peripapillary optical coherent tomography (OCT) may offer an alternative objective method to monitor progressive optic neuropathy. Peripapillary OCT is swift, easy to perform, and is highly correlated to anatomical-functional changes. The aim of this study is to compare repeated OCT measurement in children with craniopharyngiomas and their visual function.

Methods:

This is a retrospective cohort of children diagnosed with craniopharyngioma in our hospital between 1999-2011. Review of medical records, imaging, and Humphrey visual fields were performed. Peripapillary nerve fiber layer (NFL) thickness measured by OCT was analyzed and compared with VA, VFs, and optic nerve pallor. Differences were analyzed between repeated tests.

Results:

20 of 26 children diagnosed with craniopharyngioma were included (10 males, 10 females). The average age at diagnosis was 6.5 ± 3.88 years. The common symptom at diagnosis was a headache. Only one complained of visual loss. BCVA Log Mar was 0.036 ± 0.06 for healthy eyes and 1.05 ± 1.45 for optic neuropathy eyes. Signs were positive RAPD (8/20 children), VA loss (7/20), temporal VF loss (bilateral 4/15, unilateral 4/15), papilledema (3/20) and uni/bilateral optic disc pallor (14/20).

Compared with normal eyes, average RNFL thickness was reduced 14-20 μ m when optic neuropathy existed. There was no change in RNFL thickness over time (mean 18 ± 14.2 months) in seven children after 2 consecutive OCT measurements. NFL thickness and VA or VF-defect were correlated.

Conclusion:

: Craniopharyngioma leads to optic neuropathy in most pediatric patients. RNFL thickness measurement correlates with such VF defects. Thus, peripapillary OCT may contribute as an objective measurement in the follow-up of these children.

References: None

Key Words: craniopharyngioma, OCT, compressive optic neuropathy, visual field defect, pediatric neuro-ophthalmology

Financial Disclosure: The authors had no disclosures.

Poster 43

Retinal Nerve Fiber Layer Thickness in an elderly population: correlation with the cognitive state in the ALIENOR Study

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Introduction:

Alzheimer disease and other cognitive impairments become a challenging public health problem. Diagnosing these diseases earlier could lead to better treatment and outcome. It has been suggested that Retinal Nerve Fiber Layer (RNFL) thickness, which is an easy and safe eye examination, is thinner in Alzheimer's patients than in normal population. Nevertheless, previous studies had been conducted on small samples of patients. The aim of our study was i) to establish normative data of RNFL thickness in the elderly population; ii) to examine relationship between RNFL thickness and cognitive performances.

Methods:

The ALIENOR (Antioxydants, Lipides Essentiels, Nutrition and maladies OculaiRes) Study is a population-based epidemiological study on nutrition and age-related eye diseases. In 2009-2010, 591 subjects, aged 77 years or more had a RNFL thickness measurement obtained with a spectral domain OCT. The MMSE (Mini Mental State Examination, global performances), Benton (visual memory), Isaacs (verbal fluency) and Free and Cued Selective Reminding (episodic memory) tests were used for neuropsychological assessment.

Results:

59 subjects were excluded because of a diagnosis of glaucoma. The mean RNFL thickness among the non-glaucoma population was 91 μm (SD 12.7). Men had significantly thinner RNFL than women (89.2; SD 12.6 versus 92.1, SD 12.6, $p=0.005$). RNFL decreased significantly with age, from 93.0 (SD 12.0) in subjects younger than 80 to 90.1 (SD 12.9) in subjects older than 85 years ($p=0.003$). After multivariate adjustment, the lowest quartile of RNFL thickness ($<78\mu\text{m}$) was associated with lower performance at the Benton test ($p=0.04$). There were no significant associations with the other tests.

Conclusion:

This study provided normative data on RNFL thickness in an elderly population. Furthermore, it demonstrated a positive correlation between lower RNFL thickness and lower Benton score.

References: None

Key Words: Retinal Nerve Fiber Layer Thickness, Alzheimer disease, OCT, epidemiological study, cognitive state

Financial Disclosure: Marie-Benedicte Rougier: Thea, Allergan, Biogen, Bausch and Lomb. Jean Francois Korobelnik: Thea, Allergan, Novartis, Bayer. The remaining authors had no disclosures.

Poster 44

Comparison of Imaging Techniques for Posterior Pole Folds in Patients with Presumed Choroidal Folds

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Introduction:

Choroidal folds are parallel grooves or striae in the choroid of the posterior pole. The purpose of this study is to evaluate the ability of confocal scanning ophthalmoscopy (SLO), optical coherence tomography (OCT) and Multi-Spectral Imaging (MSI) to characterize posterior pole folds in pathologies associated with choroidal folds.

Methods:

Patients with posterior pole folds on clinical exam were identified in the Neuro-Ophthalmology and Retina subspecialty clinics at the Ottawa Eye Institute. Three dimensional colour and red-free fundus photographs of the disc-macula were obtained using a retinal camera. MSI of the retina and underlying choroid along with OCT topographical T-Scans of the central 29 degrees along with horizontal and vertical B-Scans through the macula were obtained for each patient. Images were analysed by staff Neuro-ophthalmologist, Clinical Electrophysiologist and Retina Specialist. Each set of images was graded for the presence or absence of folds and the layer of the folds.

Results:

Thirteen of 16 eyes with presumed choroidal folds on clinical exam also had choroidal folds on SLO/OCT and MSI. Two eyes (all idiopathic intracranial hypertension (IIH)) had retinal folds in the absence of choroidal folds on imaging.

Conclusion:

Clinical examination alone may not correctly distinguish choroidal from retinal folds. In IIH, folds classically thought to be choroidal, may actually be retinal without involvement of the choroid. OCT/SLO and MSI are valuable methods for examining chorioretinal folds. This study represents the largest collection of OCT and MSI data for patients with Choroidal Folds.

References: None

Key Words: Choroidal Folds, Multi-Spectral Fundus Imaging, OCT

Financial Disclosure: The authors had no disclosures.

Poster 45

Treated Papilledema in Children: Subclinical Optic Atrophy Detected by OCT

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Introduction:

Papilledema can cause visually-significant optic nerve head (ONH) damage. This can be hard to detect in children who cannot reliably perform visual fields. Optical coherence tomography (OCT) has been used to objectively grade the amount of swelling in papilledema, however, there is no objective clinical tool to differentiate resolving papilledema from superimposed optic atrophy.

Methods:

Children with treated IIH and residual papilledema grade ≤ 1 on the Frisen scale and normal controls had a complete ophthalmic examination and SD-OCT (Heidelberg-Engineering, Germany) measurements of the peripapillary Retinal Nerve Fiber Layer (RNFL) and macular thickness/volume. Segmentation of inner retinal layers to measure volumes of macular ganglion cell-inner plexiform layer (GC-IPL) and Nerve Fiber Layer (NFL) was done. Pale ONH were excluded.

Results:

19 patients and 19 normals were enrolled. Mean age 11.5 ± 4.6 yrs. 63% were prepubertal; 36% patients had moderate to severe pre-treatment papilledema (Grade ≥ 3) on presentation: 15 were treated medically, 2 required optic nerve sheath fenestration, 3/38 ONH were pale. Time from diagnosis to stabilization/resolution of papilledema was 75 days. RNFL was thicker in papilledema vs. controls (RNFL = $116 \pm 25 \mu\text{m}$ vs. $103 \pm 10 \mu\text{m}$, respectively, $p=0.05$). Nasal RNFL was thicker in papilledema vs. normals (95 ± 35 vs. $72 \pm 15 \mu\text{m}$, $p=0.04$), there was a trend towards thinner temporal RNFL in papilledema vs. normals (67 ± 15 vs. $77 \pm 10 \mu\text{m}$, $p=0.16$). Macular thickness was thinner in papilledema vs. normals in 4/8 sectors studied (Average thickness 336 ± 16 vs. $344 \pm 13 \mu\text{m}$ ($p < 0.05$)). GC-IPL volume was thinner in papilledema vs. normal (GC-IPL = $2.0 \pm 0.2 \text{ mm}^3$ vs. $2.2 \pm 0.1 \text{ mm}^3$, $p=0.03$). There was no difference at the level of the macular NFL.

Conclusion:

In children with treated IIH, OCT showed thinning at the level of the macula especially at the GC-IPL layer in mildly swollen despite no signs of ONH pallor.

References: None

Key Words: pediatric, idiopathic intracranial hypertension, optical coherence tomography (OCT), optic atrophy, segmentation of OCT

Financial Disclosure: The authors had no disclosures.

Poster 46

Optic disc edema does not always correlate with retinal nerve fiber layer (RNFL) thickening

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Introduction:

Optical coherence tomography (OCT) allows precise anatomical assessment of the retina and optic nerve head (ONH). The thickness of the retinal nerve fiber layer (RNFL) surrounding the optic nerve has been validated as a measure of the integrity of the retinal ganglion cell (RGC) axons that are its chief constituent. The RNFL may be thickened in conditions associated with visible optic disc edema (ODE), including papillitis and papilledema associated with idiopathic intracranial hypertension (IIH). I sought to develop a standardized method of quantifying ONH volume, comparing values with those of the RNFL in patients with ODE.

Methods:

OCT was measured by an experienced technician with a spectral domain unit (Spectralis, Heidelberg Engineering). All tests were interpreted personally. All recordings were of high quality. The RNFL was assessed with a standard 3.4mm circle scan centered on the optic disc. The ONH was measured with a series of parallel vertical scans that allowed 3-dimensional reconstruction. I defined cylinders of 2mm and 3mm diameter centered on the ONH and established the volumes of tissue contained therein, from the level of Bruch's membrane to the internal limiting membrane.

Results:

Using normal subject-derived cutoffs of 1.3mm^3 for 2mm diameters and 2.7mm^3 for 3mm diameters, I identified 64 eyes with enlarged ONHs in 41 subjects. Of these 64 eyes, 21 demonstrated normal or reduced mean RNFL thickness, only six of which showed increased thickness in any quadrant. Diagnoses in these eyes included IIH (8), hydrocephalus (4), drusen (4) and optic neuritis (2).

Conclusion:

Mismatches between optic disc volume and RNFL thickness may be explained by limited intraocular extent of axoplasmic engorgement, by mixed populations of dead and swollen RGC axons, or by intrusions of non-neural material in the ONH with drusen. I encourage the development of standardized measures of optic disc integrity to further this comparative analysis.

References: None

Key Words: optical coherence tomography, retinal nerve fiber layer, optic nerve head, papilledema, optic nerve drusen

Financial Disclosure: Dr. Morrow has served as a consultant for Biogen-Idec and a speaker for the National Multiple Sclerosis Society. He has received research support from Biogen-Idec and Novartis.

Poster 47

Spectral Oximetry Measured with Ultra-high Resolution Optical Coherence Tomography in Multiple Sclerosis

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Introduction

Multiple sclerosis (MS) is a devastating neurological disorder affecting young adults. Cerebral hypoperfusion in the normal-appearing white matter of MS patients have been reported.¹⁻⁷ The retinal vasculature has been used as a surrogate for *in vivo* studies of the brain microvasculature.⁸⁻¹⁰ The purpose of this project is to determine whether retinal vascular dysfunction occurred in MS.

Methods:

A slit-lamp adapted ultra-high resolution optical coherence tomography was developed and spectral analysis was performed. A-scans at the central wavelengths of 805 and 855 nm of the retinal artery and vein were analyzed for calculating optical density ratios (ODR), representing oxygen saturation in the retinal vessels. Four clinically definite remitting relapsing MS patients and four age and sex matched healthy subjects were recruited. Both eyes of MS patients and normal controls were scanned. None of these four MS patients had a history of optic neuritis or other medical problems. The range of their expanded disability status scale (EDSS) was from 1.0 to 1.5. Their visual acuity, visual fields and Retinal nerve fibre layer (RNFL) thickness were within normal ranges.

Results:

ODR of the retinal artery was 0.15 ± 1.09 (mean \pm SD) in MS patients, significantly lower than that in healthy controls (3.68 ± 4.42 , $P < 0.05$). ODR of the retinal vein was -0.16 ± 2.63 in MS, which was not significantly different compared to healthy controls (-0.22 ± 1.12 , $P > 0.05$). The usage of oxygen (ODR difference between artery and vein) was 0.32 ± 3.51 in MS which was significantly lower than that in normal controls (3.47 ± 3.54 , $P < 0.05$).

Conclusion:

We demonstrated that oxygen saturation levels of the retinal artery were lower in MS patients compared to that in normal controls, indicating retinal hypoperfusion in MS patients. Further studies will be needed to correlate the ocular findings to brain hypoperfusion with a large sample size.

References:

1. Saindane AM, Law M, Ge Y, et al. Correlation of diffusion tensor and dynamic perfusion MR imaging metrics in normal-appearing corpus callosum: support for primary hypoperfusion in multiple sclerosis. *AJNR Am J Neuroradiol.* 2007;28:767-772.
2. Lycke J, Wikkelso C, Bergh AC, Jacobsson L, Andersen O. Regional cerebral blood flow in multiple sclerosis measured by single photon emission tomography with technetium-99m hexamethylpropyleneamine oxime. *Eur Neurol.* 1993;33:163-167.
3. Inglese M, Adhya S, Johnson G, et al. Perfusion magnetic resonance imaging correlates of neuropsychological impairment in multiple sclerosis. *J Cereb Blood Flow Metab.* 2008;28:164-171.
4. Adhya S, Johnson G, Herbert J, et al. Pattern of hemodynamic impairment in multiple sclerosis: dynamic susceptibility contrast perfusion MR imaging at 3.0 T. *Neuroimage.* 2006;33:1029-1035.
5. Varga AW, Johnson G, Babb JS, et al. White matter hemodynamic abnormalities precede sub-cortical gray matter changes in multiple sclerosis. *J Neurol Sci.* 2009;282:28-33.
6. De Keyser J, Steen C, Mostert JP, Koch MW. Hypoperfusion of the cerebral white matter in multiple sclerosis: possible mechanisms and pathophysiological significance. *J Cereb Blood Flow Metab.* 2008;28:1645-1651.

7. Law M, Saindane AM, Ge Y, et al. Microvascular abnormality in relapsing-remitting multiple sclerosis: perfusion MR imaging findings in normal-appearing white matter. *Radiology*. 2004;231:645-652.
8. Baker ML, Hand PJ, Wang JJ, Wong TY. Retinal signs and stroke: revisiting the link between the eye and brain. *Stroke*. 2008;39:1371-1379.
9. Patton N, Aslam T, MacGillivray T, et al. Retinal vascular image analysis as a potential screening tool for cerebrovascular disease: a rationale based on homology between cerebral and retinal microvasculatures. *J Anat*. 2005;206:319-348.
10. Baker ML, Hand PJ, Liew G, et al. Retinal microvascular signs may provide clues to the underlying vasculopathy in patients with deep intracerebral hemorrhage. *Stroke*. 2010;41:618-623.

Key Words: Ultra-high resolution optical coherence tomography, Multiple sclerosis, spectral oximetry, retinal vascular dysfunction, hypoperfusion

Financial Disclosure: The authors had no financial disclosures.

Poster 48

Correlation of Peripapillary Retinal Nerve Fiber Layer Thickness as Measured by SD-OCT with Severe Vision Loss in End Stage Optic Neuropathy.

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Introduction:

Optical Coherence Tomography (OCT) is useful in monitoring optic nerve disease. Few studies have looked at the utility of OCT in severe vision loss (SVL) states [1,2]. Visual acuity in patients with end stage optic neuropathy may be classified using the semiquantitative scale of counting finger (CF), hand motion (HM), light perception (LP), and no light perception (NLP). Patients notice a difference in the ability to perform daily tasks when visual acuity changes at this level (e.g. from CF to HM) [3]. Detecting morphologic and perimetric changes among these SVL states is difficult. Correlating peripapillary retinal nerve fiber layer thickness (PRNFLT) with SVL states in patients with optic neuropathies may help determine threshold for change.

Methods:

A retrospective chart review was performed of patients with a diagnosis of optic neuropathy with a best corrected visual acuity (BCVA) of CF, HM, LP and NLP. Inclusion criteria were a stable BCVA, disease state present for at least one year and previous PRNFLT obtained. Exclusion criteria were a Q number of <15 and other pathology affecting PRNFLT results or visual function. Spectral Domain (SD) OCT Mean PRNFLT and quadrant data were analysed with paired T-test and non-parametric Spearman correlation.

Results:

41 eyes in 38 patients were reviewed. Five eyes were excluded. Of the 36 eyes, 14 had SVL related to glaucoma while 22 had SVL related to nonglaucomatous optic neuropathies. Statistical analysis indicated there was no significant difference in PRNFLT among the four SVL groups.

Conclusion:

Our small data set suggests no significant difference in SD-OCT PRNFLT among SVL states in end stage optic neuropathy. With severe thinning of PRNFLT, non-axonal content such as glial tissue and blood vessels may be significantly contributing to the measured PRNFLT. Eliminating all non-axonal contributions to PRNFLT on SD-OCT may provide more accurate measurements in severe optic neuropathies.

References:

1. Sihota, Sony, Gupta, Dada, Singh. Diagnostic Capability of Optical Coherence Tomography in Evaluating the Degree of Glaucomatous Retinal Nerve Fiber Damage. *Invest Ophthalmol Vis Sci.* 47:2006-10; 2006.
2. Chan, Miller. Peripapillary Nerve Fiber Layer Thickness Measured by Optical Coherence Tomography in Patients With No Light Perception From Long-Standing Nonglaucomatous Optic Neuropathies. *J Neuro-Ophthalmol.* 27:176-79; 2007.
3. Grover, Fishman, Anderson, Tozatti, Heckenlively, et al. Visual Acuity Impairment in Patients with Retinitis Pigmentosa at Age 45 years or Older. *Ophthalmology* 106:1780-85; 1999.

Key Words: Optic Neuropathy, Low vision, Optical Coherence Tomography

Financial Disclosure: The authors have no disclosures.

Poster 49

Optical coherence tomography findings in patients with dominant optic atrophy

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Introduction:

To report optical coherence tomography (OCT) findings measured by time-domain OCT in patients with dominant optic atrophy (DOA)

Methods:

Cross-sectional study. Four patients with DOA and twelve age-matched control subjects were enrolled. Detailed ophthalmologic examinations were performed including measurements of optic rim area, disc area, cup volume, peripapillary retinal nerve fiber layer (RNFL) thickness and macular thickness by time-domain OCT.

Results:

Mean age of DOA patients was 9.7 years. Patients with DOA had mean best-corrected visual acuity of logMAR 0.35. The optic nerve head (ONH) analysis of DOA patients showed a significantly smaller optic disc area (1.8 vs. 2.1 mm²) and rim area (1.34 vs. 1.84 mm²), compared with controls. OCT using a peripapillary fast RNFL program showed a decrease in average RNFL thickness in DOA patients (101.3 um vs. 108.8 um) compared with controls.

Conclusion:

This cross-sectional study demonstrated a significantly smaller ONH and thinner RNFL compared with the values observed in control subjects. These features may have relevance for DOA pathogenesis.

References: None

Key Words: dominant optic atrophy, optic nerve head size, retinal nerve fiber layer, optical coherence tomography

Financial Disclosure: The author has no disclosures.

Poster 50

Optical Coherence Tomography (OCT) monitoring in shunt failure in a child with stiff ventricle syndrome and normal imaging.

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Introduction:

Stiff ventricular wall syndrome is a rare entity that occurs in patients with shunted hydrocephalus. The radiological signs of high ICP (ventricle enlargement, sulcal or cisternal obliteration) aren't present.

Methods:

An 11-year-old boy with Arnold-Chiari malformation was diagnosed at age five and treated with decompression. At age ten, headaches with associated vertigo ensued and evaluation for shunt blockage was negative. Migraine medication improved his symptoms. A year later, he complained of headaches and RE pain along with blurred vision. His first visit to our center revealed bilateral papilledema, vessel engorgement, and tortuosity and retinal striae. His OCT values were 133 μ m RE, 119 μ m LE which suggested asymmetric retinal nerve fiber layer (RNFL) edema. His head CT scan was normal and his neurosurgeon decided to not intervene.

Results:

Two weeks later, he developed progressive eye pain and headaches. His visual fields weren't reliable. His OCT was 188 μ m RE and 182 μ m LE, confirming the worsening of his papilledema. Shunt failure was now thought to be definite, with the increase in RNFL bilaterally despite normal brain imaging and therefore a second neurosurgical consultation was sought.

An LP revealed opening pressure of 39 cmH₂O. A shunt tap showed an obstruction by calcification. A new shunt valve was placed. One week after, he developed a tonic seizure along with LE pain and blurred vision. He was admitted and had his shunt tapped, which revealed bloody CSF and the pressure was beyond 50cc H₂O. Neurosurgery revised his shunt and his symptoms dissipated. Two weeks later he was seen for follow up and his OCT showed improved RNFL thickness to values of, 134 μ m RE and 133 μ m LE.

Conclusion:

OCT is a helpful tool in quantifying papilledema, especially in children where other methods may be less objective. It shows change over time parallel to observation of disc edema ophthalmoscopically. OCT can objectify optic disc swelling and evaluate this syndrome at an early stage which may be sight and also life preserving.

References:

1. Trobe JD, Papilledema, The Vexing Issues, *J Neuro-Ophthalmol* 2011; 31:175-186
2. Mizrachi IB, Trobe JD, Gebarski SS, Garton HJ., Papilledema in the assessment of ventriculomegaly. *J Neuroophthalmol.* 2006 Dec; 26(4):260-3.
3. Heidary G, Rizzo JF III. Use of the Optical Coherence Tomography to evaluate papilledema and pseudopapilledema. *Semin Ophthalmol.* 2010 Sep-Nov;25(5-6):198-205. Review.

Key Words: Optical Coherence Tomography (OCT), Papilledema, Stiff ventricle syndrome, Shunt malfunction

Financial Disclosure: Rosa Ana Tang: none for topic. Principal investigator for IIHTT-NORDIC at University of Houston. The remaining authors had no disclosures.

Poster 51

Optical Coherence Tomography Value in Subclinical Optic Neuritis

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Introduction:

To determine the utility of optical coherence tomography (OCT) in patients with subclinical optic neuritis.

Methods:

OCT was performed on 4 patients (6 eyes) with subclinical optic neuritis. The parameters used to define an eye has subclinical optic neuritis was the neurophthalmic examination, visual fields and photos of the fundus of the eye

Results:

Four patients with subclinical optic neuritis by multiple sclerosis were observed during the follow consultation as a causal. Three patients developed unilateral optic neuritis and one patient bilateral optic neuritis. The visual field demonstrated paracental scotomas; the optic disc photo showed decreased excavation during the crisis and increased the cup after event resolved. The optical coherence tomography demonstrated decreased the measurement parameter of the optic disc with incremented of the values after the optic neuritis disappear. Also, a bigger cup was observed than the initial size.

Conclusion:

Optic disc cup OCT measurement could be a good indicator of subclinical optic neuritis.

References: None

Key Words: Optic neuritis, Optical Coherence Tomography, Subclinical optic neuritis, Multiple Sclerosis, OCT

Financial Disclosure: The authors had no disclosures.

Poster 52

Novel Acquisition Protocol with Spectral-Domain Optical Coherence Tomography: Using Star-Pattern Scans for the Evaluation of Papilledema

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Introduction:

Optical coherence tomography (OCT) is currently used as an adjunctive, quantitative tool in the evaluation of patients with papilledema. Past studies have used analysis of the retinal nerve fiber layer thickness and the peripapillary total retinal thickness (PTR) for optic nerve edema. We describe an acquisition protocol for papillary thickness (PT) with star-pattern scans (SPS), which, in our experience, seems more precise for the diagnosis and follow-up of papilledema.

Methods:

A case series of eight eyes from four patients with papilledema. PTR and PT were evaluated with Spectral-Domain (SD-) OCT using two methods: macular cube acquisition protocol centred on the optic disc for PTR and SPS using forty-eight radial slices through the optic disc for PT, measured from the end of Bruch's membrane. Thickness values were calculated in four quadrants and compared between them. Wilcoxon rank test was used for statistical analysis.

Results:

The mean PT measured using SPS were 647 (± 87.7) μm , 441 (± 83.7) μm , 609 (± 104.7) μm and 614 (± 92.8) μm for the superior (S), temporal (T), inferior (I) and nasal (N) quadrants, respectively. The mean PTR measured using macular cube acquisition were 572 (± 103.5) μm , 376 (± 37.9) μm , 527 (± 85.7) μm and 489 (± 74.0) μm for the S, T, I and N quadrants. In each quadrant, the values of PT were higher than that of PTR (S: 75 μm ; T: 65 μm ; I: 82 μm ; and N: 125 μm) and these differences were statistically significant ($P = 0.012 - 0.036$).

Conclusion:

PT measured with SPS acquisition protocol on SD-OCT is significantly higher than PTR. We speculate that PT could be a more sensitive sign to monitor disease progression and response to treatment than PTR. This is a pilot study to validate the measurement method of our future prospective studies that will compare the changes in PT and PTR during the evolution of papilledema.

References: None

Key Words: Spectral-Domain Optical Coherence Tomography, Papilledema, Star-Pattern Scans, Papillary Thickness

Financial Disclosure: The authors had no disclosures.

Poster 53

Congruous Homonymous Sectoranopia in Multiple Sclerosis: an Atypical Cause of Lateral Geniculate Body Lesion

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Introduction:

Multiple sclerosis may cause demyelination at any level of the visual sensory pathway; however, less than 10% of patients develop homonymous visual field defects. We describe the case of a patient who presented to our clinic complaining of loss of vision on her left side. Goldman visual-field testing demonstrated left congruous homonymous sectoranopia.

Methods:

Pubmed database was reviewed for causes of congruous homonymous sectoranopia: to our knowledge, this is the only case of a true wedge-shaped congruous horizontal sectoranopia (CHS) secondary to a demyelinating lesion.

Results:

Brain magnetic resonance imaging revealed 15 supratentorial white-matter signal anomalies, with a 20-mm demyelinating lesion on the lateral aspect of the left thalamic geniculate body (LGN).

Most cases of CHS are caused by stroke with infarction in the territory of the lateral choroidal artery. Only three cases of atypical CHS have been described: head trauma, arteriovenous malformation and tumor infiltration. To our knowledge, demyelination has never been previously reported.

Conclusion:

Yet another presenting symptom of multiple sclerosis has been unveiled. This case emphasizes the fact that demyelinating lesions may arise at any level of the nervous system, including in the LGN with manifestation of sectoranopia.

References: None

Key Words: Sectoranopia, Multiple Sclerosis, Lateral Geniculate Body

Financial Disclosure: The authors had no disclosures.

Poster 55

Retinal Nerve Fiber Layer Thickness Estimation: Humans and Machines

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Introduction:

Optic disc pallor is a clinical description. We aimed to determine if the clinical estimation of optic disc pallor correlates to the OCT measurement. We hypothesized that historical values of RNFL thickness concordance of agreement using OCT would be higher than concordance of agreement for different physicians estimating RNFL thickness by examining fundus photos.

Methods:

Digital photographs and OCTs were obtained from 37 patients with a range of RNFL thickness between 35 and 110 microns. Disc photos were sent to three neuro-ophthalmologists with clinical experience of 21, 26, and 29 years who were asked to estimate the RNFL thickness based on the fundus photos.

Results:

OCT RNFL thickness ranged from 37.7 microns to 104.6 microns. The agreement indices with OCT based on the concordance correlation coefficient for the three physicians were 0.53, 0.43, and 0.72. Between physicians, there was a 57.7% chance of estimating an RNFL value within 10 microns of each other, and 83.8% chance within 20 microns. There was a 52.3% chance among all physicians of an estimation within 10 microns of the OCT, and 82.0% chance within 20 microns. The chance of underestimating and overestimating the OCT result for all physicians was equal at 50%.

28 photos represented eyes with an OCT RNFL of 75 microns or less. 63/84 (75%) of these estimations were judged as 75 microns or less by the physicians. For RNFL <54 microns, 100% (15/15) of estimations judged the RNFL as less than or equal to 75 microns.

Conclusion:

To our knowledge, there is no prior study examining the ability of a neuro-ophthalmologist to estimate the average peripapillary RNFL thickness or comparing this estimation to OCT. Although an experienced neuro-ophthalmologist has an excellent chance of estimating the approximate RNFL thickness, OCT appears more consistent in RNFL measurement precision.

References: None

Key Words: Optical coherence tomography, Optic atrophy, Optic neuropathy, Retinal nerve fiber layer thickness

Financial Disclosure: The authors had no disclosures.

Poster 56

Oscillopsia and Orbital Pulsations Due to Post-Traumatic Compromise of the Orbital Roof

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Introduction:

Orbital pulsations can occur from cavernous carotid fistula, orbital venous varix, orbital arteriovenous malformation, orbital tumor, intraorbital ophthalmic artery aneurysm, tricuspid regurgitation, or an orbital wall defect, which is usually associated with neurofibromatosis.

Methods:

A 21-year-old man was an unrestrained front seat passenger in a motor vehicle accident. He was intubated on hospital arrival with GCS 8. CT head showed bifrontal haemorrhage with subfacial and posteroinferior herniation and comminuted, complex fractures of the skull base, facial bones, and frontal bones. CT of facial bones showed a complex nasal ethmoidal smash injury with fracture through the left sphenoid sinus walls. He underwent a bifrontal craniectomy with elevation of the depressed skull fracture and a frontal external ventricular drain insertion. There was no surgical intervention for his facial fractures. He had a bifrontal cranioplasty approximately 40 days later.

Results:

Approximately 60 days after his initial injury, once able to communicate, he reported left monocular vertical oscillopsia, and a throbbing sensation around his left eye. On exam, his left globe showed nystagmoid movements corresponding to visible pulsations of the globe, and Doppler ultrasound of the radial artery confirmed timing of the movements concomitant to arterial pulsations. CT of the facial bones revealed absence of the orbital roof, without bony separation of the orbital contents from the frontal lobe of the brain. Mesh was not placed during surgical interventions due to lack of structural mesh support.

Conclusion:

The cause of orbital pulsations in this patient is due to the transmission of intracranial CSF pulsations through the bony defect in the orbital roof through to the orbital contents. Orbital pulsations have been rarely reported due to incompetent orbital bone structure, and we believe our report is the first example of oscillopsia and orbital pulsations resulting from a post-traumatic mechanically compromised orbital roof.

References: None

Key Words: Oscillopsia, Orbital Pulsations, Orbital roof fracture

Financial Disclosure: The authors had no disclosures.

Poster 57

The Clinical manifestations of Intracranial Tumor Presenting with Visual Disturbances

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Introduction:

To evaluate ophthalmological features in patients with intracranial tumors presenting with visual problems and assess the usefulness of various examinations.

Methods:

35 patients with intracranial tumors (22 males and 13 females) diagnosed by magnetic resonance imaging (MRI) were enrolled. Only the patients who were initially diagnosed at our hospital and confirmed by MRI were enrolled, and previously known intracranial tumor patients and patients with strabismus or proptosis were excluded. Best corrected visual acuity (BCVA), pupillary reflex test, fundus examination, color vision test and visual field test were done to evaluate visual impairments. We investigated the incidence of intracranial tumors and sensitivity of tests.

Results:

Mean age was 43.7 years old (6-77 years old). The most common tumors were pituitary tumors (60.0%), followed by meningioma (20.0%), optic nerve glioma (5.7%) and Schwannoma, metastatic tumor, craniopharyngioma, meningocele and unspecified tumor (respectively 1 patient, 2.9%). The BCVA of affected eyes ranged from 1.0 to no light perception. The visual acuity of 3 patients was 1.0 in both eyes. Positive relative afferent pupillary defect (RAPD) was seen in 69.0% and abnormal disc findings including swelling, high cup/disc ratio and pallor were found in 70% of patients. The color vision test was more specific (sensitivity: 85.7%), and all patients had abnormal visual field defects.

Conclusion:

Pituitary tumor is the most common intracranial tumor presenting with visual problems in this secondary referral hospital. Among tests, a visual field test is a more sensitive test than visual acuity, color vision, RAPD and fundus examination for detecting compressive optic neuropathy.

References: None

Key Words: intracranial tumor, compressive optic neuropathy, visual field

Financial Disclosure: The authors had no disclosures.

Poster 58

Neurosyphilis presenting as a unilateral Adie's tonic pupil

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Introduction:

Pupil abnormalities are relatively common in patients with neurosyphilis. The most common pupil abnormality being light-near dissociation in pupils that are either bilaterally small (Argyll Robertson pupils) or bilaterally large (dilated tonic pupils).(1,2) To our knowledge, there have only been 2 previous case reports of neurosyphilis presenting with an isolated, unilateral, dilated tonic pupil.(4,5)

Methods:

Case report

Results:

A 38 year-old female presented with an acute unilateral dilated tonic pupil, responsive to dilute pilocarpine, but without other ocular or neurological findings and a negative review of systems. This was presumed to represent an Adie's tonic pupil and no further investigations were conducted at presentation. Two months later, she returned with bilateral dilated tonic pupils and an exotropia. Further testing demonstrated a reactive serum Rapid Plasma Reagin and a positive Treponema Pallidum Enzyme Immunoassay. Cerebrospinal fluid analysis demonstrated an elevated protein and white blood cell count, as well as a positive Fluorescent Treponemal Antibody Absorption and Venereal Disease Research Laboratory test. MRI/MRA brain with Gadolinium was unremarkable.

Conclusion:

Testing for syphilis should be considered in cases of isolated, unilateral, dilated tonic pupils (presumed Adie's tonic pupil).

References:

1. Englestein ES, Ruderman MI, Troiano RA, Digiovanni VJ; Dilated tonic pupils in neurosyphilis. *Journal of Neurology, Neurosurgery, and Psychiatry*: 49:1455-1461, 1986.
2. Fletcher WA, Sharpe JA. Tonic pupils in neurosyphilis. *Neurology*: 36(2):188-192, 1986.
3. Camoriano GD, Kassab J, Suchak A, Gimbel HV; Neurosyphilis masquerading as an acute Adie's tonic pupil: Report of a Case. *Case reports in ophthalmology*: 201;2205-2010, 2011
4. Zeligs MA, Joseph GF. Unilateral internal ophthalmoplegia: sole clinical sign in patient with syphilitic meningitis. *Ach Neurol Psychiatry*; 54:389-390, 1945.

Key Words: Pupil, Neurosyphilis

Financial Disclosure: The authors had no disclosures.

Poster 59

Two Cases of Orbital Hemorrhage Associated with Pregnancy and Labor

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Introduction:

Orbital hemorrhage is divided into the more common intraorbital hemorrhage and the less common subperiosteal hemorrhage. The most common cause for orbital hemorrhage is direct facial or orbital trauma. Nontraumatic orbital hemorrhage has been reported in association with sudden elevation of cranial venous pressure and venous congestion, systemic diseases associated with bleeding tendencies, and paranasal sinusitis. Occasionally, no associated cause has been found. We report two cases of orbital hemorrhage associated with pregnancy and labor.

Methods:

Patient 1 presented with diplopia and upper eyelid swelling during childbirth. On exam, she was noted to have a left afferent pupillary defect, limitation in extraocular motility (supraduction, abduction, and adduction of the left eye), and proptosis. MRI of the brain revealed a 1.0 x 3.3 x 2.6 cm hemorrhage in the superior aspect of the left orbit. Patient 2 presented with diplopia and eye swelling one month postpartum. On exam, she was noted to have marked proptosis of the left eye. CT scan of the head and orbits revealed bilateral subperiosteal lesions inferiorly in both orbits, left (3.3 x 2.6 x 2.2 cm) greater than right.

Results:

Patient 1 was monitored closely and symptoms completely resolved by 6 weeks postpartum without treatment. Patient 2 underwent left anterior orbitotomy with hematoma drainage and resolution of symptoms.

Conclusion:

A high index of clinical suspicion is required for an orbital hemorrhage both during and immediately after pregnancy. Such hemorrhages have been reported during labor; our second case demonstrates that the coagulopathy of the puerperium also may give rise to this condition. Vertical diplopia, eyelid ptosis, proptosis, globe displacement, and periorbital pain can be presenting clinical features in cases of orbital hemorrhage associated with pregnancy. Careful monitoring of these patients is essential for proper management and treatment, as prompt intervention may be needed to prevent visual decline.

References:

1. Atalla ML, McNab AA, Sullivan TJ, et al. Nontraumatic Subperiosteal Hemorrhage. *Ophthalmology*. 108(1): 183-9, 2001.
2. Polito E, Leccisotti A. Diagnosis and treatment of orbital hemorrhagic lesions. *Ann Ophthalmol*. 26(3): 85-93, 1994.
3. Sullivan TJ, Wright JE. Non-traumatic orbital haemorrhage. *Clin Exp Ophthalmol*. 28(1): 26-31, 2000.
4. Tan JC, Chan EW, Looi A. Bilateral orbital subperiosteal hemorrhage following labor. *Ophthalm Plast Reconstr Surg*. 27(3): 59-63, 2011.

Key Words: Orbital Hemorrhage, Subperiosteal Hemorrhage, Pregnancy, Orbitotomy

Financial Disclosure: The authors had no disclosures.

Poster 60

Intraorbital Optic Nerve Enhancement on Magnetic Resonance Imaging in Postoperative Ischemic Optic Neuropathy

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Introduction:

Postoperative ischemic optic neuropathy is a rare but devastating complication of prolonged surgical procedures. It is often bilateral and in cases of posterior ischemic optic neuropathy (PION), the optic nerves initially appear normal and no afferent pupillary defect may be evident. Magnetic resonance imaging (MRI) of the brain and optic nerves is normal in most cases of PION, which helps distinguish it from giant cell arteritis or optic neuritis, but does not support the clinical diagnosis.

Methods:

We report a case of bilateral PION after abdominal surgery with magnetic resonance imaging (MRI) demonstrating restricted diffusion within both optic nerves.

Results:

A 62 year-old female was complaining of decreased vision following multiple surgeries for mesenteric ischemia. The patient's vision was no light perception (NLP) in the right eye and hand motion (HM) at 1' in the left eye. Both pupils were nonreactive and no relative afferent pupillary defect was noted. Fundus examination was normal. T1- and T2 weighted MRI imaging of the brain was normal, but diffusion weighted imaging (DWI) revealed areas of bright signal, indicating infarction within the posterior aspect of both intraorbital optic nerves.

Conclusion:

Postoperative PION is a difficult clinical diagnosis, especially when bilateral. The posterior segment of the intraorbital optic nerve is particularly susceptible to ischemia. In acute ischemia, the development of cytotoxic edema causes a shift of water molecules from the extracellular to the intracellular space, resulting in a bright signal on DWI within minutes of the event. In contrast to infarctions within the brain parenchyma, where DWI is used routinely to locate areas of ischemic damage, in PION, MRI changes have only been described in a few cases. In our patient, we were able to support the clinical diagnosis and confirm the bilateral optic nerve infarction using DWI.

References:

1. Newman, NJ. Perioperative Visual Loss After Nonocular Surgeries. *Am J Ophthalmol* 2008; 145(4): 604-610.
2. Purvin, V. Kuzma, B. Intraorbital Optic Nerve Signal Hyperintensity on Magnetic Resonance Imaging Sequences in Perioperative Hypotensive Ischemic Optic Neuropathy. *J Neuro Ophthalmol* 2005; 25: 202-204.
3. Sadda, SR. Nee, M. Miller, NR et al. Clinical Spectrum of Posterior Ischemic Optic Neuropathy. *Am J Ophthalmol* 2001; 132: 743-750.

Key Words: Ischemic Optic Neuropathy, Magnetic Resonance Imaging

Financial Disclosure: The authors had no disclosures.

Poster 61

Etiology and Prognosis of Fourth Cranial Nerve Palsy

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Introduction:

Although there are previous studies investigating the etiology and prognosis of fourth cranial nerve palsy, most are small studies from the pre-MRI era which often included other cranial nerve palsies. We performed a large, retrospective, post-MRI era study with more detailed comparison by age than previous studies.

Methods:

All records coded as fourth cranial nerve palsy in our database from 1995 to 2011 were reviewed. We developed criteria to help differentiate congenital cases from longstanding and other idiopathic causes.

Results:

Of 952 records reviewed, there were 629 confirmed cases of fourth cranial nerve palsy. Age of onset ranged from birth to 95 years. According to our criteria, 28% of cases were longstanding (177/629), and 22% were congenital (136/629) accounting for 50% of cases overall (313/629). No congenital or longstanding cases showed significant recovery. Of the remaining 316 acquired cases, microvascular ischemia was the most common etiology in 38% (121/316); 28% of microvascular cases were in the 50-64 age group (34/121) and 66% were in the 65+ age group (80/121). 95% of microvascular cases showed recovery. Trauma was the cause in 28% (88/316); 58% of traumatic cases were in the 19-49 age group (46/88), and 74% of traumatic cases showed recovery. Only 2.5% of cases were due to neoplasm (8/316), with 50% recovery. 1.5% were due to demyelination (5/316) with 100% recovery. A further 10% of cases were idiopathic (62/316), and showed 80% recovery. 5% of cases were from various miscellaneous causes (32/629) with mixed recovery.

Conclusion:

Longstanding and congenital cases account for approximately 50% of fourth cranial nerve palsies, and generally show no recovery. This post-MRI era study shows a higher incidence of microvascular than traumatic etiology compared to pre-MRI era studies. Etiology depends on age, but prognosis depends on etiology.

References:

1. Mollan SP, Edwards JH, Price A, Abbott J, Burdon MA. Aetiology and outcomes of adult superior oblique palsies: a modern series. *Eye (Lond)*, 23(3):640-4, 2009
2. Tollefson MM, Mohny BG, Diehl NN, Burke JP. Incidence and types of childhood hypertropia: a population-based study. *Ophthalmology*. 113(7):1142-5, 2006
3. Bianchi-Marzoli S, Brancato R. Third, fourth, and sixth cranial nerve palsies. *Curr Opin Ophthalmol*. 8(6):45-51, 1997
4. Khawam E, Scott AB, Jampolsky A. Acquired superior oblique palsy. Diagnosis and management. *Arch Ophthalmol*. 77(6):761-8. 1967

Key Words: Cranial Nerve Diseases, Prognosis, Diplopia, Ocular Motility Disorders

Financial Disclosure: The authors had no disclosures.

Poster 62

Diffraction Enhanced Imaging of the Leporine Optic Nerve

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Introduction:

Diffraction Enhanced Imaging (DEI) is a novel synchrotron radiation (SR) technique that provides high spatial resolution images without serious radiation exposure. This allows for investigation of micro structures inside weakly absorbing samples, and has potential for finely detailed imaging of the optic nerve.

Methods:

We explored preliminary DEI techniques using explanted previously frozen specimens of rabbit ocular structures including optic nerves and chiasm preserved in 10% formalin. A double crystal monochromator was used to select a very small energy band from the polychromatic synchrotron beam. This monochromatic beam was passed through the samples, and an analyzer crystal similar to that in the monochromator was placed after the samples.

Results:

The analyzer crystal was sensitive to the refractive effects within the samples in addition to absorption and scattering effects, thereby creating images of the optic nerve and anterior visual pathway apparatus with various contrast properties. The silicon crystal used in DEI was selected for refraction and apparent absorption, and by changing the angle of the crystal, various images showed either apparent absorption or refraction properties.

Conclusion:

Synchrotron radiation techniques such as DEI can be used to provide images of the minute structures of the optic nerve, with minimal radiation exposure. We are the only synchrotron research group in the world attempting to image the optic nerve using DEI, and the work is highly experimental. We will continue to refine our methods and techniques of image acquisition, with a view towards in vivo imaging for the development of live animal models of various optic neuropathies.

References:

1. Kelly, M., Dustin, J., Coupal, R., Beavis, C., Schultke, E, et al. Diffraction-enhanced imaging of a porcine eye. *Canadian Journal of Ophthalmology*, 42 (5),731-733, 2007
2. Kim, B.I., Kim, KH., Youn, H.S., Jheon, S., Kim, J.K. et al. High resolution X-ray phase contrast synchrotron imaging of normal and ligation damaged rat sciatic nerves. *Microcopy and Research Technique*,71:443-447,2008
3. Gao, X., Liu B., Xu, M., Yaun, Q., Gao, X., et al. A micro-tomography method based on X-ray diffraction enhanced imaging for the visualization of micro-organs and soft tissues. *Computerized Medical Imaging and Graphics*, 30 (6-7), 339–347, 2006

Key Words: Optic Nerve, Optic Nerve Diseases, Synchrotrons, Diagnostic Imaging, Radiation

Financial Disclosure: Edward J Atkins - Dr Atkins is supported by a Saskatchewan Health Research Foundation New Investigator Grant. The remaining authors had no disclosures.

Poster 63

Chronic Progressive External Ophthalmoplegia Related to Long-Term Highly Active Antiretroviral Therapy in HIV-Infected Patients

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Introduction:

The purpose of this study is to report a unique chronic progressive external ophthalmoplegia (CPEO)-like disorder in HIV infected patients undergoing long-term treatment with highly active antiretroviral therapy (HAART), and to further describe factors that may provide insight into the pathogenesis.

Methods:

Five adult patients with symptomatic HIV infection who have been treated with HAART for more than 10 years underwent ocular motility examination. High-resolution orbital magnetic resonance imaging (MRI) was performed.

Results:

All patients presented with bilateral progressive external ophthalmoplegia and blepharoptosis. Acetylcholine receptor antibody titers were not elevated, and none of the patients met criteria for the diagnosis of myasthenia gravis. Electrocardiograms were all normal. The remainder of the ophthalmologic examination was normal for all patients. Brain MRI was unremarkable. Orbital MRI showed "spongiform" bright signal inside the extraocular muscles that had conserved volume. The findings on the orbital MRI have been previously described in patients with CPEO. The HAART regimen for all patients included a nucleoside reverse transcriptase inhibitor, which is known to interfere with mitochondrial DNA polymerase gamma. Mutations in mitochondrial DNA polymerase gamma are also known to cause CPEO.

Conclusion:

Patients under long-term HAART may develop functional abnormalities of structurally normal extraocular muscles similar to that observed in chronic progressive external ophthalmoplegia. The use of nucleoside reverse transcriptase inhibitors may have caused interference with mitochondrial DNA polymerase gamma, a polymerase in which mutations are known to be associated with CPEO. This unique constellation of signs and symptoms provides insight into the mechanism behind a CPEO-like syndrome in HIV infected individuals on HAART.

References: None

Key Words: strabismus, ophthalmoplegia, human immunodeficiency virus

Financial Disclosure: The authors had no disclosures.

Poster 64

Third Ventricular Width and Retinal Nerve Fiber Layer Thickness in Multiple Sclerosis.

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Introduction:

Optical coherence tomography (OCT) has been used in multiple sclerosis (MS) to provide a surrogate marker for axonal loss. Radiographic changes in MS include an increase in the maximum width of the third ventricle. We postulated that there is a correlation between third ventricular width and thickness of the retinal nerve fiber layer (RNFL), and between third ventricular width and total macular volume in patients with MS.

Methods:

The average RNFL thickness and total macular volume from 58 eyes in patients with MS and no history of optic neuritis were examined using Stratus OCT. All patients had brain MRI performed within six months of OCT. The maximum width of the third ventricle in these subjects was recorded using axial MRI sequences.

Results:

The Pearson correlation suggesting a relationship between RNFL thickness and maximum axial third ventricular width was -0.449, with a p value <0.0001 that a correlation exists between the two variables; Spearman's rho was -0.366 (p <0.005). For macular thickness and third ventricular width, the Pearson correlation was -0.270, (p value <0.05; Spearman's rho was 0.214 (p 0.106).

Conclusion:

There appears to be a correlation between RNFL thickness and third ventricular maximal width in patients with MS. As the RNFL thickness decreases, there is an increase in the maximal width of the third ventricle. Similarly, as total macular volume decreases, there is an increase in third ventricular maximum width. This correlation may be valuable in future MS research, particularly because unconventional MRI sequences or analysis software is not required.

References: None

Key Words: Optical coherence tomography, Multiple sclerosis, MRI

Financial Disclosure: The authors had no disclosures.

Poster 65

The Syndrome of Infantile-Onset Saccade Initiation Delay (Congenital Ocular Motor Apraxia)

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Introduction:

Infantile-onset saccade initiation delay (ISID) is a defect in initiating volitional saccades. Several abnormalities have been reported in this condition including developmental delay and ataxia. The aim of this study is to quantify these abnormalities.

Methods:

Detailed review of the English medical literature between 1952 and 2010 revealed 383 patients (67 studies) with possible ISID. Patients with inadequate information, Joubert syndrome, neurodegenerative disorders or acquired SID were excluded whenever possible. Details on the remaining 325 patients (age range 2.5 months-38 years, 173 males) were analyzed. Six percent were siblings, 1.5% parent-child, and 1.2% monozygotic twins. The percentage of abnormalities in all patients was reported to estimate their minimum prevalence.

Results:

Head thrusts were reported in 80.6% of patients, 91% of whom were horizontal and bilateral. Blinks without head thrusts were used to initiate saccades in 47%. Spontaneous saccades were present in 17.2%. Impairment of smooth ocular pursuit was reported in 37.5%, strabismus in 15.7%, and nystagmus in 10.2%. Deficits in the fast phases of the vestibulo-ocular reflex and optokinetic response were reported in 41.2% and 72.6% respectively. Hypotonia was reported in 38.8%, developmental delay in 45.2%, cognitive delay in 15.4%, speech/ language delay in 41.2%, reading difficulties in 8%, motor delay in 53%, and ataxia/ clumsiness in 53%. Neuroimaging (93.2% CT/MRI/ or both) was done on 234 patients. Imaging was normal in 32.9%. Infratentorial abnormalities were reported in 40.2%, cerebrum in 18.8%, and corpus callosum in 7.7%. Sensitivity analysis on 288 patients had minimal impact on the results (-6.8% maximum).

Conclusion:

Significant neuro-ophthalmological, neurological, and neuroradiological abnormalities occur commonly in patients with ISID. This suggests that ISID is a neurological syndrome rather than primarily a disorder of saccadic initiation. The high prevalence of various abnormalities in these patients is consistent with widespread brain dysfunction.

References: None

Key Words: Eye movements, Saccades, Ocular motor apraxia, Clinical spectrum of condition, Children

Financial Disclosure: The authors had no disclosures.

Poster 66

Blink Rates in Children During Different Types of Eye Movements

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Introduction:

The spontaneous blink rate (SBR) is variable in humans. It increases rapidly during childhood before reaching a plateau in adulthood at 10-20 blinks/minute. The SBR depends on the ocular surface status, mental activity, cognitive and emotional factors, and the presence of specific neurologic and psychiatric diseases. Our aim was to quantify and compare the SBR during visual fixation and different types of eye movements in healthy children.

Methods:

Thirty-nine normal participants (M=21, mean age (SD) 13.6(3.5) years), made horizontal and vertical visually-guided saccades to 5-15° target steps, tracked a horizontal and vertical visual target moving in a sinusoidal and smooth fashion at 15.5 and 31°/s peak target velocities, performed horizontal and vertical active sinusoidal head rotations in light and darkness at 0.5 and 2Hz while looking straight ahead (VOR task) or fixated straight ahead and in four directions of gaze. The eyes of each participant were video-taped and reviewed twice to determine the SBR. Parametric and non-parametric tests were used to analyze the data.

Results:

The median SBR during fixation straight ahead was 7 blinks/ minute (range 2-20.5), which was similar to the median SBR during fixation in four directions of gaze, horizontal saccades and horizontal smooth pursuit tasks. The median SBR during vertical smooth pursuit and vertical saccades was significantly lower than during fixation ($p \leq 0.042$). The median SBR during the VOR in light (vertical and horizontal) and dark (horizontal) was significantly higher than during fixation ($p = 0.019-0.024$).

Conclusion:

The SBR during visual fixation was lower than that reported previously in young children (14 blinks/minute), and even lower during vertical smooth pursuit and saccades tasks. This may be due to differences in concentration required for visual fixation in general and vertical visual tasks more specifically. The higher SBR during the VOR may have been caused by drying of the eyes during head shaking.

References: None

Key Words: Blink rate, Children, Eye movements

Financial Disclosure: The authors had no disclosures.

Poster 67

Development of Increased Vertical Fusional Amplitudes in Patients with Thyroid Eye Disease

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Introduction:

Increased vertical fusional amplitudes are considered the sine qua non of congenital fourth nerve palsies, and not thought to be present in cases of acquired diplopia. We present 9 cases of patients with thyroid eye disease (TED) who developed increased vertical fusional amplitudes over the course of their disease, in several cases leading to resolution of their diplopia. In reviewing both standard texts and a Pubmed search we found no other similar reported cases.

Methods:

Review of charts from neuro-ophthalmology practice at a tertiary referral center. The diagnosis of TED was made on clinical criteria (proptosis, lid retraction, lid lag) and neuroimaging.

Results:

Of the 9 cases reviewed, all were female with ages ranging from 43-81 years. All presented with persistent or intermittent diplopia and one also complained of vision loss. Duration of disease was variable from 6 months to 10 years. One was described only as "years" duration. Vertical fusional amplitudes ranged from 5-25 diopters. Two of the cases developed a head posture which helped them fuse. Prisms were recommended during the course of disease in three cases but were only utilized in two cases.

Conclusion:

From these cases it can be seen that large vertical fusional amplitudes can develop over time in patients with TED. One thought is that these patients had a head posture in which they could fuse, allowing them to develop increased fusion and increase their range of single binocular vision. As the most common longstanding acquired diplopia, versus ischemic cranial nerve palsies which resolve quickly, this gives insight into the plasticity of the vergence system and suggests that perhaps prism therapy and longer followup would reduce the number of patients who need surgical correction or reduce the amount of surgery needed.

References: None

Key Words: Thyroid, Diplopia, Vergence, Vertical, Fusion

Financial Disclosure: The authors had no disclosures.

Poster 68

Use of the Hess screen in diplopia evaluation.

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Introduction:

Quantitation has been an important part of afferent system function analysis since the introduction of Snellen's optotypes in 1862. Little data exists on quantitative assessment of ocular motility. Nine cardinal position measurements are standard, but require an orthoptist, and are often poorly reproducible unless standardized direction. In 1908, Walter Rudolph Hess used red green glasses to separate out the movements of the two eyes. Variations of this concept include the Lancaster Red-green test, and Lee's test. These have been available for more than 100 years although tend to be under utilized.

Methods:

Retrospective review of patients presenting with diplopia, orbital pathology, or cranial nerve palsies and evaluated with a Hess screen. The role of the Hess screen was analyzed in diagnosis, quantitative follow up, and surgical planning.

Results:

A total of 2,000 patients over the last 10 years were reviewed. Not all patients with diplopia were able to do Hess screen although the vast majority were. Patients were analyzed for specific application. Pattern deviation was often helpful in establishing a diagnosis. Quantitative assessment was particularly useful in following patients with both restrictive and paretic deviations, and surgical planning could be aided. The Hess screen does not distinguish between restrictive and paretic patterns; thus forced ductions or intraocular pressure elevations in eccentric gaze are still a part of evaluation.

Conclusion:

Other than the teaching value of the Hess screen in allowing presentations to indicate separate movements of the two eyes, the particular usefulness of the Hess screen falls into three categories: 1. Pattern recognition (often aiding in the diagnosis). 2. Quantitative assessments of ocular motility (allowing for a detailed study of natural disease history or response to treatment). 3. Surgical planning (Hess screen is often a major advantage in deciding on surgery in the contralateral eye to maximize binocularity).

References:

1. Hess WR. Ein einfaches messendes Verfahren zur Motilitätsprüfung der Augen. Zeitschrift Fur Augenheilkunde. 1916; 15: 201-219.
2. Newman SA: The use of the Hess screen in the evaluation of diplopia. Presented at the 147th Meeting of the American Ophthalmology Society Meeting, Greenbrier West Virginia May 2010.

Key Words: ocular motility, quantitation, diplopia, restrictive strabismus, cranial nerve palsies

Financial Disclosure: The author had no disclosures.

Poster 69

Retrospective analysis of ocular myasthenia gravis patients in Kuwait

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Introduction:

The statistics of ocular myasthenia gravis is fairly well established in western literature. This study was aimed to examine the rates of conversion of ocular myasthenia to generalized myasthenia in Kuwait and their response to treatment.

Methods:

Case files of myasthenia patients who were seen in a major neurology/ophthalmology centre in Kuwait were collected and those presenting with ocular myasthenia were identified. These cases were then analysed and their rates of conversion to generalized myasthenia (if any) were determined; in addition to other parameters such as their response to various forms of treatment.

Results:

Forty-two patients with myasthenia were identified, out of these twenty-six (56%) presented with ocular myasthenia with a mean follow up of 46 months. Of the twenty-six 19% converted to generalized myasthenia, 57% received steroids, 34% went on to receive immune-modulatory therapy.

Conclusion:

Our retrospective analysis of a small number of ocular myasthenia patients revealed a 19% conversion rate to generalized myasthenia. However a larger, and ideally, a prospective analysis should be done in order to better document the rates of conversion to generalized myasthenia and if immune therapy affects this rate.

References: None

Key Words: Myasthenia gravis, Conversion, ocular, treatment

Financial Disclosure: The authors had no disclosures.

Poster 70

Results Of Performing Extraocular Muscle Surgery On Patients Diagnosed With Myasthenia Gravis

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Introduction:

The purpose of this study was to report the postoperative results of strabismus surgery on patients with stable myasthenia gravis (MG).

Methods:

A retrospective review of the records of patients diagnosed with MG who underwent strabismus surgery was performed. Analysis included age, disease stability, strabismic deviation, diagnostic test results, surgical procedures, and surgical results. Final ocular alignment was determined at the last follow-up.

Results:

Nine patients (44% male) were included with age at first surgery of 49.9 years (range 9.7-74.3) and post-operative follow-up of 5.7 years (range 0.7-10.7). Two patients had horizontal strabismus alone, three had vertical strabismus alone, three had vertical and horizontal strabismus, and one patient had vertical and torsional strabismus. For patients for whom stability of measurement data was available (n=8) the pre-operative length of ocular alignment stability was 1.0 ± 1.3 years. Mean preoperative horizontal and vertical deviations were 27 ± 32.7 and 25.6 ± 36.7 PD, respectively. All surgeries were performed with adjustable sutures. Average length of follow-up after the first surgery was 5.7 ± 4.2 years. Mean final horizontal and vertical deviations were 5.7 ± 12.0 (p=0.07, compared to pre-operative mean horizontal deviation) and 5.2 ± 4.7 PD (p=0.1, compared to pre-operative mean vertical deviation), respectively. Success (deviation within 10 PD of orthotropia) was achieved in 8 (89%) of patients after one surgery. Four (44%) of these patients developed a second deviation. Five patients (56%) underwent a second surgery. Time to second operation ranged from 0.4 to 5.0 years. All patients achieved success after two operations. Two patients (22%) at the final visit subsequently had treatment failure.

Conclusion:

Patients with quiescent MG undergoing strabismus surgery can successfully achieve good alignment, and can maintain good alignment over time. However these patients may require re-operation to achieve success.

References: None

Key Words: strabismus, myasthenia gravis, adjustable sutures, ocular alignment

Financial Disclosure: The authors had no disclosures.

Poster 71

Ocular torsion and morphological anomalies of semicircular canals

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Introduction:

Ocular torsions (OT) are closely related to dysfunctions of the oculo-labyrinthic system: peripheral acquired lesions or surgery of labyrinths induce cyclotorsion. We observed that many patients suffering from scoliosis can also be affected with OT and semi-circular canals (ducts) anomalies. The aim of this study is to establish a feasible lateralization of OT versus canal anomalies in a set of 50 scoliotic patients.

Methods:

The 50 selected patients had ophthalmologic examination and OT measurement with SLO or RNM. Each had MRI and 3D modeling of the fluid inside the labyrinthic ducts. We compared the sided OT to the side and number of affected ducts. OT was evaluated following two criteria: absolute value of the OT angle on right and left eye referred to normal values and difference of the angle value between right and left eye.

Results:

Compared to normal population, means of both criteria of evaluation were increased. Patients with several affected ducts were the ones with highest OT and the side of OT was correlated with the side of the number of affected ducts. The lateral duct was the most frequently affected and with the most abnormal shape. Statistically, compared to a normal group, in case of unilateral anomaly, OT is different ($p < 0.05$)

Conclusion:

These first results on 50 patients show a significant relationship between ducts anomalies chiefly located on lateral duct and OT. The implication of these two factors in scoliosis could be explained through an abnormal direct vestibulospinal output added to consequences of OT in vertical visual perception. But we have also to take into account the role of growth and the complexity of equilibrium networks in the brain

References: None

Key Words: Ocular torsion, Oculo labyrinthic system, Semi circular canals, Scanning Laser Ophthalmoscopy, Non Mydriatic Retinography

Financial Disclosure: The authors had no disclosures.

Poster 72

The Incidence of Rebound Nystagmus in Normal Subjects Under Different Physiologic Conditions

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Introduction:

Although originally thought to be always pathologic, rebound nystagmus (RN) has more recently been reported to be present in normal subjects when recorded in the dark. This experiment was done to determine the incidence of RN in different conditions including in the dark and while focusing either at near or at distance in the light.

Methods:

Eye movements were measured in eighteen normal subjects while fixating for 40 seconds in primary position, extreme gaze, primary position, opposite extreme gaze and again in primary position. It was done in the light using a distant (10') target by nine subjects and at near (3') target by 13 subjects as well as in the dark by 11 subjects. The average number of trials for each person was 2.5 (range 1 –9). Not all subjects did the testing under all conditions. A trial was considered positive for RN if at least 5 beats of nystagmus were recorded after return to primary position.

Results:

When tested in the light with a near target one patient had no RN after gaze to the left on two trials but both times had RN after right gaze. 12 others had RN from either side on all 37 trials. For dark trials RN was present only unilaterally in 3 patients (two trials each) but was bilateral in 8 others on each of 17 trials. At distance a single person had no RN to either side on either of two trials. Two others had only unilateral RN and five had bilateral RN on all of 14 trials. The amplitude of the nystagmus was the same in all conditions with an average range of 0.8-2.0 degrees

Conclusion:

RN can be consistently seen in normal individuals following extreme gaze. Further testing needs to be done to identify features that distinguish physiologic from pathophysiologic rebound nystagmus.

References: None

Key Words: rebound nystagmus

Financial Disclosure: The authors had no disclosures.

Poster 73

Isolated Unilateral Ptosis due to Mesencephalic Stroke

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Introduction:

Since the work of Warwick in 1953, the anatomy of the oculomotor nucleus, its sub nuclei arrangement and the fascicles within the midbrain has been a source of extensive investigation and debate.

The location of the oculomotor fascicles has been derived from correlation of individual deficits and the midbrain lesions identified by neuroimaging.

To the best of our knowledge, there is no earlier documentation in the literature of isolated unilateral ptosis due to midbrain infarct.

Methods:

Clinical-radiologic correlation.

Results:

A 69y/o man with hypertension, diabetes type II, dyslipidemia, and coronary artery disease, who had a partial occlusive thrombus within the distal left vertebral artery that produced acute infarcts in the left cerebellar hemisphere, right dorsal midbrain tegmentum and right thalamus.

Neurological examination revealed partial third nerve palsy with isolated complete ptosis of the right eye, Extra ocular movements including adduction, supraduction, infraduction and inferior oblique function were all intact. The left eye was not ptotic. The pupils were symmetric in size and pupillary reflex intact bilaterally.

Conclusion:

This is a unique case and we concluded that the isolated unilateral right lid ptosis was due to an ipsilateral fascicular lesion located in the levator palpebrae fascicle after the axons exited from its sub nucleus in the midline.

The location of the Magnetic Resonance imaging (MRI) lesion and the isolated unilateral ptosis supports Warwick's model that proposes the levator palpebrae nucleus is located on the central caudal aspect of the mesencephalon. Additionally, the unilateral deficit is also consistent with the topographical fascicular model proposed by Ksiazek and colleagues.

References:

1. Castro O, Johnson LN, Mamourian AC. Isolated inferior oblique paresis from brain-stem infarction. Perspective on oculomotor fascicular organization in the ventral midbrain tegmentum. *Arch Neurol* 1990; 47: 235–237.
2. Ksiazek SM, Slamovits TL, Rosen CE, Burde RM, Parisi F. Fascicular arrangement in partial oculomotor paresis. *American Journal of Ophthalmology* 1994; 118: 97–103.
3. Martin TJ, Corbett JJ, Babikian PV, Crawford SC, Currier RD. Bilateral ptosis due to mesencephalic lesions with relative preservation of ocular motility. *J Neuro-ophthalmol* 1992; 16:258-263.
4. Saeki N, Murai H, Mine S, Yamaura A. Fascicular arrangement within the oculomotor nerve MRI analysis of a midbrain infarct. *J Clin Neurosci*. 2000;7:268-270.
5. Warwick R. Representation of extra-ocular muscles in oculomotor nuclei of monkey. *J Comp Neurol* 1953; 98: 449–495.

Key Words: Midbrain stroke, Partial third nerve palsy, Unilateral ptosis

Financial Disclosure: The authors had no disclosures.

Poster 74

Brain Activation Pattern Indicates Intentional Saccadic Oscillation in Voluntary Nystagmus: A PET Study

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Introduction:

The mechanisms and involved brain areas remain unknown in voluntary nystagmus that is characterized by volitional bursts of small back-to-back saccades without intersaccadic intervals. The aim of this study was to identify the brain areas involved in the generation of voluntary nystagmus using metabolic network pattern analysis of F-18-fluorodeoxyglucose (FDG) brain PET.

Methods:

Seven soldiers (all men, age range=18-25, mean age=21) with voluntary nystagmus underwent 3-dimensional recording of eye movements, EEG, and MRI and F-18-fluorodeoxyglucose (FDG)-PET of the brain. FDG-PET images of 13 age-matched healthy men were used as normal controls. Characteristic brain metabolic network was investigated using scaled subprofile model/principal component analysis (SSM/PCA). Additionally, regional metabolic difference between the two groups was determined using statistical parametric mapping analyses (SPM, unpaired *t*-test).

Results:

The ocular oscillation was mostly in the horizontal plane, with a mild facial grimacing and efforts of convergence. Brain MRIs were all normal. SSM/PCA analysis revealed a significant spatial covariance pattern that discriminated the patients with voluntary nystagmus from the controls ($p < 0.05$). This voluntary nystagmus-related pattern accounted for 17% of the subject X voxel variance and is characterized by hypermetabolism in the bilateral prefrontal cortices, precuneus, cuneus, and posterior cingulate cortices. In contrast, metabolic activity was relatively reduced in the bilateral cerebellum. SPM analyses ($p < 0.005$, uncorrected) also disclosed increased metabolism in the bilateral superior frontal and parietal cortices, precentral gyri and right precuneus.

Conclusion:

The characteristic metabolic network pattern supports that voluntary nystagmus is intentional saccadic oscillation. PET may provide an important clue in differentiating voluntary nystagmus from more devastating ocular flutter/opsoclonus, especially in cases with financial compensation or legal/military mitigation.

References: None

Key Words: Voluntary nystagmus, Saccadic oscillation, Metabolism, PET

Financial Disclosure: The authors have no disclosures.

Poster 75

Acute Saccadic and Supranuclear Palsy, Case Presentation with Pathologic Evaluation and Literature Review

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Introduction:

Selective saccadic palsy (SSP) and a “PSP-like phenotype” (PSP-I) have both been reported following cardiac and aortic procedures. Pathologic and MRI evidence has been reported demonstrating lesions in the dorso-medial pons in SSP (1,2) and bilateral basal ganglia and pons in PSP-I (3). We present a case of impaired pursuits, volitional saccades, dysarthria and imbalance following cardiac surgery with autopsy evaluation.

Methods:

A 78-year-old male underwent scheduled aortic valve replacement with post op complications of atrial fibrillation and cardiac tamponade. Following extubation he demonstrated a supranuclear ophthalmoparesis with absent voluntary saccades and pursuits. His speech became sparse and dysarthric and he required assistance to walk. Forty-four days following his initial surgery he died due to respiratory compromise. An autopsy was performed.

Results:

Pathologic evaluation identified subacute right thalamic and remote right frontal lacunar infarcts. No lesion was identified in the pons or midbrain, including the previously reported dorso-medial pons. Lewy bodies were found in the substantia nigra, locus ceruleus and vagus motor nucleus.

Conclusion:

SSP and PSP-I present with variable features of impaired saccades, supranuclear palsy, dysarthria and imbalance. These may represent a spectrum of the same process or distinct processes. The absence of pontine lesion in this case is consistent with the latter.

References:

1. Hanson MR, Hamid MA, Tomsak RL, Chou SS, Leigh JR. Selective Saccadic Palsy Caused by Pontine Lesions: Clinical, Physiologic and Pathological Correlations. *Ann Neurol* 1986;20:209-217
2. Eggers SDZ, Moster ML, Cranmer K. Selective Saccadic Palsy After Cardiac Surgery *Neurology*, 2008 Jan 22;70(4):318-20.
3. Kim HT, Shields S, Bhatia KP, Quinn N. Progressive Supranuclear Palsy-like Phenotype Associated with Bilateral hypoxic-Ischemic Striopallidal Lesions. *Mov Disord* 2005 Jun;20(6):755-7.

Key Words: Selective Saccadic Palsy, Asaccadia, Progressive Supranuclear Palsy

Financial Disclosure: Eric Eggenberger: grants or consulting for Biogen, Teva, Serono, Bayer, Allergan, Novartis. The remaining authors had no disclosures.

Poster 76

Ophthalmoplegia associated with polymyositis/dermatomyositis

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Introduction:

Polymyositis is an idiopathic inflammatory myopathy that with cutaneous findings is called dermatomyositis. Associated ophthalmoplegia is rare with only one confirmed reported case in the English literature. We describe two additional patients.

Methods:

Case 1 is a 49 year old woman with a persistent skin rash and diplopia secondary to a variable exotropia and hypertropia. Case 2 is a 50 year old woman with weight loss and diplopia secondary to a pseudo intranuclear ophthalmoplegia. Neither women had orbital inflammatory signs or symptoms.

Results:

Both patients had an elevated ANA and creatine kinase, and a muscle biopsy consistent with polymyositis/dermatomyositis. Case 1 was treated with prednisone, chloroquine and methotrexate, while case 2 with prednisone and azathioprine. Both patients had subsequent resolution of the diplopia.

Conclusion:

We describe two additional patients who developed ophthalmoplegia secondary to polymyositis/dermatomyositis. Treatments of their underlying systemic illness lead to resolution of the diplopia.

References:

1. Susac, Garcia-Mullin, Glaser, Ophthalmoplegia in dermatomyositis, Neurology, 23, 305, 1973

Key Words: Ophthalmoplegia, Polymyositis/dermatomyositis, Diplopia, Pseudo intranuclear ophthalmoplegia, Exotropia

Financial Disclosure: The authors had no disclosures.

Poster 77

Increasing interval between injections after prolonged onabotulinum toxin type A use for blepharospasm

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Introduction:

We have been using onabotulinum toxin type A (BTA) to treat blepharospasm (BEB) for over 25 years and noted that some patients seem to have an increase in efficacy over time as seen by longer intervals between injections. The change in duration of response to therapy has been described in cervical dystonia and hemifacial spasm. There is limited data regarding changes in dosing intervals after prolonged BTA use in patients with BEB.

Methods:

We retrospectively reviewed the charts of patients that continued to follow in our clinic and were receiving BTA for BEB. Clinical information from the initial and most recent injections were recorded into a database including: sex, age at the start of treatment, current age, initial frequency of injections, current frequency of injections.

Results:

We identified 9 cases where the interval between injections had changed. The average age at first injection was 59.8 years and average current age was 74.4 years. Two patients were male and 7 were female. Average duration of toxin use was 15 years (range=8-26 years). The average dosing interval at the start of treatment was 2.8 ± 0.4 months, which increased to 5.0 ± 1.3 months ($p < 0.001$). The average duration of toxin use prior to the increase in dosing interval was 9.8 ± 5.2 years ($n=6$); however, this data is limited because of variable access to records over 10 years old.

Conclusion:

The dosing interval for BTA may increase over time in patients treated for BEB. The change in dosing interval may occur more readily in BEB patients treated with BTA for approximately 10 years.

References:

1. Mejia N, Vuong K, Jankovic J. Long-term botulinum toxin efficacy, safety, and immunogenicity. *Movement Disorders* 2005; 20: 592-97.
2. Naumann M, Albanese A, Heinen F, et al. Safety and efficacy of botulinum toxin type A following long-term use. *European Journal of Neurology* 2006; 13: 35-40.

Key Words: Botulinum toxin A, Blepharospasm

Financial Disclosure: The authors had no disclosures.

Poster 78

Bilateral abducens nerve palsy as a manifestation of Epstein Barr virus (EBV) encephalitis in children

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Introduction:

Clinical features of EBV encephalitis are diverse. However, the involvement of ocular motility is rare. We report 2 cases of bilateral abducens nerve palsy associated with EVB encephalitis in children.

Methods:

Case report

Results:

A 14-month-old boy presented with fever and esodeviation of left eye. Forty five prism diopters (PD) esotropia was observed by modified Krimsky methods. Duction and version test revealed limitation of abduction in both eyes. MRI of brain exhibited lesions at right anterior portion of pons, tegmentum and medulla. Serologic test for viral infection showed elevated EVNA, EVB VCA IgG and IgM. PCR from CSF was positive for EBV. He was treated with systemic acyclovir and prednisolone. Four months later, abduction limitation of both eyes was partially improved and he showed 25 PD esotropia in the left eye. A 13-year-old boy presented with headache, fever and altered consciousness. MRI of brain showed meningo-encephalitis at bilateral temporal, frontal, and parietal area. Serologic test revealed elevated EVB EA DR IgM. He complained diplopia and had 30PD esotropia with bilateral limitation of abduction. With supportive care, abduction limitation recovered completely but he had seizure at 4 months follow-up.

Conclusion:

EBV encephalitis can accompany acquired bilateral abducens nerve palsy in children. Abducens palsy and other neurologic sequelae can remain after several months.

References: None

Key Words: Abducens nerve palsy, Epstein Barr virus, Encephalitis, esotropia

Financial Disclosure: The authors had no disclosures.

Poster 79

Downbeat Nystagmus In Patients On Lamotrigine In Combination With Other CNS Active Drugs

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Introduction:

Three patients presented with downbeat nystagmus (DBN), two of whom had severe gait difficulty, while on lamotrigine in addition to other CNS drugs. Despite reports of anticonvulsant - associated with DBN the treating doctors maintained that the DBN and gait disorders were not drug related. One patient was on lamotrigine and lacosamide for seizures, one was on lamotrigine, lithium and duloxetine and the third patient was on lamotrigine, topiramate, amantadine, trileptal and propranolol for migraines.

The first patients' lamotrigine and lacosamide were gradually reduced over a period of months and the DBN disappeared and his gait returned to normal for the first time in two years. The second patients' lamotrigine was stopped and the lithium and duloxetine were unchanged and the DBN disappeared in one week. The third patient had her all medications rapidly reduced and then stopped and her downbeat nystagmus and ataxia resolved between one - two weeks. Anticonvulsants and lithium are known causes of downbeat nystagmus as are structural abnormalities of the cerebellum and posterior fossa such as Arnold-Chiari, syringobulbia, cerebellar tumors, and posterior fossa strokes. Amongst the commonest causes of DBN are hereditary, toxic, metabolic, inflammatory, and paraneoplastic cerebellar degenerations. The cerebellum inhibits vestibular circuits mediating upward but not downward eye movements.¹ With loss of cerebellar inhibition upward drifts of the eyes could occur which would elicit fast downward corrections i.e. downbeat nystagmus.

Methods:

Results:

Conclusion:

Lamotrigine has been associated with DBN. Decreasing lamotrigine alone, or in combination, or even altering the ratio of CNS active medications has been shown to alleviate the symptoms of DBN and ataxia. The etiology of this phenomenon alone is unknown however, CNS receptor inhibition or altered pharmacokinetics leading to elevated serum levels could contribute. This side effect of CNS active medications is essential to identify and recognize.

References:

1. Walsh and Hoyt's clinical neuro-ophthalmology. ed Neil Miller. 6th edition, P1145, 1161
2. O'Donnell, John et al. Lamotrigine overdose in an adult Journal of Clinical Toxicology.2000 pp. 659-660
3. Sun-Young, Oh, et al. Downbeat, Positional, and Perverted Head-Shaking Nystagmus Associated with Lamotrigine Toxicity. Journal of Clinical Neurology. Vol2, December 2006, pp. 283-285.
4. AlKawi, Ammar et al. Downbeat nystagmus as a result of lamotrigine toxicity. Epilepsy Research 63 (2005) 85-88.
5. Arif. H et al. The effect of age and comedication on lamotrigine clearance tolerability, and efficacy. Epilepsia. 2011 Oct;52(10):1905-13. doi: 10.1111/j.1528-1167.2011.03217. x Epub 2011 Aug 12.

Key Words: nystagmus, lamotrigine, CNS active medications, ataxia

Financial Disclosure: The authors had no disclosures.

Poster 80

Painful Ophthalmoplegia Secondary to Chronic Bacterial Infection

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Introduction:

We present a rare and previously unreported case of a 74 year old male who was diagnosed with a left cavernous sinus and Meckel's cave lesion which was initially thought to be a tumor, but which turned out to be an unusual infection with Gram positive cocci in clusters.

Methods:

Case report

Results:

8 years ago, this man presented with what seemed to be an isolated painful left sixth cranial nerve palsy which was attributed to microvascular ischemia. His abduction deficit did not improve and the pain became chronic. Imaging showed a left Meckel's cave and cavernous sinus mass. Partial resection was performed and a chordoma or meningioma was favoured by both radiology and neurosurgery. Pathology was non-diagnostic. The lesion recurred, and invaded the skull base, resulting in a thrombosed jugular vein. A second partial resection for repeat biopsy was done; however cultures were not performed, since the surgical specimen was thought to be tumor and was preserved in formalin to be sent to the Mayo Clinic for diagnostic pathology. The pathology report described an interesting pattern of granulomatous and suppurative inflammation with the presence of colonies of gram positive cocci. Serial imaging has shown that the infectious mass persists, but has decreased in size, and his symptoms are slowly improving on long term antibiotic therapy.

Conclusion:

There are several case reports of cavernous sinus thrombosis and infection associated with ipsilateral sinus disease or Lemierre's syndrome. Invasive infectious lesions involving the cavernous sinus and extending to the skull base are well known, but we are not aware of similar cases presenting as such an indolent process, in the absence of any identifiable pathology other than a focal intracranial mass of Gram positive cocci in clusters, and showing slow improvement on long-term antibiotic therapy over the course of several years.

References: None

Key Words: Infections, Ocular Motility Disorders, Ophthalmoplegia, Cavernous Sinus Syndrome

Financial Disclosure: The authors had no disclosures.

Poster 81

Another AION? Wegener's Presenting as Isolated Optic Neuropathy.

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Introduction:

There are few reports of isolated optic neuritis as an initial presentation of Wegener's granulomatosis.¹⁻⁵ We present a case of an isolated optic neuropathy as the only manifestation of ANCA positive Wegener's.

Methods:

Retrospective case report.

Results:

A 36 year old man awoke with vision loss (< 15 seconds) in his right eye and a headache. His PCP injected steroid which improved the headache. He had two weeks of stuttering episodes of brief vision loss. He then awoke with loss of superior field OD. Vision was 20/25 OD and 20/20 OS. Lab studies included ESR, CRP, ANA, RPR, HLA-B27, all of which were within normal limits. A head and neck MRI and MRA were normal. CSF studies were unremarkable. He returned 4 weeks later with NLP vision OD and 20/20 OS. C-ANCA was positive with significantly elevated myeloperoxidase IgG antibody (92.0 EU/ml; <5.90 EU/ml normal). Treatment was started on 80mg of prednisone for 2 days, then switched to 1gram of IV solumedrol daily for 3 days. He was started on cyclophosphamide and continued on oral prednisone. He also underwent evaluation for respiratory and kidney involvement, with negative chest xray, urinalysis, and normal renal function. After six months of cyclophosphamide and a slow oral prednisone taper, he was switched to mycophenylate. One year after presentation, his vision had improved to 20/40 and automated perimetry had improved. His exam was significant for a RAPD and his fundus exam showed temporal pallor.

Conclusion:

This presentation of Wegener's, limited to an optic neuropathy, describes an insidious onset with dramatic sudden vision loss, followed by a gradual improving course on continued immunosuppressive treatment. Treatment under the guidance of rheumatology gave significant but gradual return of vision.

References:

1. Moubayed SP, Black DO. Optic neuritis as an initial presentation of Wegener's granulomatosis. *Can J Ophthalmol*, 44, e59, 2009.
2. Monteiro ML, Borges WI, do Val Ferreira Ramos C, Lucato LT, Leite CC. Bilateral optic neuritis in Wegener granulomatosis. *J Neuroophthalmol*, 25, 25-8, 2005.
3. Duran E, Merkel PA, Sweet S, Swan N, Babikian VL. ANCA-associated small vessel vasculitis presenting with ischemic optic neuropathy. *Neurology*, 62, 162-163, 2004.
4. Belden CJ, Hamed LM, Macuso AA. Bilateral isolated retrobulbar optic neuropathy in limited Wegener's granulomatosis. *J Clin Neuroophthalmol*, 13, 119-123, 1993.
5. Niskopoulou M, Du Toit N. Optic neuritis as a feature of Wegener's granulomatosis. *Eye*, 1, 320-321, 2002.

Key Words: optic neuropathy, Wegener's, immunosuppression, visual field defect, visual recovery

Financial Disclosure: Louise Mawn, MD grant support from NIH and Research to Prevent Blindness for unrelated projects. The remaining author had no disclosures.

Poster 82

High Dose Botulinum Toxin For Benign Essential Blepharospasm

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Introduction:

Botulinum toxin is a primary treatment for benign essential blepharospasm (BEB). Standard starting doses for BEB range from 25-50 units for both eyes. There is little data available on the use of doses of 90 units or greater. This study aims to evaluate the effectiveness and side effects of repeated, high dose botulinum toxin for treatment of BEB.

Methods:

The charts of consecutive patients treated for benign essential blepharospasm with high-dose botulinum toxin (greater than 90 units for BEB) by one injector were reviewed. Initial starting dose, final dose, number of injections at high dose, degree of symptom relief, and side effects were recorded. To be included patients had to have received high dose injections on at least 4 visits.

Results:

A total of 7 patients were treated with repeat doses of high dose botulinum toxin during this time period. These patients had received a range of 7 to 41 repeat injections over periods of 1.5 to 10 years. Doses of botulinum toxin at the most recent treatment ranged from 92.5 to 135 units. The most common adverse effect was ptosis. 3 patients reported upper lid ptosis, with the right eye affected in 5 instances and the left eye affected in 3 instances. The treatment produced good resolution of blepharospasm symptoms with 6 out of 7 patients reporting excellent relief and 1 reporting moderate improvement of symptoms. All patients had started at our standard dose of 32.5 units and been gradually titrated to their current dosage.

Conclusion:

High dose botulinum toxin for blepharospasm is effective with minimal adverse effects in patients who do not respond adequately to standard doses.

References:

1. Levy RL, Berman D, Parikh M, Miller NR, Supramaximal doses of botulinum toxin for refractory blepharospasm, *Ophthalmology*, 113, 1665-8, 2006.
2. Pang AL, O'Day J, Use of high-dose botulinum A toxin in essential blepharospasm: is too high too much?, *Clin Experiment Ophthalmol*, 34, 441-4, 2006.

Key Words: Benign Essential Blepharospasm, Botulinum Toxin, High Dose

Financial Disclosure: The author had no disclosures.

Poster 83

Mechanical Oscillopsia After Lower Eyelid Blepharoplasty With Fat Repositioning

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Introduction:

Blepharoplasty with fat repositioning is a technique used to fill the tear trough in the aging lower eyelid. We present a patient who underwent transcutaneous lower eyelid blepharoplasty with fat repositioning and subsequently developed mechanical oscillopsia exacerbated by movement of the face.

Methods:

A Retrospective Case Report.

Results:

A 71-year-old female complained of distorted vision, present for two years. This symptom began immediately after a four eyelid blepharoplasty. The lower lid fat had been secured with sutures over the infraorbital rim. The image from the right eye was moving, consistent with vertical oscillopsia exacerbated by lower facial movement. Medical history was non-contributory. She was not on any medications. She sought numerous ophthalmic, neurologic and plastic surgery consultations without explanation or treatment for her symptom.

Examination demonstrated visual acuity of 20/30 OD and 20/25 OS and full color plates OU. Confrontational visual fields were full OU. No RAPD was present. Ductions were full. Maddox rod testing demonstrated 3 degrees of excyclotorsion and vertical deviation; there was increased displacement of the double Maddox rod lines with cheek movement. External exam did not show ptosis, lagophthalmos or an eyelid movement abnormality. Cranial nerve examination was otherwise normal. Review of all prior extensive imaging was unremarkable. There was suspicion for incarceration of the inferior oblique in the anterior lamella closure of the right eye. Orbitotomy was performed and multiple areas of cicatrix were released in the right lower lid. Our patient immediately noted amelioration of her visual distortion and at > 3 month follow-up remains asymptomatic.

Conclusion:

Our patient developed cicatricial tethering of the inferior oblique muscle to the superficial musculoaponeurotic system as a complication of transcutaneous lower eyelid blepharoplasty with fat repositioning. Excision of the scar bands led to immediate amelioration of symptoms. Neuro-ophthalmologists need to consider this anatomical explanation for visual disturbance after eyelid surgery.

References:

1. Hamra ST. Arcus marginalis release and orbital fat preservation in midface rejuvenation, *Plast Reconstr Surg*, 96:354-362, 1995.
2. Goldberg RA. Transconjunctival orbital fat repositioning: transposition of orbital fat pedicles into a subperiosteal pocket, *Plast Reconstr Surg*, 105:743-748; discussion 749-751, 2000.
3. Goldberg RA, Yuen VH. Restricted ocular movements following lower eyelid fat repositioning. *Plast Reconstr Surg*, 110:302-305; discussion 306-308, 2002.
4. Harley RD, Nelson LB, Flanagan JC, Calhoun JH. Ocular motility disturbances following cosmetic blepharoplasty, *Arch Ophthalmol*, 104:542-544, 1986.
5. Syniuta LA, Goldberg RA, Thacker NM, Rosenbaum AL. Acquired strabismus following cosmetic blepharoplasty, *Plast Reconstr Surg*, 111:2053-2059, 2003.

Key Words: Oscillopsia, Blepharoplasty, Anatomy, Surgery, Inferior Oblique Muscle

Financial Disclosure: Dr. Louise Mawn, support from NIH and RPB for unrelated projects. The remaining authors had no disclosures.

Poster 84

Challenges in Recognizing Dural Puncture-induced Intracranial Hypotension as the Cause of Sixth Cranial Nerve Palsy

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Introduction:

Inadvertent dural perforation is not uncommon during epidural pain management. CSF leakage may lead to the development of intracranial hypotension and sixth cranial nerve palsy. Clinicians may overlook the causal connection between the presumed epidural tear and the sixth cranial nerve palsy for a variety of reasons.

Methods:

A retrospective review disclosed 6 cases examined at the University of Michigan between 2009 and 2011 with sixth cranial nerve palsies attributable to intracranial hypotension (IH) following epidural pain management related to surgical procedures (5 cases) or parturition (1 case).

Results:

Diplopia was reported within 1 to 14 days after the placement of the epidural system. Four patients had unilateral and two had bilateral sixth nerve palsies. Complete recovery was seen in 2 weeks in 1 patient, 4 weeks in 1 patient, and much longer in the other 4 patients. The establishment of IH as the cause of the palsy was confounded by one or more features, including lack of awareness on the part of ophthalmologists that an epidural pain control system had been used, lack of headache, or clinical and brain MRI findings that left open the possibility of inflammatory or neoplastic meningitis.

Conclusion:

We conclude that when sixth cranial nerve palsy develops in a patient who has recently undergone a surgical procedure or parturition, clinicians should inquire about the use of an epidural pain management system and maintain a high suspicion for dural puncture-induced IH before embarking on far-flung diagnostic studies, including lumbar puncture.

References:

1. Nishio I, Williams BA, Williams JP. Diplopia: a complication of dural puncture. *Anesthesiology*. 2004 Jan;100(1):158-64.
2. Béchar P, Perron G, Laroche D, Lacroix M, Labourdette A, Dolbec P. Case report: epidural blood patch in the treatment of abducens palsy after dural puncture. *Can J Anaesth*. 2007 Feb;54(2):146-50.
3. Arcand G, Girard F, McCormack M, Chouinard P, Boudreault D, Williams S. Bilateral sixth cranial nerve palsy after unintentional dural puncture. *Can J Anaesth*. 2004 Oct;51(8):821-3.
4. Szokol JW, Falleroni MJ: Lack of efficacy of an epidural blood patch in treating abducens nerve palsy after an unintentional dural puncture. *Reg Anesth Pain Med* 1999; 24:470-2
5. Thorsen G. Neurological complications after spinal anesthesia. *Acta Chir Scand Suppl* 1947;121:1-272.
6. Corbonnois G, O'Neill T, Brabis-Henner A, Schmitt E, Hubert I, Bouaziz H. Unrecognized dural puncture during epidural analgesia in obstetrics later confirmed by brain imaging. *Ann Fr Anesth Reanim*. 2010 Jul-Aug;29(7-8):584-8.
7. Yatziv Y, Stolowitch C, Segev Y, Kesler A. Acute esotropia after epidural anesthesia. *Obstet Gynecol*. 2008 Feb;111(2 Pt 2):540-1.
8. Niedermüller U, Trinkka E, Bauer G. Abducens palsy after lumbar puncture. *Clin Neurol Neurosurg*. 2002 Jan;104(1):61-3.

Key Words: Sixth nerve palsy, Intracranial hypotension, Epidural anesthesia, Dural perforation, Pachymeningitis

Financial Disclosure: The authors had no disclosures.

Poster 85

Hemifacial Spasm Following Gamma Knife Radiosurgery For Vestibular Schwannoma

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Introduction:

Hemifacial spasm is an uncommon complication following stereotactic radiosurgery for vestibular schwannoma.

Methods:

We report two patients who developed hemifacial spasm after gamma knife radiosurgery (GKRS) for vestibular schwannoma.

Results:

Case 1 is a healthy 58 year old man who presented with unilateral hearing loss and tinnitus. Cranial imaging showed vestibular schwannoma. He underwent GKRS and received a total dose of 12.1 Gy. Seventeen months after treatment, he developed episodic hemifacial twitching associated with very mild hemifacial weakness ipsilateral to the side of the tumor. Repeat cranial imaging showed a radiologically stable tumor. Case 2 is a 64 year old woman who presented with balance difficulties and right-sided hearing loss. A right internal acoustic canal tumor was found on cranial imaging. She underwent GKRS. The radiation dose delivered 13 Gy. She developed right hemifacial spasms several days after GKRS which subsequently improved. However, 8 months after GKRS, right hemifacial spasms recurred and were worse. Follow-up scans showed a radiologically stable tumor. She was treated with Botulinum toxin A injections.

Conclusion:

Hemifacial spasm is a delayed complication of stereotactic radiosurgery for vestibular schwannoma. It is amenable to Botulinum toxin A injections but tumor regrowth should first be ruled out.

References:

1. Battista, RA. Gamma knife radiosurgery for vestibular schwannoma, *Otolaryngol Clin N Am* 42, 635-654, 2009
2. Hayhurst, C et al, Predicting non-auditory adverse radiation effects following radiosurgery for vestibular schwannoma: A volume and dosimetric analysis. *Int J Radiation Oncology Biol Phys*, 1-6, 2011.
3. Lee, DJ, WH Westra, H Staecker, D Long and J Niparko. Clinical and histopathologic features of recurrent vestibular schwannoma (acoustic neuroma) after stereotactic radiosurgery. *Otology & Neurotology*, 24: 650-660, 2003.
4. Pollock, BE. Management of vestibular schwannomas that enlarge after stereotactic radiosurgery: Treatment recommendations based on a 15 year experience. *Neurosurgery*, 58, 2, 241-248, 2006.
5. Slattery, WH III. Microsurgery after radiosurgery or radiotherapy for vestibular schwannomas. *Otolaryngol Clin N Am* 42, 707-715, 2009

Key Words: hemifacial spasm, stereotactic surgery, vestibular schwannoma, Botox injection, gamma knife radiosurgery

Financial Disclosure: The authors had no disclosures.

Poster 87

The effect of horizontal rectus muscle surgery on distance-near incomitance

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Introduction:

To determine the effect of horizontal rectus muscle surgery on distance-near incomitance.

Methods:

Prospective evaluation of patients who had horizontal rectus muscle surgery between 12/09 and 9/11. Prism and alternate cover testing was performed at distance (6 meters) and near (0.3 meters) before and after > 45 minutes of monocular occlusion at the pre-operative and post-operative examinations. The change in distance-near incomitance was calculated. The choice of strabismus surgery was at the surgeon's discretion. Patients with muscle fibrosis/paralysis were excluded.

Results:

Twenty-three patients met inclusion criteria. Seven patients had esotropia treated with medial rectus recessions. Sixteen patients had exotropia treated with either a recess/resect procedure (5 patients), lateral rectus recessions (9 patients), or medial rectus resection (2 patients). Within one week after surgery, the change in distance-near incomitance was clinically insignificant in all patients, (< 5 prism diopters in 19 patients, < 8 prism diopters in all 23 patients). Sixteen of 17 patients that had measurements > 2 months after surgery had reduced distance-near incomitance prior to monocular occlusion. In seven patients, distance-near incomitance increased to the preoperative amount after fusional vergence was eliminated by monocular occlusion. Nine patients maintained their reduction in distance-near incomitance after occlusion, indicating that muscle/anatomic changes reduced distance-near incomitance.

Conclusion:

Horizontal rectus muscle surgery does not have a clinically significant effect on distance-near incomitance. Therefore, it is not necessary to consider distance-near incomitance when choosing between medial rectus and lateral rectus muscle surgery. Two months after surgery, distance-near incomitance often decreases from fusional vergence, muscle/anatomic changes, or both.

References: None

Key Words: strabismus surgery, strabismus, distance-near incomitance, horizontal rectus surgery

Financial Disclosure: The authors had no disclosures.

Poster 88

Alternating Windmill Nystagmus, 28 Years & Still the Only Case Seen

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Introduction

This is a case seen in 1973 of an elderly woman who lost sight due to an acquired eye disease and developed alternating windmill nystagmus.

Methods:

This is the only case report of an alternating windmill nystagmus.

Results:

Review of this case showed that alternating windmill nystagmus could occur due to acquired blindness without any other neurologic cause seen at that time.

Conclusion:

Pathologic alternating nystagmus can occur in eye diseases that disrupt the input towards the brain and its capability to keep eyes on the target, and change in direction may be caused by a change in the null point due to the tonic activity of the vestibular nuclei.

References:

1. Sanders MD, Glaser JS, Alternating windmill nystagmus, Neuro-ophthalmology, Vol 7, St Louis: CV Mosby, 1973:133-136
2. Hall E, Gordon J, Abel LA, Hainline L, Abramov I; Nystagmus Waveforms In Blindness, Visual Impairment Research 2000: vol 2, No. 2 pp. 65-73

Key Words: Alternating nystagmus, windmill, blindness, periodic alternating nystagmus, vestibular nucleus

Financial Disclosure: The authors had no disclosures.

Poster 89

Safety of Prednisone for Ocular Myasthenia Gravis

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Introduction:

Treatment with chronic corticosteroids has been associated with frequent significant adverse effects. We hypothesized that the major side effect profile of a long-term, low-dose prednisone regimen for OMG was low. We attempted to project the number of complications that would be observed in randomized controlled trial of OMG using such a regimen.

Methods:

Consecutive OMG patients, evaluated and managed at a single institution between 10/1984 and 12/2010 and treated with ≥ 1 month of daily prednisone, were included. Complications of steroids were defined as the development of or worsening of any conditions that required an alteration to medical therapy. Serious complications included conditions requiring hospitalization, surgery, or emergency care. Statistical methods included univariate analyses and survival analysis.

Results:

83 patients were included (median age: 61 years, 72% men). Follow-up ranged from 1-271 months (median: 58). Fifty-eight (70%) patients had follow-up ≥ 24 months. The maximum prednisone dose ranged from 10-60 mg. Tapering to ≤ 10 mg per day required ≤ 4 months for all but two patients. The median average daily dose following the initial course was 5 mg daily (interquartile range: 4-7.5). During the first two years, there were 24.5 complications per 100 person-years. Only one patient had a serious complication within the first two years (2 year cumulative risk: 1%), but he was not following the recommended regimen, instead taking 20-40 mg daily.

Conclusion:

Low-dose prednisone for OMG appears to be safe and causes few serious complications (~1% two year risk). However, patients need monitoring for the early detection of the relatively common, but less serious complications (~39% two year risk) to ensure the timely adjustment of medical therapy. In particular, a low-dose prednisone regimen is safe within the context of a clinical trial for the treatment of OMG where patients will be on standardized protocol with defined study evaluations.

References:

1. Pascuzzi RM, Coslett HB, Johns TR. Long-term corticosteroid treatment of myasthenia gravis: report of 116 patients. *Ann Neurol* 1984;15:291-298.
2. Caldwell JR, Furst DE. The efficacy and safety of low-dose corticosteroids for rheumatoid arthritis. *Semin Arthritis Rheum* 1991;21:1-11.
3. Kupersmith MJ, Ying G. Ocular motor dysfunction and ptosis in ocular myasthenia gravis: effects of treatment. *Br J Ophthalmol* 2005;89:1330-1334.

Key Words: ocular myasthenia gravis, prednisone, clinical trials, safety

Financial Disclosure: The authors have no disclosures.

Poster 90

Non-mydratric Ocular Fundus Photography Read by Emergency Department (ED) Physicians: FOTO-ED Study

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Introduction:

In the first phase of the FOTO-ED Study, 12.6% of 350 ED patients with complaints or conditions warranting ocular fundus examination had findings, such as papilledema, that should have altered their management or disposition. Disturbingly, only 14% of these 350 patients had direct ophthalmoscopy performed, and nearly all of the relevant findings were missed and identified solely by fundus photography reviewed by neuro-ophthalmologists. Our aim was to compare non-mydratric ocular fundus photography read by ED physicians to their direct ophthalmoscopy.

Methods:

Patients presenting to our ED with headache, focal neurologic defect, visual change, or diastolic blood pressure ≥ 120 were prospectively enrolled. Fundus photography was performed by a nurse practitioner or medical student using a non-mydratric fundus camera (Kowa nonmyd-alpha-D). ED physicians were notified that photographs were available on the electronic medical record, but were not required to review the photographs.

Results:

354 patients were included from among 473 assessed for eligibility (exclusions: 85 ineligible, 33 refused participation). Photographs of 239 (68%) were reviewed by ED physicians. 35 patients (10%) had relevant findings identified by neuro-ophthalmologist review (6 disc edema, 6 grade III/IV hypertensive retinopathy, 7 isolated hemorrhages, 15 optic disc pallor, and 1 retinal vascular occlusion). 16 of those 35 relevant findings (46%) were identified by the ED physicians, eight (24%) occurred in patients whose photographs were not reviewed by ED physicians, and the remaining were reviewed but recorded as "likely normal" or "normal". The ED physicians reported that the photographs were helpful for 125 patients (35%).

Conclusion:

Non-mydratric fundus photographs were used more frequently than direct ophthalmoscopy by ED physicians, and were more sensitive to relevant abnormalities. Ocular fundus photography was often helpful in ED patient evaluations, even when normal. We next plan to investigate whether training in the interpretation of fundus photographs improves ED physician performance and facilitates appropriate ED management.

References:

1. Bruce BB, Lamirel C, Wright DW, Ward A, Heilpern KL, et al. Nonmydratric ocular fundus photography in the emergency department. *N Engl J Med*. 2011 Jan 27;364:387-9.
2. Bruce BB, Lamirel C, Biousse V, Ward A, Heilpern KL, et al. Feasibility of Nonmydratric Ocular Fundus Photography in the Emergency Department: Phase I of the FOTO-ED Study. *Acad Emerg Med*. 2011 Sep;18:928-33.
3. Lamirel C, Bruce BB, Wright DW, Delaney KP, Newman NJ, et. al. Quality of non-mydratric digital fundus photography obtained by nurse practitioners in the emergency department: the FOTO-ED study. *Ophthalmology*. In press.

Key Words: ocular fundus, non-mydratric fundus photography, emergency medicine, papilledema

Financial Disclosure: the authors have no disclosures.

Poster 91

Ophthalmic evaluations in clinical studies of fingolimod (FTY720) in multiple sclerosis (MS)

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Introduction:

Fingolimod is a sphingosine 1-phosphate receptor modulator used to treat relapsing MS. The results of ophthalmic evaluations during clinical studies of fingolimod in patients with MS are reported.

Methods:

Analyses were based on data from MS patients enrolled in FREEDOMS, a 2-year, placebo controlled study (n=1272), and on pooled safety data from FREEDOMS, TRANSFORMS (1-year interferon beta-1a-controlled), plus extension data up to 24 months, and phase 2 data up to 60 months (n=2615, "All Studies Group"). Patients with diabetes mellitus, a risk factor for macular edema (ME), were excluded.

Results:

In the All Studies Group, 16 cases of ME were confirmed by the retinal expert on the Data and Safety Monitoring Board (fingolimod 0.5 mg, n=2, 0.2%; 1.25 mg, n=14, 1.1%). One case (fingolimod 0.5mg) had an alternative possible cause (retinal vein branch occlusion associated with long-standing hypertension). Most patients with ME (n=12, 75%) presented with blurred vision or decreased visual acuity; most (n=12, 75%) were diagnosed within 3–4 months of treatment initiation. ME usually improved or resolved after drug discontinuation; there were no reports of further deterioration in vision after stopping study drug. A history of uveitis in four (25%) cases of confirmed ME (vs 1% in All Studies Group), suggests that uveitis may be a risk factor. Optical coherence tomography provides a sensitive measure of central foveal thickness (CFT) and ME is usually associated with a CFT ≥ 300 μm . In FREEDOMS, slight increases in CFT from a baseline mean of 169 μm were observed in the fingolimod groups (0.5 mg, +4.1 μm ; 1.25mg, +5.5 μm ; placebo, +0.9 μm). Visual acuity was similar between treatment groups and stable throughout the study.

Conclusion:

Fingolimod 0.5mg is associated with a low incidence of clinically significant ME. A slight increase in mean CFT over 24 months is not considered to be clinically relevant.

References: None

Key Words: multiple sclerosis, fingolimod, ophthalmic evaluation

Financial Disclosure: Dr. Zarbin has served as a consultant for Novartis, Iridex, Pfizer/Wyeth, Celgene, Allergan, Genentech, Eli Lilly, and Alimera Sciences and has received grant support from Advanced Cell Technology, Foundation Fighting Blindness, Research to Prevent Blindness, Lincy Foundation, and the National Eye Institute. Dr. Collins is an employee of Novartis AG. Drs. Francis and Zhang are employees of Novartis Pharmaceuticals Corporation.

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Poster 92

Sub-Retinal Fluid Is Common in Experimental Anterior Ischemic Optic Neuropathy

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Introduction:

Anterior ischemic optic neuropathy (AION) is due to ischemia of the optic nerve head, leading to significant vision loss. Sub-retinal fluid has been reported in about 10% of patients.¹

Methods:

We used a murine photochemical thrombosis model of AION.²⁻⁴ We performed vivo quantification with spectral domain optical coherence tomography (SD-OCT) in 8 adult C57BL/6 mice one day following AION using the ring scan vs. volume scan mode to assess the frequency of sub-retinal fluid following acute AION. The ring scan consisted of diameter of 12 deg, and the volume scan included 25 horizontal B-scans spanning a 15°x15° area centered on the disc.

Results:

Within one day after experimental AION, there was optic nerve head edema on histology and staining of the disc on fluorescein angiography. As measured by the ring scan mode on SD-OCT, there was significant swelling of the peripapillary retina following AION, with 30% increased in the inner retinal thickness (retinal nerve fiber layer + retinal ganglion cell layer + inner plexiform layer) (AION eyes: $100.1 \pm 2.2 \mu\text{m}$, control eyes: $77.2 \pm 0.4 \mu\text{m}$, $P < 0.0001$). On day one, ring scan of the peripapillary retina showed no evidence of sub-retinal fluid on SD-OCT. In contrast, volume scan of the optic disc revealed sub-retinal fluid in 100% of AION eyes. No sub-retinal fluid was seen in the contralateral control eyes with either scan mode.

Conclusion:

While the ring scan mode of SD-OCT allowed measurement of the retinal nerve fiber layer, it underestimated the presence of sub-retinal fluid in acute AION. Volume scan significantly increased the sensitivity of detecting sub-retinal fluid. Based on the animal data, we now routinely perform retinal nerve fiber layer ring scan, volume scan, and macular scan to assess patients with acute AION and have found increased incidences of sub-retinal fluid.

References:

1. Hedges TR, Vuong LN, Gonzalez-Garia AO, Mendoza-Santiesteban CE, Amaro-Quierza ML. Subretinal Fluid From Anterior Ischemic Optic Neuropathy Demonstrated by Optical Coherence Tomography. *Arch Ophthalmol.* 126(6):812-815, 2008.
2. Bernstein SL, Guo Y, Kelman SE, Flower RW, Johnson MA. Functional and cellular responses in a novel rodent model of anterior ischemic optic neuropathy. *Invest Ophthalmol Vis Sci.* 44:4153-4162, 2003.
3. Goldenberg-Cohen N, Guo Y, Margolis F, et al. Oligodendrocyte dysfunction after induction of experimental anterior optic nerve ischemia. *Invest Ophthalmol Vis Sci.* 46:2716-2725, 2005.
4. Pangratz-Fuehrer S, Kaur K, Ousman SS, Steinman L, Liao YJ. Functional rescue of experimental ischemic optic neuropathy with alphaB-crystallin. *Eye (Lond).* 25:809-817, 2011.

Key Words: anterior ischemic optic neuropathy, optical coherence tomography, subretinal fluid, animal model

Financial Disclosure: The authors had no disclosures.

Poster 93

Quantification of retinal neural loss in patients with neuromyelitis optica and multiple sclerosis with or without history of optic neuritis using Fourier-domain optical coherence tomography

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Introduction:

Optical coherence tomography (OCT) may be used to detect a large array of retinal abnormalities, including those resulting from axonal loss in diseases affecting the anterior optic pathway, such as multiple sclerosis (MS) and neuromyelitis optica (NMO). The purpose of this study was to compare retinal nerve fiber layer (RNFL) and macular thickness measurements in patients with MS and NMO with or without history of optic neuritis and in controls using Fourier-domain (FD) optical coherence tomography (OCT).

Methods:

Patients with MS (n=60), NMO (n=33), longitudinal extensive transverse myelitis (LETM) (n=28) and healthy controls (n=41) were submitted to ophthalmic examination, including automated perimetry, and to FD-OCT RNFL and macular thickness measurements. Five groups of eyes were compared: MS with (Group 1) or without (Group 2) previous optic neuritis, NMO (Group 3), LETM (Group 4) and controls (Group 5). Correlation between OCT and VF findings was investigated.

Results:

With regard to most parameters, RNFL and macular thickness measurements were significantly smaller in Groups 1 to 4 than in Group 5. Groups 1 and 2 did not differ significantly, but measurements were smaller in Group 3 than in all other groups. RNFL (but not macular thickness) measurements were significantly smaller in Group 4 than in controls. While OCT abnormalities were significantly correlated with VF loss in both NMO/LETM and MS, the correlation was much stronger in the former.

Conclusion:

Although FD-OCT RNFL and macular thickness measurements can reveal subclinical or optic neuritis-related abnormalities in both NMO-spectrum and MS patients, abnormalities are predominant in the macula of MS patients and in RNFL measurements in NMO patients. The correlation between OCT and VF abnormalities was stronger in NMO than in MS, suggesting the two conditions differ with regard to structural and functional damage.

References:

1. Fisher JB, Jacobs DA, Markowitz CE, et al. Relation of visual function to retinal nerve fiber layer thickness in multiple sclerosis. *Ophthalmology* 2006;113(2):324-32.
2. Ratchford JN, Quigg ME, Conger A, et al. Optical coherence tomography helps differentiate neuromyelitis optica and MS optic neuropathies. *Neurology* 2009;73(4):302-8.
3. Naismith RT, Tutlam NT, Xu J, et al. Optical coherence tomography differs in neuromyelitis optica compared with multiple sclerosis. *Neurology* 2009;72(12):1077-82.
4. Henderson AP, Trip SA, Schlottmann PG, et al. An investigation of the retinal nerve fibre layer in progressive multiple sclerosis using optical coherence tomography. *Brain* 2008;131(Pt 1):277-87.
5. Cheng H, Laron M, Schiffman JS, et al. The relationship between visual field and retinal nerve fiber layer measurements in patients with multiple sclerosis. *Invest Ophthalmol Vis Sci* 2007;48(12):5798-805.
6. Saidha S, Syc SB, Ibrahim MA, et al. Primary retinal pathology in multiple sclerosis as detected by optical coherence tomography. *Brain* 2011;134(Pt 2):518-33.
7. Costello F, Hodge W, Pan YI, et al. Tracking retinal nerve fiber layer loss after optic neuritis: a prospective study using optical coherence tomography. *Mult Scler* 2008;14(7):893-905.
8. de Seze J, Blanc F, Jeanjean L, et al. Optical coherence tomography in neuromyelitis optica. *Arch Neurol* 2008;65(7):920-3.
9. Merle H, Olindo S, Donnio A, et al. Retinal peripapillary nerve fiber layer thickness in neuromyelitis optica. *Invest Ophthalmol Vis Sci* 2008;49(10):4412-7.
10. Moura FC, Fernandes DB, Apóstolos-Pereira SL, et al. Optical coherence tomography evaluation of retinal nerve fiber layer in longitudinally extensive transverse myelitis. *Arq Neuropsiquiatr* 2011;69(1):69-73.

Key Words: Neuromyelitis Optica, Multiple Sclerosis, Axonal loss, Optical coherence tomography, retinal nerve fiber layer

Financial Disclosure: The authors had no disclosures.

Poster 94

Clinical and immunological heterogeneity of optic-spinal multiple sclerosis in China

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Introduction:

Whether optic-spinal multiple sclerosis(OSMS) is belonged to MS or neuromyelitis optica(NMO) is still controversial.

Methods:

The medical records of consecutive OSMS patients hospitalized in Department of Neurology, Beijing Tongren Hospital, Capital Medical University, during May, 2009 to April 2011, were reviewed. Clinical features, brain and spinal MRI, CSF examinations, blood testes including systemic autoimmune diseases related antibodies and anti-AQP4 antibody(AQP4-Ab) positivity were recorded. Patients who met the revised NMO diagnostic criteria were further classified as "relapsing NMO", then statistically compared with the other OSMS patients.

Results:

Totally 62 OSMS patients with intact clinical data were included, 8 male and 54 female. Age ranged 9 to 61 year, averaged 36.3 ± 13.3 years. EDSS was 2.8 ± 1.4 . Severe optic neuritis with poor outcome was found in 59 of 116(50.9%) affected eyes. Transverse myelitis was found on 25 cases(40.3%). In totally 100 brain MRI performed, white matter lesion was found in 49(49.0%). Barkhof brain lesions was found in 2 cases(3.2%) while the other 2 cases(3.2%) showed "NMO-like" brain lesions. In 38 spinal cord MRI performed during myelitis episode, abnormalities were found on 37(97.3%), of which 15(40.5%) was longitudinal extensive spinal cord lesion(LESCL). OB and MBP were positive in 3(8.8%) and 6(17.6%) of 34 patients with CSF examination. Serum AQP4-Ab was tested for 44 cases, in which 25 cases(56.8%) was positive. Thirty-four(54.8%) patients met the revised NMO-2006 diagnostic criteria(relapsing-NMO). Compared with the other 28 cases, "relapsing-NMO" had significantly higher EDSS(3.3 ± 1.5 vs 2.2 ± 1.1), more transverse myelitis(52.9% vs 25.0%), more LESCL (50% vs 0%) and higher positivity of AQP4-Ab (82.1% vs 6.7%). The 2 patients with NMO-like brain lesion were in "relapsing NMO" group.

Conclusion:

Clinical and immunological heterogeneities exist in OSMS in China.

References:

1. Weinshenker BG, Wingerchunk DM. Japanese optica-spinal MS-is is MS or neuromyelitis optica and does the answer dictate the treatment? *Neurology*, 2010, 75: 1404-1405.
2. Kira J. Multiple sclerosis in Japanese population. *Lancet Neurol*. 2003, 2:117-27.

Key Words: optica-spinal multiple sclerosis, neuromyelitis optica, AQP4 antibody, MRI, China

Financial Disclosure: The authors had no disclosures.

Poster 95

Gender and retinal nerve fiber layer thickness in patients with non-acute optic neuritis

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Introduction:

Multiple sclerosis (MS) is a demyelinating disease of the central nervous system. Females are three times as likely to develop MS compared to men. Optic neuritis (ON) is common in MS patients, and the effects of inflammation in the anterior visual pathway can be readily quantified with optical coherence tomography (OCT). A recent meta-analysis has shown that time domain OCT (TD-OCT)- measured retinal nerve fiber layer (RNFL) values are approximately 20 microns lower in MS ON eyes ($p < 0.0001$), and approximately 7 microns ($p < 0.0001$) lower in MS non ON eyes. The impact of gender on RNFL values has not been established. Therefore the purpose of this study was to longitudinally compare the male and female TD-OCT-measured RNFL thickness in a cohort of MS patients, excluding patient with acute ON (within 6-months).

Methods:

As part of an ongoing longitudinal study at the University of Calgary, patients underwent repeat OCT (Stratus III, Zeiss Meditech, Dublin, CA), best-corrected VA (EDTRS charts) and low contrast letter acuity (2.5 and 1.25% Sloan charts) testing.

Results:

Data were analysed from 68 men (59% ON patients) and 164 women (54% ON patients). RNFL thinning was significantly greater for male ON eyes (5.7 microns, SD=9.0) compared to female ON eyes (1.5 microns, SD=7.0, $p < 0.05$), ($p < 0.05$) over a period of 1 year. No differences were found between the sexes in the fellow eyes of ON patients or eyes of non ON patients.

Conclusion:

Gender appears to impact RNFL thinning in ON affected eyes, with evidence of better RNFL preservation in women. Sex hormones including estrogen are known to have neuroprotective benefits, but whether this can explain gender differences in RNFL thinning after ON awaits further study.

References: None

Key Words: Multiple Sclerosis, Optic Neuritis, Retinal nerve fiber layer thickness, Gender

Financial Disclosure: Dr. Fiona Costello has received advisory board compensation from Novartis and Biogen Idec. The remaining author has nothing to disclose.

Poster 96

Hyperbaric oxygen treatment for ischemia of the visual pathways even beyond the acute phase

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Introduction:

Hyperbaric oxygen treatment (HBOT) involves the administration of oxygen at supra-atmospheric pressures at high concentrations. It has been suggested for the treatment of acute brain ischemia, various retinal pathological conditions and optic neuropathies, including radiation-induced. Purpose: To describe 2 cases, one with brain ischemia and one with radiation induced optic neuropathy (radAION), that improved after HBOT beyond the acute ischemic phase.

Methods:

Report of 2 cases treated by HBOT and review of the literature.

Results:

Case 1: A 3 year-old girl, severe preterm 29 weeks/934gr, was diagnosed and operated urgently for brain herniation caused by VP shunt malfunction. After surgery she had wandering eyes with no response to light; pupils reacted to light and normal fundoscopy except for mild disc palor. MRI showed bilateral occipital stroke. The child was sent for HBOT 1 week after the operation. Following the 10th treatment visual acuity improved, but she remained with complete left hemianopsia.

Case 2: A 63-year-old woman suffered from severe optic and retinal ischemia 4 years after being treated with brachytherapy for choroidal melanoma. HBOT improved visual function, especially the visual field.

Conclusion:

Recent studies in human and animal models of brain ischemia and infarction [1-5] reported improved visual outcome using HBOT via a protective anti-apoptotic effect [1,2], related to the immediacy of its administration after an acute ischemic event and the extent of necrosis occurring immediately after injury. In animal models, HBO treatment administered immediately after injury induction protects neuronal retinal cells from apoptosis, as shown both histologically and molecularly, without toxic side effects. The cases presented demonstrate that HBOT administration even after the acute phase of ischemic damage to the visual pathways, may lead to improvement in visual functions.

References:

1. Avraham-Lubin BR., Dratviman-Storobinsky O., Dadon-Bar El S., Hasanreisoglu M., Goldenberg-Cohen N. Neuroprotective effect of hyperbaric oxygen therapy on anterior ischemic optic neuropathy. *Front Neurol.* 13;2:23, 2011.
2. Gaydar V., Ezrachi D., Dratviman-Storobinsky O., et al. Reduction of apoptosis in ischemic retinas of two mouse models using hyperbaric oxygen treatment. *Invest Ophthalmic Vis Sci*, in press, 2011
3. Gibson AJ, Davis FM. Hyperbaric oxygen therapy in the treatment of post cardiac surgical strokes--a case series and review of the literature. *Anaesth Intensive Care.* 38:175-184, 2010.
4. Matchett GA, Martin RD, Zhang JH. Hyperbaric oxygen therapy and cerebral ischemia: neuroprotective mechanisms. *Neurol Res.* 31:114-121,2009.
5. Yin D, Zhou C, Kusaka I, et al. Inhibition of apoptosis by hyperbaric oxygen in a rat focal cerebral ischemic model. *J Cereb Blood Flow Metab.* 23:855-864, 2003.

Key Words: hyperbaric oxygen treatment, acute brain ischemia, radiation optic neuropathy, visual outcome, case report

Financial Disclosure: The authors had no disclosures.

Poster 97

A Retrospective Chart Review Study Comparing Elevated and Normal C-Reactive Protein (C-RP) Groups in NAION Patients: Demographics, Visual Functions on Admission and Follow-up.

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Introduction:

Elevated C-RP/ ESR in a patient with swollen disc and visual loss is suggestive of temporal arteritis (arteritic AION). Still there are patients who present as NAION with elevated C-RP with no clinical signs of temporal arteritis, and uneventful follow-up.

Methods:

We carried out a retrospective chart review study of all NAION patients admitted to our department between 1.2005 and 1.2010. Inclusion criteria: unilateral visual loss, swollen disc, C-RP level taken at time of admission, and follow-up compatible with NAION. Exclusion criteria: Patients with temporal arteritis, known or discovered rheumatologic or inflammatory disease, known malignancy or systemic infection.

Results:

40 out of 51 NAION patients were included in the study: eleven (27.5%) had elevated CRP (>5) mean 12.798+/- 6.35. There was a significant difference in visual fields (VFs) mean deviation (MD) at presentation (but not visual acuity), in the elevated C-RP group 100% had MD less than -10 compared to 66.67% of the normal C-RP group (P=0.0356). Diabetes mellitus was more prevalent in the normal C-RP group compared to elevated C-RP (P=0.0028). Elevated CRP was more prevalent in the Israeli Arab patients (43.47%) compared to the Israeli Jewish patients(5.88%) (p=0.0111). Mean age at presentation was significantly lower in Israeli Arab compared to Israeli Jewish patients (57.53+/-12.03yrs and 70.18+/-11.27yrs respectively) (p=0.0027).

Conclusion:

Our pilot study demonstrates that elevated C-RP is not rare in NAION patients (27%). Elevated CRP may be an independent risk factor for the development of NAION in general or for its appearance at a younger age, this effect might be similar to its role in ischemic heart disease. Patients with elevated C-RP/ESR should however be considered to have arteritic AION until proven otherwise. A prospective study with a larger number of patients is required in order to give more reliable results.

References: None

Key Words: NAION, Elevated C-RP, Risk Factor, retrospective

Financial Disclosure: The authors had no disclosures.

Poster 98

Optic neuropathy after discontinuing biotin supplementation

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Introduction:

Biotin is an essential vitamin which is important for synthesis of fatty acids, catabolism of certain amino acids and gluconeogenesis.¹ It is particularly important for mitochondrial function.² Biotinidase deficiency is an autosomal recessive condition in which the enzyme responsible for normal recycling of biotin is deficient resulting in a loss of biotin. Patients with profound biotinidase deficiency will develop several neurologic problems early in life if untreated. This includes seizures, hypotonia, feeding problems, ataxia, cognitive deficits, sensorineural hearing loss and optic atrophy.³ In the United States, and many countries, biotinidase deficiency is included in newborn screening programs, which has dramatically decreased the number of symptomatic cases.

Methods:

A case is presented of a patient who had been identified with biotin deficiency on newborn screening and provided with appropriate supplementation of biotin. However, at approximately age 10 supplements were discontinued. At age 19 he presented for evaluation of subacute onset of bilateral vision loss.

Results:

The patient was found to have visual acuities of 20/70 and 20/25 with bilateral cecocentral scotomas. Critical flicker fusion was reduced. There was optic pallor bilaterally and optic atrophy measured with OCT. Biotin levels were undetectable and biotinidase activity was reduced. He was placed on biotin 20 mg daily. Within one month the patient appreciated some improvement of his vision. After 4 months of supplementation he had significant improvement and near normalization of visual function testing.

Conclusion:

Biotinidase deficiency is well known to cause optic atrophy. This case is unique as the patient was appropriately supplemented during infancy and childhood but stopped supplementation in adolescence. The only neurologic symptom detected was the optic nerve dysfunction. This case may also be significant as there are several parallels with other mitochondrial disorders such as Leber's hereditary optic neuropathy.

References:

1. Zempleni J, Hassan YI, Wijeratne SS, Biotin and biotinidase deficiency, *Expert Rev Endocrinol Metab*, 3, 715-723, 2008
2. Wolf B, The neurology of biotinidase deficiency, *Mol Genet Metab*, 104, 27-34, 2011
3. Atamna H, Newberry J, Erlitzki R, Schultz CS, Ames BN, Biotin deficiency inhibits heme synthesis and impairs mitochondria in human lung fibroblasts, *J Nutr*, 137, 25-30, 2007

Key Words: optic neuropathy, biotin, biotinidase, mitochondria

Financial Disclosure: The authors had no disclosures.

Poster 99

Analysis of Therapeutic Agents and Their Effectiveness in Sarcoid Optic Neuropathy

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Introduction:

Sarcoidosis of the anterior visual pathway is uncommon, involving 1-5% of cases of sarcoidosis. Although it is said to be a steroid responsive disease with good visual outcomes, the basis for this is largely anecdotal. Prior research has shown the beneficial role of steroid therapy in suppressing active disease. For patients who either fail or cannot tolerate corticosteroid therapy; no single alternate regimen is clearly preferred. Previous case reports have shown good response to cyclosporine A or Methotrexate in some patients, but there are no studies in the literature that compare the visual function outcome, doses, and duration of various therapeutic regimens.

Methods:

Retrospective chart review of all patients with sarcoid optic neuropathy seen in neuro-ophthalmic consultation by 2 neuro-ophthalmologists at NJMS in the period between 1989-2011. We identified 82 patients with sarcoid optic neuropathy. We were able to retrieve 33 charts. Criteria for inclusion were: diagnosis of sarcoid optic neuropathy and 6 months follow up after therapy initiation. Eleven patients met all study criteria. Clinical characters, therapeutic regimens, initial and final visual functions on each regimen was recorded along with the time course, dosages, duration, and side effects for each drug used¹⁻³

Results:

Of the 11 patients, one only required a single agent: prednisone. The remaining 10 patients needed more than 1 drug to stabilize. Prednisone was one of the combined regimens in all 10. Three stabilized on prednisone and methotrexate, two on prednisone and cyclosporine, one on prednisone and thalidomide, one on prednisone and mycophenolate, and one on prednisone and hydroxychloroquine. One patient needed more than 2 drugs: prednisone, cyclosporine, thalidomide, and remicade. Other therapeutic combinations were tried in most patients but had to be discontinued either due to progression or intolerance.

Conclusion:

Despite prior statements that sarcoid optic neuropathy is typically extremely steroid sensitive, such is not always the case. This may be related to complexity of individual case diagnosis leading to diagnosis at an advanced stage. Multiple agents are effective as secondary agents but none is clearly the best adjunct agent in sarcoid optic neuropathy.

References:

1. Maust HA, Foroosan R, Sergott RC, Niazi S, Weibel S, Savino PJ. Use of methotrexate in sarcoid-associated optic neuropathy. *Ophthalmology* 2003;110:559-563.
2. Bielory L, Holland C, Gascon P, Frohman L. Uveitis, cutaneous and neurosarcoid: treatment with low-dose cyclosporine A. *Transplant Proc* 1988;20:144-148.
3. Frohman LP, Guirgis M, Turbin RE, Bielory L. Sarcoidosis of the anterior visual pathway: 24 new cases. *J Neuroophthalmol* 2003;23:190-197.

Key Words: sarcoidosis, optic neuropathy, management

Financial Disclosure: The authors had no disclosures.

Poster 100

Transorbital Drug Delivery for Treatment of Optic Neuropathies

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Introduction:

We have developed an image guided (IG) flexible orbital endoscope for neuroprotective treatment of the optic nerve and tested the system in human phantoms and in a live porcine model.

Methods:

Targets were placed in the orbits in human phantoms. The physician studied the CT images and located the target. Each user was randomized to starting image-guided (IG) versus non IG and randomized to the first eye. Accuracy of color identification and time to target were recorded. 28 physicians were trialed on two phantoms (4 eyes) each. We then tested the system using a living porcine model. In an IACUC approved study, the pigs were anesthetized and glass spheres containing both a coloring agent and a contrast agent were placed in different retrobulbar locations. The color of the target sphere was blind to the surgeon. Three pigs were imaged on a 3T MRI machine. The tomograms were loaded into our proprietary IG surgery system and the image and physical space registered. The surgeon located the target and stated its color.

Results:

Several clear trends emerged in the phantom project. Time to target decreased across the four eye trials for all grades of users. Accuracy was better with both guidance and experience. With IG 84.6% of the trials resulted in a correct color identification where without IG 78.6% were correct. All of the errors with IG were to the closest "distractor" ball where the unguided errors "located" all possible colors. Medical students made 1.33 errors per trial (out of a possible 4) whereas residents made 0.78 errors per trial. Attending physicians made 0.67 errors per trial. Fiducial Registration Error (FRE) was approximately 2 mm \pm 0.45 mm. In the pig trials we have 100% correct identification.

Conclusion:

The designed IG endoscopic system provides accurate guidance, short learning curves and safe delivery to the retrobulbar space.

References:

1. Mawn LA, Shen JH, Jordan DR, Joos KM. Development of an orbital endoscope for use with the free electron laser. *Ophthalmic Plastic and Reconstructive Surgery* 20(2):150-7, 2004.
2. Atuegwu NC, Galloway RL. Sensitivity analysis of fiducial placement on transorbital target registration error. *IJCARS*, Vol 2(6), pp 397-404, 2008.
3. Atuegwu NC, Mawn LA, Galloway RL. Transorbital Image Guidance. *IEEE-EMBS* pp 4663-4666, 2007.
4. Ingram MC, Atuegwu NC, Mawn LA, Galloway RL. Transorbital therapy delivery: phantom testing, *Proc. SPIE 7964*, 79642A, 2011.

Key Words: Image guidance, Neuroprotection, Endoscope, Animal trial, Technology

Financial Disclosure: Dr. Louise Mawn NIH and Research to Prevent Blindness. Dr. Galloway NIH. The remaining authors had no disclosures.

Poster 101

Bilateral optic atrophy with novel MTTL 3268 A>G homoplasmy - LHON variant or not?

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Introduction:

Leber's Hereditary Optic Neuropathy is well known as a maternally inherited eye disease with symptoms of bilateral visual loss that affect generally young male, related predominantly with mitochondrial DNA mutations such as ND1 G3460A, ND4 G11778A and ND6 T14484C. With data accumulated and bio-molecular-genetic methods developed, several novel mutations were reported. We present a case of bilateral optic neuropathy with novel 3268 mutation

Methods:

Case report

Results:

This 56 year-old female with bilateral, insidious, painless visual loss was referred to our clinic for evaluation of visual dysfunction. On exam, visual acuity was OD 20/50, OS 20/60, bilateral optic disc pallor with preserved pupillary reflex and abnormal visual field defects. OCT revealed significant retinal nerve fiber thinning with around 50 micrometer OU. We requested LHON mutation study and revealed abnormality in Mitochondrial DNA.

Conclusion:

We found MTTL 3268 A>G homoplasmy mutation in bilateral optic atrophy patient using PCR and sequencing. This the first report of 3268 mitochondrial mutation in a possible LHON spectrum.

References:

1. Abu-Amro KK. Leber's Hereditary Optic Neuropathy: The Mitochondrial Connection Revisited. *Middle East Afr J Ophthalmol.* 2011 Jan-Mar; 18(1): 17–23
2. Finsterer J. Inherited mitochondrial neuropathies. *Journal of the Neurological Sciences* 2011 304: 9–16.
3. Koilkonda RD, Guy J. Leber's Hereditary Optic Neuropathy-Gene Therapy: From Benchtop to Bedside. *Ophthalmol.* 2011; 2011:179412.

Key Words: LHON, Bilateral Optic atrophy, novel mutation

Financial Disclosure: The authors had no disclosures.

Poster 102

Optic Neuropathy Associated With Copper Deficiency After Gastric Bypass Surgery

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Introduction:

The number of gastric bypass surgeries in the United States, has increased to over 100,000 annually and the increased risk of nutritional deficiencies status post gastric bypass is well known. In addition to folic acid, fat soluble vitamins and zinc, copper levels can become deficient leading to various neurological manifestations. Herein we report a patient with progressive optic neuropathy due to copper deficiency occurring after gastric bypass surgery.

Methods:

A case report of a 35-year old woman with a history of gastric bypass surgery noted to have progressive visual decline in both eyes.

Results:

Ophthalmic examination showed temporal pallor of the optic nerve heads bilaterally. Laboratory work-up revealed copper deficiency. There was a vast improvement in vision with mineral replenishment.

Conclusion:

Nutritional deficiencies frequently occur in patients after bariatric procedures due to reduced gastric and enteral absorption. Vitamin B12 and folic acid deficiency are common causes of nutritional optic neuropathy. Copper deficiency in the body occurs in rare instances and usually takes decades to manifest. Based on case reports the prognosis for visual recovery is poor despite replenishing the mineral. Our case is unique in that copper deficiency was noted as early as 3 years after gastric bypass surgery. Moreover rapid mineral replenishment led to a dramatic recovery of vision. Our case underscores the need for maintaining a high suspicion when evaluating patients with suspected optic neuropathy occurring after gastric bypass surgery since prompt diagnosis and treatment may lead to reversal in visual decline.

References:

1. Folope V, Coeffier M, Dechelotte P. Nutritional deficiencies associated with bariatric surgery. *Gastroenterol Clin Biol*;31(4):369-77, 2007.
2. Livingston EH. The incidence of bariatric surgery has plateaued in the U.S. *Am J Surg* 200(3):378-85, 2010.
3. Naismith RT, Shepherd JB, Weihl CC, et al. Acute and bilateral blindness due to optic neuropathy associated with copper deficiency. *Arch Neurol*;66(8):1025-7, 2009.
4. Pineles SL, Wilson CA, Balcer LJ, et al. Combined optic neuropathy and myelopathy secondary to copper deficiency. *Surv Ophthalmol*;55(4):386-92, 2010.

Key Words: Optic Neuropathy, Copper Deficiency, Gastric Bypass Surgery, Nutritional Optic Neuropathy

Financial Disclosure: The authors had no disclosures.

Poster 103

Unexplained Hydrocephalus in a Patient with Neuromyelitis Optica

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Introduction:

The discovery of antibodies to the aquaporin 4 water channel transformed the understanding of Neuromyelitis optica. The effects of these antibodies on brain-water homeostasis are unknown. We present a case of unexplained hydrocephalus in a patient with Neuromyelitis optica, and speculate as to whether aquaporin-4 dysfunction played a role.

Methods:

A 54 year old blind, quadriparetic female with antibody-positive Neuromyelitis optica was admitted to our hospital for seizure activity, which was controlled with a higher dose of levetiracetam. She was doing well until the third day of admission when she became unresponsive.

Head CT revealed new ventriculomegaly. An external ventricular drain was placed, and her intracranial pressure was measured to be 29cmH₂O. Cerebrospinal fluid drawn from the drain was non-diagnostic and a lumbar puncture was not performed. MRI of the brain demonstrated an enhancing lesion in the peri-callosal white matter consistent with a demyelinating lesion. Her intracranial pressure gradually and spontaneously improved. Four days after insertion, her drain was clamped and subsequently removed. During this time, she became more alert and was able to interact appropriately.

Results:

On the 14th day of admission she started a course of plasma exchange followed by her usual dose of cyclophosphamide. She gradually regained her previous baseline. Serial head CTs showed improvement in her ventriculomegaly.

Conclusion:

No clear etiology of our patient's hydrocephalus was ever revealed. Several authors have implicated aquaporin 4 dysfunction in the pathogenesis and compensation of hydrocephalus in mice. This evidence raises the question: do aquaporin 4 auto-antibodies in Neuromyelitis optica cause or predispose patients to hydrocephalus? We consider this case to be a compelling argument for further research regarding the role that aquaporin 4 auto-antibodies play in hydrocephalus.

References:

1. Bloch et al, Accelerated progression of kaolin-induced hydrocephalus in aquaporin-4-deficient mice, *Journal of Cerebral Blood Flow and Metabolism*, 26, 1527-1537, 2006.
2. Brian et al, Aquaporins: relevance to cerebrospinal fluid physiology and therapeutic potential in hydrocephalus, *Cerebrospinal Fluid Research*, 7:15, 2010.
3. Feng et al, Sporadic Obstructive Hydrocephalus in Aqp4 Null Mice, *Journal of Neuroscience Research*, 87, 1150-1155, 2009.
4. Mao et al, Aquaporin 4 changes in rat brain with severe hydrocephalus, *European Journal of Neuroscience*, 23, 2929-2936, 2006.

Key Words: Neuromyelitis Optica, Hydrocephalus, Aquaporin 4

Financial Disclosure: Dr. Carlos Mora was a one time consultant to Teva Neuroscience in September 2011. The remaining author had no disclosures.

Poster 104

Ipilimumab-associated optic neuropathy

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Introduction:

We report a unique case of ipilimumab-associated optic neuropathy in a patient with metastatic melanoma.

Methods:

Case report.

Results:

A 53-year-old man with metastatic melanoma experienced sudden left eye vision loss. He was previously treated with ipilimumab which was recently discontinued because of hypophysitis. Examination revealed no light perception (NLP) vision in the left eye, afferent pupillary defect, and left optic disc swelling. Brain MRI demonstrated pituitary gland swelling without left optic nerve enhancement. MRA head/neck, echocardiogram, and carotid dopplers were normal. He was treated with enoxaparin for a suspected retinal thromboembolic event. Five weeks later he developed blurred vision in his right eye. Examination revealed 20/50 vision in the right eye with visual field constriction and optic disc swelling. Orbital MRI demonstrated bilateral perineural optic nerve enhancement and central right optic nerve enhancement. CSF showed lymphocytosis, elevated protein, and negative cytology. He was treated with high dose steroids and had improvement in right eye vision and disc swelling. Attempts at tapering steroids resulted in worsening vision. Mycophenolate mofetil was initiated with slow steroid taper. Vision stabilized at 20/20 in the right eye with resolution of optic nerve swelling. Left eye vision remained NLP. Extensive work-up for other etiologies of his optic neuropathy was unrevealing.

Conclusion:

Ipilimumab is a monoclonal antibody that potentiates an antitumor T-cell response by blocking cytotoxic T-lymphocyte antigen-4.¹ It is associated with autoimmune-related adverse effects including enterocolitis, hepatitis, and hypophysitis.² Our patient experienced hypophysitis and bilateral optic neuritis related to ipilimumab, presumably on an auto-immune basis. He was treated with steroids and mycophenolate mofetil, which led to resolution of his optic neuropathy. Optic neuritis appears to be a rare immune-related adverse effect of ipilimumab therapy which, to our knowledge, has not been reported previously.

References:

1. Wolchok JD, Weber JS, Hamid O, et al. Ipilimumab efficacy and safety in patients with advanced melanoma: a retrospective analysis of HLA subtype from four trials. *Cancer Immunity*, 10, 9-14, 2010.
2. Weber J. Ipilimumab: controversies in its development, utility and autoimmune adverse events. *Cancer Immunol Immunother*, 58, 823-830, 2009.

Key Words: Optic neuropathy, Ipilimumab, Hypophysitis, Auto-immune optic neuritis, Vision loss

Financial Disclosure: The authors had no disclosures.

Poster 105

Case Report of Muslin-induced Optic Neuropathy and Review of The Literature

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Introduction:

Intracranial aneurysms are commonly treated with neurosurgical clipping or endovascular therapy via either balloon occlusion or packing with detachable coils. Many aneurysms require reinforcement of the vessel wall by coating or wrapping with muslin (cotton gauze). A rare complication of this wall stabilization is a foreign body inflammatory reaction, which can involve the optic chiasm and/or optic nerve and lead to significant decreased visual acuity. There have been 35 total cases reported in the literature of optic neuropathy caused by intracranial muslin.

Methods:

We present a challenging case of optic neuropathy due to intracranial muslin and review the literature of cases reported.

Results:

In this case report, we describe a 45-year-old man who presented with bilateral painless and progressive vision loss 5 months after aneurysm clipping and wrapping for a subarachnoid hemorrhage. After extensive workup, muslin-induced optic neuropathy was determined to be the cause of his findings. His visual symptoms resolved after high dose dexamethasone. Upon each attempt to taper the steroids, his visual symptoms would worsen until higher doses were re-introduced. This dramatic response to steroids along with the pathology from repeat craniotomy support an underlying chronic inflammatory reaction targeted at the muslin wrapping.

There are 35 cases in the literature that report the use of intracranial muslin or “cotton gauze” and subsequent optic neuropathy, with the exception of two cases that report subsequent CN III palsy and one case that reports headaches and seizures. The delay in the onset of visual symptoms ranges from 1 to 54 months after surgical aneurysm repair. The majority of patients reported have been female (28/35) and treatment responses have varied from surgical intervention, to steroids, and cyclophosphamide.

Conclusion:

The case presented illustrates a 45-year-old man with optic neuropathy secondary to a foreign body inflammatory reaction from intracranial muslin introduced during prior aneurysm clipping procedure.

References: None

Key Words: Optic Neuropathy, Muslinoma, Gauzoma, Foreign body inflammatory reaction, Intracranial aneurysm repair

Financial Disclosure: The authors had no disclosures.

Poster 106

Charles Bonnet Syndrome and Periodic Alternating Nystagmus: Moving Visual Hallucinations

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Introduction:

Charles Bonnet syndrome (CBS), complex visual hallucinations with insight, has been reported in patients with profound visual loss. Acquired periodic alternating nystagmus (PAN) is a rare condition of continuous horizontal jerk nystagmus that periodically reverses direction. It is frequently associated with posterior fossa lesions but has also been described in patients with binocular visual sensory deprivation secondary to ocular disease^{1,2}.

Methods:

We present a case series of 2 patients with visual loss, PAN and CBS syndrome who experience moving visual hallucinations. Video recordings demonstrate that their description of the speed, direction and periodicity of changes in direction of the hallucinations mirror the same features of, and are in time with, their nystagmus.

Results:

A 62 year old woman with visual loss secondary to bilateral retinal detachments and an 85 year old man with visual loss secondary to anterior segment and retinal disease experienced visual hallucinations of a rotating wall and a rotating row of houses respectively. As the slow phase of their nystagmus was to the right, they described the wall/houses as moving to the right. The movement of the wall/houses slowed as the amplitude of the nystagmus reduced and it was stationary at the null point, with the man able to 'see' between the houses. The rotation of the wall/houses then changed direction with the slow phase of the nystagmus.

Conclusion:

The 'sensory deprivation' theory of CBS suggests that sub-threshold visual input leads to production of spontaneous images from the visual association cortex, resulting in visual hallucinations³. Acquired PAN occurring after visual loss has been postulated to be secondary to a latent congenital PAN becoming manifest after loss of fixation², or of a pre-existing brainstem/cerebellar pathology causing ocular instability once normal retinal signals are lost⁴. This is the first report of the perception of visual hallucinations reflecting such ocular movement.

References:

1. Cross S, Smith J, Norton E. Periodic Alternating Nystagmus Clearing after Vitrectomy. *J Clin Neuro-ophthalmol*, 2:5-11, 1982
2. Jay W, Williams B, Chicchis A. Periodic Alternating Nystagmus Clearing after Cataract Surgery. *J Clin Neuro-ophthalmol*, 4:149-152, 1985
3. Miller N. Walsh and Hoyt's Clinical Neuro-Ophthalmology, Vol 2. Williams & Wilkins, Baltimore. pp903-905, 1984
4. Vukicevic M, Fitzmaurice K. Butterflies and black lacy patterns: the prevalence and characteristics of Charles Bonnet hallucinations in an Australian population. *Clin Experiment Ophthalmol*, 36:659-665, 2008

Key Words: Charles Bonnet Syndrome, Periodic Alternating Nystagmus, Hallucinations

Financial Disclosure: The authors had no disclosures.

Poster 107

Diagnostic Uncertainty Due to Optic Disc Drusen

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Introduction:

Optic disc drusen (ODD) are a frequent cause of optic disc elevation raising the possibility of optic disc edema. Papillary and peripapillary hemorrhages with or without optic disc edema may occur with ODD further confounding the clinical picture. We present such a patient.

Methods:

Case study

Results:

An 80-year-old otherwise healthy woman complained of headaches but denied other symptoms of giant cell arteritis. Visual acuity was 20/30 bilaterally and color vision and pupillary reactions were normal. Eye movements were full and slit lamp examination was unremarkable. Both optic discs contained drusen and were edematous with peripapillary hemorrhages. Automated visual fields demonstrated peripheral nerve fiber bundle defects in each eye. Fluorescein angiography confirmed the presence of optic disc drusen (ODD) and showed dye leakage in the late phases of the angiogram, more on the right disc than the left.

With good visual acuity and intact color vision, increased intracranial pressure was suspected and an MRI of the brain was performed. This study showed only white matter ischemic changes consistent with the patient's age. Complete blood count, metabolic panel, and Westergren sedimentation rate were normal. Lumbar puncture showed an opening pressure of 17 cm H₂O with normal cerebrospinal fluid analysis. The patient was prescribed gabapentin with relief of her headaches. Examination three months later showed resolution of the optic disc edema and a decrease in the size of the peripapillary hemorrhages.

Conclusion:

Hemorrhage associated with ODD include; 1) small nerve fiber hemorrhages localized to the optic disc, 2) optic nerve head hemorrhages extending into the vitreous, 3) subretinal peripapillary hemorrhages with or without associated choroidal neovascularization.^{1,2,3} Optic disc edema associated with ODD may occur in; 1) nonarteritic anterior ischemic optic neuropathy (NAION),⁴ 2) raised intracranial pressure,⁵ 3) associated with hemorrhagic events as described above. It is in this last setting that our patient's optic edema developed. The presence of ODD in evaluating the optic disc almost always obviates the need for further patient testing. However, in our patient, the combination of bilateral optic disc edema with good visual acuity mimicking papilledema necessitated additional work-up.

References:

1. Romero J, Sowka J, Shechtman D. Hemorrhagic complications of optic disc drusen and available treatment options. *Optometry*. 2008;79:496-500.
2. Auw-Haedrich C, Staubach F, Witschel H. Optic disk drusen. *Surv Ophthalmol*. 2002;47:515-532.
3. Sanders TE, Gay AJ, Newman M. Hemorrhagic complications of drusen of the optic disk. *Am J Ophthalmol*. 1971;71:204-217.
4. Hayreh S.S. Ischemic optic neuropathy, *Prog Retin Eye Res*. 2009;28:34-62.
5. Granger RH, Bonnelame T, Daubenton J, Dreyer M, McCartney P. Optic nerve head drusen and idiopathic intracranial hypertension in a 14-year-old girl. *J Pediatr Ophthalmol Strabismus*. 2009; 46:238-240.

Key Words: Optic disc drusen, Papilledema, NAION, Peripapillary hemorrhages, Choroidal neovascularization

Financial Disclosure: The author had no disclosures.

Poster 108

Diffusion MRI in a Case of Posterior Ischemic Optic Neuropathy

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Introduction:

PION (posterior ischemic optic neuropathy) is retrobulbar ION (ischemic optic neuropathy) involving the optic nerve and or optic chiasm, but the optic disc is not swollen. PION is much less common than AION (anterior ION) and should be differentiated from any retrobulbar optic nerve pathology including compressive, inflammatory, and infiltrative optic neuropathies. PION generally occurs in either the perioperative setting or in the setting of GCA or other vasculitides. Despite the efficacy of diffusion imaging in detecting acute ischemia of the CNS, there are extremely rare reports of the application of DWI in the setting of ION.

Methods:

A 46-year-old man presented with unilateral painless blindness (OS). Because of the funduscopy showed normal fundus and neuroophthalmologic examination showed RAPD (OS), he was diagnosed with PION. The brain MRI showed high signal intensity in the left optic nerve on diffusion weighted imaging (DWI) with decreased apparent diffusion coefficient (ADC) map. His vision did not recover for years.

Results:

Conclusion:

The DWI and ADC on brain MRI may play an important role in demonstrating optic nerve infarction in PION.

References:

1. Kennard C, Leigh RJ. *Neuro-Ophthalmology*. Elsevier Science Health Science div, 2011.
2. Verma A, Jain KK, Mohan S, Phadke RV. Diffusion-weighted MR imaging in posterior ischemic optic neuropathy. *AJNR Am J Neuroradiol* 2007;28:1839-1840.

Key Words: optic neuropathy, Diffusion weighted imaging, posterior ischemic optic neuropathy

Financial Disclosure: The authors had no disclosures.

Poster 109

Acute Zonal Occult Outer Retinopathy Mimicking Retrobulbar Optic Neuritis

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Introduction:

We would like to report acute zonal occult outer retinopathy (AZOOR) misdiagnosed as retrobulbar optic neuritis in pregnant woman.

Methods:

A 27-year-old pregnant woman suffered a sudden visual disturbance in the right eye. The right eye had a visual acuity of 0.02 and a relative afferent pupillary defect. A visual-field examination revealed an inferotemporal field defect in the right eye. Brain MRIs without gadolinium enhancement were normal. On ophthalmoscopic examination, visual evoked potential, flicker electro-oculogram and Hess test were normal.

Results:

Multifocal electroretinography showed decreased responses in the right eye at the corresponding area with the visual-field defect. SD-OCT demonstrated photoreceptor inner-outer segment junctional disruption and loss. The patient was diagnosed as acute zonal occult outer retinopathy.

Conclusion:

In the cases of subtle retinopathies misdiagnosed as an optic neuropathy, therefore, awareness about the possibility of AZOOR complex in unusually presentation of optic neuropathy is essential to ensure accurate diagnosis and further treatment.

References: None

Key Words: AZOOR, Optic neuritis, SD-OCT, Multifocal ERG, Pregnancy

Financial Disclosure: The authors had no disclosures.

Poster 110

Optic neuritis masquerades

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Introduction:

We present a series of cases of varying aetiology masquerading as optic neuritis and to highlight the importance of a meticulous approach and a high index of suspicion in handling such cases.

Methods:

We present a series of four cases that were referred to us as optic neuritis. The patients were subjected to a detailed examination including slit lamp exam and a dilated fundus examination. Relevant investigations were also performed. In each of the cases the initial diagnosis was disproved and a variety of retinal, inflammatory and metabolic pathologies were localized and appropriately managed.

Results:

Various pathologies can masquerade as optic neuritis and a correct and early diagnosis is necessary to ensure optimal further investigations and treatment.

Conclusion:

A meticulous approach and high index of suspicion are necessary while dealing with cases that present with symptoms of optic neuritis, and a thorough workup should be done to rule out all other causes before a diagnosis of idiopathic optic neuritis can be made.

References: None

Key Words: optic neuritis, investigative algorithm, approach, masquerades, diagnosis

Financial Disclosure: The authors had no disclosures.

Poster 111

Cross sectional survey of visual acuity and self-reported visual function in patients with amyotrophic lateral sclerosis

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Introduction:

Binocular visual acuity (VA) is reduced in amyotrophic lateral sclerosis (ALS) (Moss et al. 2011). Lesions in the afferent visual pathway, abnormal ocular motility and/or limited ophthalmic care might account for this observation. Further characterization is necessary to develop vision as a marker of non-motor neuron impairment in ALS. Therefore we sought to determine if monocular VA, binocular summation (the difference between binocular and monocular acuity) and self reported vision related function are reduced in ALS patients compared with control subjects.

Methods:

Patients attending a multidisciplinary ALS clinic (n=15, age 48.4 +/- 3.1 years) and their caregivers, serving as controls (n=8, age 48.5 +/- 3.1 years) participated in this study. Monocular and binocular VA were assessed using high (100%) and low (2.5%, 1.25%) contrast charts (Sloan) under controlled lighting conditions following refraction. Binocular summation was calculated as the difference between binocular and best monocular acuity scores. Subjects completed the National Eye Institute visual functioning questionnaire-25, a reliable and valid assessment of visual function.

Results:

Low contrast VA was reduced by approximately 1 line in ALS patients versus control subjects in both binocular (OR 0.79 (0.62-0.98), p=0.032 logistic regression) and monocular (OR 0.78 (0.59-0.97), p=0.029, logistic regression) conditions. High contrast VA and high and low contrast binocular summation were not different between groups. There was a trend towards lower overall self-reported health related function in ALS patients (p=0.056), but no difference in self-reported vision related function when compared with the control group (p=0.39).

Conclusion:

We confirm that binocular VA is decreased in ALS patients and also find this to be true for monocular testing. Binocular summation is not affected. Self reported vision related function is not decreased in ALS patients in this small sample. Further study is warranted regarding the pathophysiologic basis for these results and their application to disease categorization and outcomes assessment.

References:

1. Moss, McLuskey, Elman, Galetta, Balcer, Hoskins, Talman, Grossman, Liu, Cross-sectional evaluation of clinical neuro-ophthalmic abnormalities in the ALS population, *Journal of the Neurosciences*, doi:10.1016/j.jns.2011.1.10.016 (in press), 2011

Key Words: vision loss, amyotrophic lateral sclerosis, low contrast acuity, vision related quality of life

Financial Disclosure: Dr. Moss has received speaker honoraria from Quark pharmaceuticals. Dr. Rowin has received research grants from Myasthenia Gravis Foundation of America, ALS Association and Muscular Dystrophy Association

Poster 112

Comparison of latency development in pattern and orientation reversal VEPs

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Introduction:

The latency of Pattern-Reversal (PR)-VEP was present at birth and the latency of its first positive peak has been found to develop rapidly, reaching the adult level around 15 weeks of age (Moskowitz & Sokol, 1983; Lee et al, 2011). However, the developmental course of Orientation-Reversal (OR)-VEP, reflecting the specific spatial organization of cortical receptive fields, still remains unknown.

Methods:

OR-VEP was tested in 81 adults at 1- 12 r/s and 145 infants (age 4-79 weeks) at 2-8 r/s. An additional 123 infants (range 4.0-20.3 wks) were analyzed with OR at 4r/s studied previously (Braddick et al, 2005). The peak latency of the initial OR response in transient recordings at 1-4 reversal/second (r/s) were extracted. For comparison, apparent latencies from the gradient of phase against temporal frequency in steady-state recording at 1- 9.6 r/s were also calculated. This measure reflects the full time course of the response beyond the initial peak.

Results:

For both adults and infants, no significant latency difference in the initial positive peak was found for low reversal rates, suggesting that they arose from a similar level of visual processing. The calculated latency was statistically longer than the transient latency. While the transient latency asymptoted to adult value of 102 ms at around age 55 weeks, the calculated latency showed much less variation across the age span.

Conclusion:

Although there was a dominant effect of transmission delay on the initial peak in infancy due to immature physiology, the fully developed visual cortex, like that in the infant, remains slower in the overall time course of responses to orientation change. Upon reaching maturity, the initial peak in the contrast and orientation VEP responses may arise from the same level of cortical processing in V1, but the overall time course reflected in phase-based measurements still shows a much more prolonged response.

References:

1. Braddick O, Birtles D, Wattam-Bell J, Atkinson J. Motion-and orientation-specific cortical responses in infancy. *Vision Res.* 45(25-26):3169-3179, 2005.
2. Lee J, Birtles D, Wattam-Bell J, Atkinson J, Braddick O. Latency of pattern-reversal and orientation-specific VEPs in adults and infants. *Percept.* 39:135:ECVP Abstract, 2010.
3. Lee J, Birtles D, Wattam-Bell J, Atkinson J, Braddick O. Latency measures of pattern-reversal VEP in adults and infants: different information from transient P1 response and steady-state phase. *Invest Ophthalmol Vis Sci.* (under review). 2011.
4. Moskowitz A, Sokol S. Developmental changes in the human visual system as reflected by the latency of the pattern reversal VEP. *Electroencephalogr Clin Neurophysiol.* 56(1):1-15, 1983.

Key Words: Orientation reversal, Visual evoked potentials, Visual development, Infant, Latency

Financial Disclosure: Jin Lee- a Thouron Fellowship. Oliver Braddick- an award from the Leverhulme Foundation & MRC Research Grant G0601007. Deirdre Birtles, John Wattam-Bell, Janette Atkinson- MRC Research Grant G0601007.

Poster 113

Why Visual Function Does Not Correlate With Optic Glioma Size Or Growth

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Introduction:

It has long been known that there exists little correlation between visual function and tumor size with respect to optic gliomas (World Health Organization Grade I juvenile pilocytic astrocytomas).^{1,2} Hence, some optic gliomas can be large congenitally, and others noted to continue to grow to great extents along considerable lengths of the visual axonal pathways, without causing any perceptible loss of visual acuity or function. Others much smaller, or gradually shrinking in size, may cause considerable visual morbidity.

A pathophysiologic mechanism is proposed to elucidate this apparent incongruity.

Methods:

Review of visual and neuroimaging findings in children with optic gliomas is combined with review of processes active during neuronal development and physiologic aspects of neuronal axon transport under pressure.

Results:

The overgrowth of glial cells with supporting tissue elements that normally surround each axon to create gliomas constitutes a lesion intrinsic to the visual pathways. Such intrinsic growths, particularly when developing in utero while apoptosis is still an ongoing process, can develop in uniform fashion without impeding axoplasmic flow and neuronal signalling. Conversely, non-uniform regression or growth of these intrinsic elements post-natally can disrupt established axonal arrangements and lead to loss of function. Analogous phenomena of paradoxical worsening of visual function despite an amelioration of other underlying neurological causes are analyzed and compared.

Conclusion:

A combination of well-understood neurophysiological phenomena of development, including neuronal proliferation and apoptosis, as well as concepts encompassing uniform versus focal neuronal responses to pressure, can fully explain apparently paradoxical visual phenomenon with respect to optic gliomas in children, without the need to invoke new or unknown processes. A limited utility of visual function or visual evoked potential testing to monitor for the progression of such tumors, or serve as a trigger for the initiation of interventional therapies, is thereby explained.

References:

1. Glaser JS, Hoyt WF, Corbett J. Visual morbidity with chiasmal glioma. Long-term studies of visual fields in untreated and irradiated cases. *Arch Ophthalmol* Jan;85(1):3-12, 1971.
2. Fletcher WA, Imes RK, Hoyt WF. Chiasmal gliomas: appearance and long-term changes demonstrated by computerized tomography. *J Neurosurg* 65(2):154-159, 1986.

Key Words: Glioma, Apoptosis, Visual evoked potential, Visual acuity, Growth

Financial Disclosure: The author has no disclosures.

Poster 114

An iPhone/ iPad App for teaching Neuro-Ophthalmology

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Introduction:

Learning Neuro-Ophthalmology is easier if fun. Sixty-nine per cent of US medical students own an iPhone as a mobile device and 72% of US physicians owned a smartphone in 2010. These are increasingly used as reference devices in preferences to books and computers. The iPhone, iPad and Android equivalents are potent learning tools for they are portable touch-based devices with built in accelerometers that can be programmed to simulate a variety of eye movement and pupillary examples of normal physiology and pathology.

Methods:

Using Apple developer XCode programming suite and Objective-C libraries from the Apple developer website, open-source Cocos-2D gaming software and other open-source software such as GIMP an IOS window based application was constructed incorporating touch control and accelerometer inputs on a standard Apple Mac computer. Subroutines were invented to simulate saccades, blinks, neuronal arc animations and Hess charts responsive to touch and gestures.

Results:

An iPhone App with modules including:

- Dynamic Hess chart simulator illustrating nerve palsies and Herings laws of equal innervation,
- Programmable eye movement simulator allowing the user to “create “ nerve palsies and strabismus.
- Eye movement simulator with animations of basic vestibular reflexes and neuron arcs
- Eye movement simulator and neuronal path animations of various nuclear, supranuclear and nystagmus disorders
- Pupil disorder simulator with animations to simulated applications off diagnostic agents and animations of corresponding neuro-muscular reactions.

Conclusion:

We believe this will be a useful tool to all health professionals interfacing with, and for students of, neuro-ophthalmology.

References: None

Key Words: Teaching, Computing, iPhone, Eye Movement disorders, Pupil Disorders

Financial Disclosure: No financial disclosures at time of submission.

Poster 115

A new method for testing letter contrast sensitivity in a clinical setting: Comparing M&S smart system II letter contrast test with Pelli-Robson chart

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Introduction:

The M&S smart system II (MSSSII; M&S Technologies Inc, Illinois) is a novel computer-based contrast sensitivity assessment tool. The purpose of this study was to compare the utility of the MSSSII to the Pelli-Robson chart as a clinical test for contrast sensitivity.

Methods:

75 participants aged 5 to 59 (19 visually normal subjects and 56 patients) were recruited. They were tested during monocular viewing using MSSSII and the Pelli-Robson chart in random order. Agreement between the two methods was assessed using the Bland-Altman test.

Results:

Bland-Altman tests demonstrated that these two methods gave similar results. For visually normal subjects, the mean contrast sensitivity (\pm standard deviation) was 1.69 ± 0.18 with MSSSII and 1.63 ± 0.07 with the Pelli-Robson chart. For patients, the mean contrast sensitivity (\pm standard deviation) was 1.51 ± 0.23 with MSSSII and 1.55 ± 0.20 with the Pelli-Robson chart. A minor difference in the values was noted due to a slight variation in the log step progression between the two methods.

Conclusion:

The MSSSII and Pelli-Robson test shows comparable contrast sensitivity values for both patients and visually normal subjects, indicating that the MSSSII is a valid tool for measuring contrast sensitivity as an alternative to the Pelli-Robson chart. Initial configuration and calibration of the MSSSII is simple. Once calibrated, the MSSSII provides reproducible, consistent, simple and easy assessment of contrast sensitivity in a wide variety of ophthalmic conditions in both adults and children as young as 5 years of age.

References: None

Key Words: Pelli-Robson chart, M&S smart system II, Letter contrast sensitivity, acuity

Financial Disclosure: The authors have no disclosures.

Poster 116

Driver Fitness in patients with Glaucoma and Cognitive Impairment

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Introduction:

Recent evidence suggests a relationship between cognitive impairment and glaucoma besides age alone. Whether impaired visual perception in glaucoma contributes to diminished cognitive function in patients with dementia, or cognitive impairment further limits visual perception due to optic nerve damage in glaucoma is unclear. Dimensions of vision beyond acuity can impair driver fitness. One objective of this study was to see if there were significant differences in age and measures of perceptual, cognitive and driving performance between older drivers with cognitive impairment and/or glaucoma and controls that still had good visual acuity. A second goal was to measure the strength of association between measures of visual, cognitive, and driving performance in these groups.

Methods:

302 older drivers were classified as having glaucoma alone (n=69), cognitive impairment alone (n=41), both (n=21) or neither (controls n=171). All participants had good visual acuity (20/40), a valid drivers license and were still driving. Demographic, health status, driving accidents and clinical tests of vision and driving performance (simulation and on-road) variables were analyzed using one-way ANOVAS and Pearson correlations.

Results:

Across clinical and driving measures, accidents and age, there were significant differences between those with cognitive impairment, with or without glaucoma, and controls. Subjects with glaucoma alone showed significant differences with controls on driving simulation tests and accidents. Differences between subjects with cognitive impairment, or glaucoma and cognitive impairment, only occurred on measures of self-reported visual health status, working memory (BVRT) and divided attention in the driving simulator. Driving simulator variables and measures of visual attention (UFOV) were significantly correlated with accidents.

Conclusion:

Driving simulation is a valid way to evaluate task performance and may be a more sensitive and salient method of detecting the additive and/or interactive effects of glaucoma and cognitive impairment in older drivers than vision and neuropsychological tests alone. More research is needed.

References:

1. Dawson, J.D., Anderson, S.W., UC, E.Y., Dastrup, E., & Rizzo, M. (2009). Predictors of driver safety in Alzheimer's disease. *Neurology*, 72:521-527.
2. Haymes SA, LeBlanc RP, Nicoleta MT, Chiasson LA, Chauhan BC. Glaucoma and on-road driving performance. *Invest Ophthalmol Vis Sci*. Jul2008;49(7):3035-3041.
3. Medeiros, F.A., Weinreb, R.N., Boer, E.R. & Rosen, P.N. (2010), Driving Simulation as a Performance-based Test of Visual Impairment in Glaucoma. *J Glaucoma* (in press)
4. Owsley C, McGwin G, Jr., Ball K. Vision impairment, eye disease, and injurious motor vehicle crashes in the elderly. *Ophthalmic Epidemiol*. Jun 1998;5(2):101-113.
5. Szlyk JP, Mahler CL, Seiple W, Edward DP, Wilensky JT. Driving performance of glaucoma patients correlates with peripheral visual field loss. *J Glaucoma*. Apr 2005;14(2):145-150. *study*. *Arch Ophthalmol*. Nov 2002;120(11):1509-1516.

Key Words: Glaucoma, Cognitive impairment, Driver fitness, Driving simulator, UFOV

Financial Disclosure: The authors had no disclosures.

Poster 117

Development of Clinical Markers to Avoid Unnecessary Temporal Artery Biopsy in Giant Cell Arteritis

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Introduction:

Giant cell arteritis (GCA) is a medium-to-large vessel vasculitis that, untreated, can cause blindness, myocardial infarction, and cerebral ischemia. Presentation may be highly non-specific, resulting in potentially morbid treatment with steroids and temporal artery biopsy (TAB). Depending on institution, only 10-30% of biopsies are positive for GCA.¹ If clinical features correlating with negative biopsy could be identified, risk-benefit ratio of biopsy could be more accurately assessed.

Methods:

This is a retrospective review of all patients over age 50 undergoing a temporal artery biopsy at our institution from 2007-2009 with a minimum follow-up of four months. Keyword search identified 186 cases. 86 did not meet inclusion criteria; 100 cases were included in final analysis. The electronic records were reviewed for biopsy result, diagnosis at final follow-up, and a total of 41 variables within the history.

Results:

100 patients were analyzed (14 positive; 84 negative, of which 13 were diagnosed with GCA in follow-up; and 2 non-ocular GCA cases). The following clinical and laboratory features showed trends toward statistical significance in differentiating true negative biopsy from positive cases: documented field deficit (p 0.02), frontal headache (p 0.04), leukocytosis with predominant neutrophils (p<0.001), thrombocytosis (p 0.04), history of renal disease with GFR<60 (p<0.001), obesity (p<0.001) and history of hyperlipidemia (p 0.01). Positive cases were distinguished from negative cases by high CRP (p 0.02), history of PMR (p 0.03), and sudden visual loss (p 0.04).

Conclusion:

Even after biopsy, deciding whether to treat for GCA may not be clear-cut. With this review we have identified a number of currently unreported features that may help distinguish patients likely to have truly negative TABs, potentially avoiding unnecessary biopsy and therapy. Next, we will examine whether an intermediate classification of pathological biopsy, in tandem with these newly identified clinical variables, might aid in more definitive diagnosis of GCA.²

References:

1. E Ypsilantis, ED Coutney, N Chopra, et al. Importance of specimen length during temporal artery biopsy. *British Journal of Surgery*. 2011; 98: 1556-1560.
2. L Zhou, K Luneau, CM Weyand, V Biouse, NJ Newman, HE Grossniklaus. Clinicopathologic Correlations in Giant Cell Arteritis: A Retrospective Study of 107 Cases. *Ophthalmology*. 2009 August; 116(8): 1574–1580.

Key Words: giant cell arteritis, temporal artery biopsy, negative biopsy

Financial Disclosure: The authors had no disclosures.

Poster 118

The Optic Nerve Head Component of the Multifocal Electroretinogram: A Novel Metric for Dissecting Pathophysiologic Mechanisms in MS

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Introduction:

To characterize abnormalities of a novel multifocal electroretinographic (mfERG) late response potential, the optic nerve head component (ONHC), in patients with MS and optic neuropathy. The retinal ganglion cell axons exit the eye through the lamina cribrosa, anatomically demarcating the transition from unmyelinated to myelinated axons, with axonal transmission properties thereby transformed from membrane to saltatory conduction.

Methods:

A novel mfERG protocol has been developed which applies global flash stimuli that are interleaved at specific intervals between the multifocal stimuli, thereby enhancing the ONHC potential. We evaluated normal subjects and MS patients with evidence of unilateral or bilateral optic neuropathy.

Results:

In contrast to normal subjects, MS eyes with optic neuropathy exhibited waveform disorganization or complete absence of the ONHC. Loss of the ONHC was correlated with reduced low contrast letter acuity (at 1.25% and 2.5%), retinal nerve fiber layer (RNFL) thinning by spectral domain OCT, and with abnormal timing responses on pattern reversal multifocal visual evoked potentials (mfVEP). Distinct quadrants of RNFL thinning topographically corresponded to localized ONHC changes within a discrete distribution of the retinal patch stimulus-response montage.

Conclusion:

We have characterized a heretofore, unrecognized electrophysiologic abnormality in MS patients with optic neuropathy; specifically the disorganization or loss of the ONHC. Future studies will be aimed at defining the utility of the ONHC for corroborating neuroprotective or even restorative effects of novel therapies. In particular, and in contrast to other assessments, this metric may represent a more sensitive and site-selective signature of chronic demyelination, and as such could be utilized as a reliable outcome in remyelination trials.

References:

Frohman, E.M., Fujimoto, J.G., Frohman, T.C., Calabresi, P.A., Cutter, G., et al. Imaging retinal architecture in multiple sclerosis with optical coherence tomography. *Nature Neurology* 2008;4:664-675. Sutter EE. The fast m-transform: a fast computation of cross-correlations with binary m-sequences. *Soc Ind Appl Math.* 1991;20:686-694. Hood DC, Seiple W, Holopigian K, Greenstein V. A comparison of the components of the multifocal and full-field ERGs. *Vis Neurosci.* 1997;14(3):533-544. Hood DC. Assessing retinal function with the multifocal technique. *Prog Retinal Eye Res.* 2000;19(5):607- 646.

Key Words: Multifocal ERG, Optic nerve head component, Retinal ganglion cells, Patter reversal stimulation, Saltatory conduction

Financial Disclosure: The authors had no disclosures.

Poster 119

Bilateral Retinal Artery Occlusion associated with Posterior Reversible Encephalopathy Syndrome

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Introduction:

Posterior reversible encephalopathy syndrome (PRES) has been described as reversible encephalopathy associated with hypertension, eclampsia, immunosuppressive agents. Transient cortical blindness is common in PRES but it is very rare that the occurrence of permanent visual loss because of a bilateral retinal artery occlusion associated with PRES.

Methods:

We described a case with a rare ocular complication and associated with PRES which has not been reported previously.

Results:

A 27-year-old primigravida patient was hospitalized at 37 weeks gestation because of pre-eclampsia. After administration, urgent cesarean section resulted in the birth of a healthy baby. Two days after surgery, her visual acuity decreased to hand motions at 30cm distance in both eyes. Magnetic resonance imaging (MRI), and magnetic resonance angiography (MRA) of the brain showed high signal intensity in bilateral occipital lobes in T2-weighted and FLAIR images and diffuse vasoconstriction of cerebral arteries. Routine ophthalmologic examinations revealed bilateral retinal artery occlusion. With a diagnosis of PRES(Posterior reversible encephalopathy syndrome), she was started on antihypertensive and conservative treatment. But the patient had a permanent bilateral visual loss.

Conclusion:

A clinical diagnosis of PRES includes the presence of headache, seizures, encephalopathy and visual disturbances, as well as radiologic findings of focal reversible vasogenic edema best seen on magnetic resonance imaging (MRI) of the brain. Both its clinical spectrum and underlying pathophysiology remain poorly defined.

Transient visual loss has been attributed to cerebral vasogenic edema secondary to PRES and it is considered as neuro-ophthalmologic emergency and important to correct causative factors. Irreversible visual loss caused by bilateral retinal artery occlusion following childbirth is a rare ocular complication and associated with PRES has not been reported previously.

References: None

Key Words: Bilateral retinal artery occlusion, Eclampsia, Posterior Reversible Encephalopathy Syndrome (PRES)

Financial Disclosure: The authors had no disclosures.

Poster 120

White Dots and Cerebral Spots: APMPE and Cerebral Vasculitis

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Introduction:

A 26-year-old man developed sequential left-sided numbness, left hand clumsiness, severe headache, and left monocular visual loss over two hours. He was found to have multiple strokes within the anterior and posterior circulations on MRI, and ophthalmologic examination was consistent with acute posterior multifocal placoid pigment epitheliopathy (APMPPE).

Methods:

Fluorescein angiography (FA), isocyanine green angiography (ICGA), optical coherence tomography (OCT), cerebral angiogram, laboratory evaluation for primary rheumatologic and infectious etiologies, and a brain biopsy were performed.

Results:

CSF examination showed a mild lymphocytic pleocytosis. FA, ICGA, and OCT were highly characteristic of APMPPE. Conventional cerebral angiogram showed vessel narrowing in a parietal branch of the right ACA and the origin of the left PICA. Brain biopsy, including immunohistochemistry, demonstrated focal areas of mild perivascular hemosiderin deposition, mild endothelial prominence, and scattered perivascular lymphocytes. Rheumatologic and infectious evaluations were notable only for low-titer ANA positivity and positive Anti-Ro ELISA 54.8 (reference 0-19.9).

Conclusion:

APMPPE is a rare inflammatory disorder of the choroid of unknown pathogenesis, and there are only 15 reported cases of associated stroke and cerebral vasculitis (1). Like our patient, the majority of cases suggest a small-vessel cerebral vasculitis with minimal signs of vessel narrowing on even conventional cerebral angiogram. The two cases of cerebral vasculitis confirmed by histopathology showed granulomatous inflammation within the walls of medium and large-size cerebral and meningeal arteries (2,3). Our pathology was suggestive of an endotheliopathy of small cerebral vessels and nearby inflammation, but it did not demonstrate granulomas. When radiographic findings suggest cerebral vasculitis, an association with APMPPE as confirmed by ICGA and FA is nearly pathognomonic of cerebral vasculitis. APMPPE-related cerebral vasculitis requires effective immunomodulatory therapy, and our patient was treated effectively with high dose intravenous corticosteroids followed by oral steroids and cyclophosphamide 2mg/kg/day. He has had no disease recurrence after twelve months.

References:

1. Luneau, Newman, Srivastava, Biousse. A Case of Acute Posterior Multifocal Placoid Pigment Epitheliopathy With Recurrent Stroke. *J Neuro-Ophthalmol*, 29: 111-118, 2009.
2. Wilson, Choromokos, Sheppard. Acute posterior multifocal placoid pigment epitheliopathy and cerebral vasculitis. *Arch Ophthalmol*, 106: 796-800, 1988.
3. De Vries, Dunnen, Timmerman. Acute posterior multifocal placoid pigment epitheliopathy with cerebral vasculitis: a multisystem granulomatous disease. *Arch Ophthalmol*, 124: 910-913, 1996.

Key Words: Acute posterior multifocal placoid pigment epitheliopathy (APMPPE), Cerebral Vasculitis, Inflammatory diseases, Immune modulatory therapy

Financial Disclosure: The authors had no disclosures.

Poster 121

The Efficacy of Toothpaste refraction in Children with Functional Visual Loss and its prognosis

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Introduction:

Children with functional visual loss are less sophisticated and highly suggestible, so may be easily coaxed with toothpaste refraction. Toothpaste refraction is a visual acuity testing start from the smallest line with suggestion. If this test is highly effective, it may reduce cost and confusion. But, it's efficacy has not been studied. Visual prognosis of these patients has not been studied also. We evaluated the efficacy of the toothpaste refraction in children with functional visual loss and the visual prognosis of the patients.

Methods:

Retrospective medical record review and prospective follow up was performed in 22 patients who were 5 to 16 years old, diagnosed with functional visual loss.

Results:

Toothpaste refraction was performed in 17 patients and 14 patients(82%) were proved to have corrected VA of 20/20 or better. The rest of the patents showed improved visual acuity more than 5 lines. Twenty patients were followed up and 17 patients(85%) of them were recovered normal VA or lost the symptom of decreased vision

Conclusion:

Toothpaste refraction is a very useful in proving non-organicity in children with functional visual loss. The prognosis for in children with functional visual loss is excellent.

References: None

Key Words: functional visual loss, toothpaste refraction, prognosis

Financial Disclosure: The authors had no disclosures.

Poster 122

Treatment of Functional Vision Loss with Duloxetine

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Introduction:

Functional Vision Loss (FVL) designates reduction in the subjective clarity of visual perception in excess of demonstrable objective visual performance. Although the etiology of FVL is uncertain, preliminary research suggests that anxiety and depression may play a role.¹ For this reason, the serotonin-norepinephrine reuptake inhibitor duloxetine may ameliorate FVL. Four patients diagnosed with FVL were treated with duloxetine and are presented here

Methods:

Four patients with FVL diagnosed by the author were treated with duloxetine and clinical examinations and follow up were recorded.

Results:

Two patients had severely reduced acuity on Snellen chart; the other two had normal acuities with subjective blur. Patient one had diabetic retinopathy with very poor visual field performance. Patient two had paracentral scotomata and a multifocal electroretinogram was non-diagnostic possibly due to eccentric fixation. Patients three and four had subjective blur and normal acuities and visual fields. Patient three had a posterior fossa arachnoid cyst; patient four had monocular aponeurotic ptosis. All patients had dramatic improvement in acuities and visual fields or subjective blur after addition of duloxetine 60 mg a day. Patient one stopped duloxetine and had recurrence of reduced acuity, abnormal visual fields, and poor visual performance.

Conclusion:

FVL is a common disorder that has been found in up to 11% of patients in a university neuro-ophthalmology practice.² It is also common in conjunction with organic illness.^{2,3} Despite this, treatment for this condition is lacking. These results suggest that addition of duloxetine, a medication with few adverse effects, may help improve visual dysfunction in patients suspected of suffering from FVL.

References:

1. Egan RA, Butler RW. Neuropsychological Profiles of Patients with Functional Vision Loss. *32nd Annual North American Neuro-Ophthalmology Society Meeting*, February 2006.
2. Scott JA, Egan RA. Concurrence of Functional Visual Loss with Organic Disease. *American Journal of Ophthalmology* 135: 670-5; 2003.
3. Ney JJ, Volpe NJ, Liu GT, Balcer LJ, Moster ML, Galetta SL. Functional Visual Loss in Idiopathic Intracranial Hypertension. *Ophthalmology*. 116(9): 1808-1813.

Key Words: Functional Vision Loss, duloxetine, functional visual loss, non-organic vision loss

Financial Disclosure: The author had no disclosures.

Poster 123

Bilateral endogenous endophthalmitis associated with meningitis in a healthy woman.

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Introduction:

Endogenous endophthalmitis is caused by the migration of the pathogen to the eye where it crosses the blood-ocular barrier and rare in immunocompetent patients. Here we describe a case of bilateral endogenous endophthalmitis secondary to bacterial meningitis with early detection and good visual recovery in a healthy patient.

Methods:

A 45-year-old woman with meningitis was referred to neuro-ophthalmologic clinic with acute visual impairment of both eyes. Moderate nuchal rigidity and continuous fever (38.5°C) were present. Her visual acuities were hand motions at 2 feet in both eyes. Ophthalmoscopy revealed inflammation in the anterior chamber and vitreous opacities of both eyes. She was diagnosed as having endogenous endophthalmitis associated with the meningitis. Vitreous sample was obtained for culture and vancomycin (1.0mg/0.1ml) and amikacin (0.4mg/0.1ml) were injected intravitreally.

Results:

Lumbar puncture revealed a cloudy cerebrospinal fluid (CSF) with leukocytes 468 cells/mm³ (72% neutrophils), glucose 34 mg/dl (116 mg/dl, serum glucose). Cerebral magnetic resonance imaging (MRI) showed diffuse prominent leptomeningeal enhancement. The pathogen responsible for the infection was not isolated in the vitreal, blood, cerebrospinal fluid samples. She was treated with intravenous ceftriaxone (6 g/day) and vancomycin (3 g/day) for 10 days. Vision improved to 20/20 in the right eye, and 5/100 in the left eye 1 month later. On fundus examination left eye showed occlusive retinal vasculitis and optic atrophy.

Conclusion:

The possibility of endogenous endophthalmitis secondary to bacterial meningitis in healthy patients should be considered. Endogenous endophthalmitis constitutes a rare complication of bacterial meningitis, and its prompt suspicious diagnosis and administration of empirical intravitreal antibiotics could lead to a more favorable visual prognosis.

References:

1. Miller JJ, Scott IU, Flynn HW Jr, Smiddy WE, Corey RP, Miller D: Endophthalmitis caused by *Streptococcus pneumoniae*. *Am J Ophthalmol*, 138, 231–236, 2004
2. Hidemasa T, Hiroshi M, Eiko S, Yuho I: Bilateral Endophthalmitis in a Patient with Bacterial Meningitis Caused by *Streptococcus pneumoniae*. *Ophthalmologica*, 222:357–359, 2008
3. Chan SM, Hodge WG, Leonard BC: Postoperative *Streptococcus pneumoniae* endophthalmitis complicated by meningitis. *Arch Ophthalmol*, 116, 951–953, 1998

Key Words: endophthalmitis, meningitis, endogenous endophthalmitis

Financial Disclosure: The authors had no disclosures.

Poster 124

Parry-Romberg Syndrome: Presentation of Two Patients; Review of Neuro-Ophthalmologic Manifestations

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Introduction:

Reports the neuro-ophthalmologic associations in patients with Parry-Romberg syndrome are infrequent.

Methods:

Case Report and Review of the Literature

Results:

A 59-yearold woman had visual loss of the right eye beginning in her 20, with ipsilateral optic atrophy and facial hemiatrophy, and, later, left sided trigeminal neuralgia. Examination noted VA OD NLP, OS 20/25, right optic atrophy, facial hemiatrophy, enophthalmos, and impaired sensation, and dysesthesias in the left trigeminal distribution.

A 64-year old woman had left facial hemiatrophy beginning in her 20s, later developing left trigeminal pain and anosmia, associated with dysguesia, dysarthria, dysphagia, Examination noted left enophthalmos, facial hemiatrophy, impaired abduction and elevation, impaired olfaction, taste, trigeminal sensation, and atrophy of the left tongue. Neuroimaging will be shown.

Conclusion:

Parry-Romberg syndrome, progressive facial atrophy, may be associated with trigeminal neuralgia, migraine, facial paralysis, epilepsy, cerebral hemiatrophy, and optic neuropathy. In one we report the association of optic nerve atrophy and in another patient various associated cranial neuropathies.

References:

1. Mazzeo, N, et al. Progressive hemifacial atrophy (Parry-Romberg syndrome): case report. *Oral Surg Med Oral Pathol Oral Radiol Endod.* 1995;79(1):30-35.
2. Stone J. Parry Romberg syndrome: a global survey of 205 patients using the Internet. *Neurology.* 2003;61(5):674-676.
3. Viana M, et al. Trigeminal neuropathic pain in a patient with progressive facial hemiatrophy (Parry-Romberg syndrome). *Arch Neurol.* 2011;68(7):938-942.

Key Words: Parry-Romberg Syndrome, Hemifacial atrophy, Optic atrophy, Enophthalmos

Financial Disclosure: The authors had no disclosures.

Poster 125

Appearance of cavernous angiomas after brain radiation therapy.

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Introduction:

Intracranial vascular formations usually run a benign course; however, they have potential for recurrent cerebral hemorrhage, epilepsy, and morbidity. An association between cerebral radiation therapy and de novo appearance of cavernous malformation has been described. We report 3 cases of cavernous angioma development following brain radiation therapy.

Methods:

This is a retrospective case series report of 3 adult patients seen in the neuro-ophthalmology clinic status post brain radiation therapy for malignancy (lymphoma, medulloblastoma) and found to have subsequent development of cerebral cavernomas.

Results:

The latency period for the detection of cavernoma following brain radiation ranged from 12 to 24 years.

Conclusion:

Radiation-induced cavernous malformations have previously been reported rarely, typically in the pediatric population. We describe 3 cases of de novo cerebral cavernoma development in adult patients following cerebral radiation therapy.

References: None

Key Words: cerebral angioma, venous angioma, cerebral malformation, radiation therapy

Financial Disclosure: The authors had no disclosures.

Poster 126

New Onset Leber's Hereditary Optic Neuropathy in a 79 year-old male

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Introduction:

Leber's Hereditary Optic Neuropathy (LHON) is a mitochondrial-inherited, degenerative, neuro-retinal disease presenting with rapid, bilateral sequential central visual loss. LHON typically affects males in their early twenties. Few cases of late onset LHON have been reported.

Methods:

We report a case of new onset LHON in a 79 year-old male. The literature on LHON is also reviewed.

Results:

A 79 year-old caucasian male presented with rapidly progressive, sequential vision loss over four weeks. His medical history included hypertension and prostate cancer. Ocular history included glaucoma and recent cataract extraction. Family history was remarkable for visual impairment in the patient's mother and maternal uncle from glaucoma.

Visual acuity was 20/400 bilaterally, with no afferent pupillary defect. Color vision was markedly reduced. The fundoscopic examination was unremarkable and visual field testing revealed ceco-central scotomata. The patient underwent an extensive workup. Genetic testing revealed a mutation at locus 11778 consistent with LHON.

Conclusion:

LHON typically presents with acute, painless, profound central vision loss in a sequential pattern. Vision loss occurs within 4-6 weeks of symptom onset and significant visual recovery is rare. One of three mitochondrial-DNA point mutations (11778 G>A, 14484 T>C, 3460 G>A) is responsible for 95% of diagnosed cases. Incomplete penetrance is common with roughly 50% of affected males, and 10% of females developing symptoms. Visual loss occurs prior to age 50 in 95% of cases. This patient represents a rare instance of late onset LHON. At the age of 79 years, he is likely the oldest known patient to manifest LHON. Awareness of late onset LHON is clinically important to avoid subjecting patients to unnecessary and potentially invasive diagnostic procedures. Detailed history and genetic testing is warranted in family members of affected patients.

References:

1. Man P, et al, Leber hereditary optic neuropathy, *Journal of Medical Genetics* 2002; 39: 162-169.
2. Newman N. Leber's hereditary optic neuropathy: New genetic considerations, *Archives of Neurology* 40, 540-548, 1993.
3. Newman N et al. Progression of visual field defects in Leber hereditary optic neuropathy: experience from the LHON treatment trial, *American Journal of Ophthalmology* 2006; 141:1061-1067.
4. Borruat FX, Late onset Leber's optic neuropathy: a case confused in ischaemic optic neuropathy. *British Journal of Ophthalmology* 1992; 76: 561-573.

Key Words: Leber's Hereditary Optic Neuropathy, Vision Loss, Elderly Patient, Genetic

Financial Disclosure: The authors had no disclosures.

Poster 127

Amaurosis fugax and stroke in a boy due to supernumary rib in the neck

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Introduction:

Cervical rib is the extra rib from the seventh cervical vertebra. Amaurosis fugax followed by stroke is a very uncommon manifestation of cervical rib.

Methods:

A fourteen years old boy presented with complaints of transient blurring of vision from right eye three weeks back. The visual blurring lasted for less than twenty minutes. Ten days later he had sudden weakness on left of body while he doing exercise. He did not give any history of headache, vomiting, seizures, loss of consciousness or neck pain. There was no history of breathlessness, chest pain and palpitation or pain in the arms. The blood pressure was not recordable in right upper limb while it was normal in left upper limb. The pulse was absent on right but was palpable on left side. Neuro-ophthalmic examination was normal. Neurological examination showed increased tone on left upper and lower limbs, power was 2/5 in left upper limb and 4/5 in left lower limb. Deep tendon reflexes were brisk and plantar was extensor on left side. Sensory examination was normal. Patient was investigated for Takayasu's arteritis or cardioembolic phenomenon. Haemogram, Blood-biochemistry, Coagulation and Vasculitis profile, ECG and Echocardiogram were normal. Computerized tomography (CT) of head showed right capsular infarct. CT angiogram revealed cervical rib causing, focal narrowing of right subclavian artery with post stenotic dilatation, arterial occlusion and collateral formation.

Results:

Transient reversal of flow in the subclavian artery together with the thrombus formation in the aneurysmal part of the subclavian artery has caused thromboembolic phenomenon in the internal carotid artery.

Conclusion:

Antegrade artery to artery thromboembolization from the aneurysmal segments of subclavian artery in the cervical rib has been reported in the literature. Retrograde artery to artery thromboembolization into the internal carotid artery may occur from aneurysmal segment of subclavian artery in cervical rib.

References: None

Key Words: Amaurosis fugax, supernumary rib, Cervical

Financial Disclosure: The authors had no disclosures.

Poster 128

Lymphocytic Hypophysitis

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Introduction:

Lymphocytic hypophysitis is a rare autoimmune condition characterized by pituitary expansion and hypopituitarism typically affecting women in the peripartum period. We report two cases of this rare condition, both initially presumed to be pituitary adenoma.

Methods:

Retrospective review of two patients with biopsy-proven lymphocytic hypophysitis.

Results:

A 34-year-old woman presented at 32-weeks gestation with episodic left temporal headaches. Visual acuity was reduced to 20/200 OS, accompanied by dyschromatopsia and a left relative afferent pupillary defect. Humphrey visual field testing revealed a bitemporal hemianopsia in conjunction with a 1.9x1.7cm pituitary mass on MRI imaging.

The patient underwent partial tumor resection for presumed pituitary adenoma. However, the procedure was halted when an intra-operative biopsy revealed a diagnosis of lymphocytic hypophysitis. As a result, she was started on treatment with oral steroids, resulting in marked visual recovery. However, she experienced recurrent vision loss following NSVD, prompting treatment with high-dose IV steroids, again with significant improvement. Interestingly, hypophysitis did not recur during a subsequent pregnancy that commenced 5 months later.

A 40-year-old woman presented with progressive occipital headaches at six years postpartum. Visual acuity and color vision were full, although a left relative afferent pupillary defect was elicited on exam. HVF testing revealed a bitemporal hemianopsia, while MRI showed the presence of a 1.0x1.2cm sellar mass.

As symptoms were refractory to treatment with bromocriptine and oral corticosteroids, surgical resection of the mass was ultimately performed. Pathology was consistent with lymphocytic hypophysitis.

While headaches and visual symptoms improved, post-operative course was complicated by marked thrombocytopenia. During a subsequent hospital admission, the patient ultimately succumbed to complications stemming from necrotizing enterocolitis.

Conclusion:

The discordance in presenting signs, treatment, and recovery in the two patients reported in our study reflects the wide spectrum of clinical presentation, management options, and disease course that may be seen in lymphocytic hypophysitis.

References: None

Key Words: Lymphocytic hypophysitis, Pituitary, Pregnancy

Financial Disclosure: The authors have no disclosures.

Poster 129

Electrophysiological Testing to Diagnose Sudden Bilateral Blindness from Cryptococcal Meningitis

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Introduction:

Cryptococcus neoformans is the leading cause of meningitis in patients with Acquired Immune Deficiency Syndrome and is associated with a high mortality rate. In patients who survive the infection, the most debilitating outcome appears to be visual impairment. Ocular complications of HIV-related cryptococcal meningitis are frequent, but complete binocular blindness is rare. We report a case of bilateral sudden blindness diagnosed with the help of electrophysiology.

Methods:

Our patient is a 45 year old African American female who presented with one month of painless, decreased vision in both eyes. She has a history of HIV and dementia. She was recently admitted for cryptococcal meningitis complicated by elevated intracranial pressure. On exam, her vision was bare light perception OU with no relative afferent pupillary defect. Extra-ocular movements were full with no response to optokinetic-nystagmus drum. Slit lamp and fundoscopic examination were unremarkable, with no papilledema or pallor visualized. MRI showed non-specific basal ganglia lesions and no mass.

Results:

Our patient underwent electrophysiologic testing to confirm the cause of visual loss. The Ganzfeld electroretinogram (ERG) was completely normal. Pattern-reversal visual evoked potentials (VEP) were extinguished, as were flash VEPs. These results, showing normal retinal function but unrecordable cortical responses, are consistent with cryptococcal meningitis.

Conclusion:

Mechanisms for binocular blindness due to cryptococcal meningitis include rapid-onset visual loss by direct fungal infiltration of the visual pathway, particularly at the chiasm, or slow-onset visual loss due to increased intracranial pressure. Our patient showed unremarkable fundoscopic examination findings with rapid onset visual loss, likely due to a retrobulbar fungal infiltration. When clinical suspicion is high for cryptococcus induced visual loss, without definitive exam findings, an ERG combined with a VEP can help elucidate the diagnosis.

References:

1. Cohen DB, Glasgow BJ. Bilateral optic nerve cryptococcosis in sudden blindness in patients with acquired immune deficiency syndrome. *Ophthalmology*. 100:1689–94; 1993
2. Golnic KC, Newman SA, Wispelway B. Cryptococcal optic neuropathy in the acquired immuno deficiency syndrome. *J Clin Neuro Ophthalmol*. 11: 96–103; 1991
3. Johnston SR, Corbett EL, Foster O, Ash S, Cohen J. Raised intracranial pressure and visual complications in AIDS patients with cryptococcal meningitis. *J Infect*. 24:185–9. 7; 1992
4. Kestelyn P, Taelman H, Bogaerts J, et al. Ophthalmic manifestations of infections with *Cryptococcus neoformans* in patients with acquiredimmuno- deficiency syndrome. *Am J Ophthalmol*. 116:721–7. 9;1993

Key Words: Cryptococcal Meningitis, Electrophysiology, Visual Evoked Potentials, Electroretinogram, Sudden Blindness

Financial Disclosure: The authors had no disclosures.

Poster 130

Occult Giant Cell Arteritis: Overcoming Hurdles to Diagnosis

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Introduction:

Giant Cell Arteritis (GCA) is a systemic vasculitis of the elderly characterized by new-onset headache, jaw claudication, and constitutional symptoms. Visual loss results from ischemic damage of the optic nerve via inflammatory and thrombotic occlusion of arterial supply. Temporal artery biopsy is the gold standard for diagnosis, usually guided by preliminary diagnostic tools. These include clinical presentation and elevated inflammatory markers of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). MR Imaging of the orbits rarely show any changes; with scant reports of optic nerve enhancement.

Methods:

We report an 84 year-old right-handed Caucasian male with no prior medical issues who presented with right eye vision loss and disc swelling. Initial ESR and CRP were within normal limits and MRI brain and orbits were unremarkable. At 3 day follow-up, worsening vision on the right prompted referral to our tertiary ophthalmology center. Patient denied any other symptoms typically associated with GCA. Ophthalmologic examination revealed bilateral disc swelling and disc hemorrhages. Visual acuity was light perception OD and 20/60 OS. Visual fields were unobtainable OD and full to confrontation OS. A right relative afferent pupillary defect was present and the left pupil reacted normally to light. Extra-ocular movements were intact. The rest of the neurologic examination was normal.

Results:

The lack of systemic symptoms and no elevation of inflammatory markers prompted a repeat MRI head which showed enhancement of the left optic nerve. Further systemic workup and lumbar puncture were normal. High sensitivity CRP (hsCRP) was elevated at 22.1 mg/L. Temporal artery biopsy confirmed GCA.

Conclusion:

Clinical ophthalmologic assessment remains critical in the diagnosis of giant cell arteritis. Optic nerve enhancement although not typical of GCA can be seen. hsCRP, an inflammatory marker used in cardiovascular and cerebrovascular disease, can be elevated in GCA, despite the lack of elevation of other inflammatory markers.

References: None

Key Words: Giant cell arteritis, Neuroimaging, High-sensitivity C-reactive protein, Occult, ophthalmologic assessment

Financial Disclosure: The authors had no disclosures.

Poster 131

Creutzfeldt-Jakob disease with cortical blindness

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Introduction:

CJD is a rare, fatal neurodegenerative disease caused by infectious prion. Visual impairment is one of the clinical manifestations. We present a patient diagnosed as CJD with typical clinical symptoms, brain imaging, abnormal EEG and cerebrospinal fluid 14-3-3 protein.

Methods:

Case report

Results:

A 56-year-old man presented with 2 months dizziness, blindness and progressive confusion. On examination, he was noncooperative, moderately impaired attention, concentration and memory, bilateral visual loss with pupils reactive to light. The optic disc was normal, a slight degenerative of bilateral macular. There was no myoclonic jerks but with rigid limbs. Blood tests were normal except elevated anti-thyroid antibody. Periodic sharp waves were found on EEG. Protein 14-3-3 was detected in CSF. The brain MRI in diffusion weighted image enhancement throughout the cortical ribbon of occipital lobe. Methylprednisolone 1g for 3d was given for 3d without any improvement. The patient died in one month after he left hospital.

Conclusion:

Cortical blindness might be a typical clinical feature to differentiate CJD from other encephalopathy diseases as Hashimoto encephalopathy, paraneoplastic and other infectious encephalopathy.

References: None

Key Words: Creutzfeldt-Jakob disease, cortical blindness, encephalopathy, paraneoplastic encephalopathy, Hashimoto encephalopathy

Financial Disclosure: The authors had no disclosures.

Poster 132

Web-based Neuro-Ophthalmic Patient Education

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Introduction:

For many patients seeking medical information, the Internet is a mixed blessing: although information is abundant, the challenge of finding reliable, useful sources can be insurmountable (Gremeaux 2010; Anderson 2008)

Methods:

We generated web-based educational content for the Neuro-Ophthalmology division webpage of our academic hospital website. The content was written by physicians and covered symptoms, diagnostic testing, treatment choices, and prognosis for common neuro-ophthalmic conditions. The question-answer format was user-friendly, visually-engaging, and free of any commercial bias. Patients seen in our hospital-based clinic were informed of the site. Interwoven Teamsite software was used to develop HTML pages. Webpage keywords were selected for search engine optimization. Google Urchin WebAnalytic software was used to track visitors to the site. Page views, duration of visit, and geographic origin were also recorded. Search terms were examined and educational content was modified to address frequent queries.

Results:

The addition of educational content to the website yielded immediate results. In the two-month periods before and after these changes, there was a 833% increase in first time visitors (from 33 per month to 308 per month), a 227% increase in average visit length (from 42s to 136s), and a 1,381% increase in page views per day (from 953 to 14,115). In this period, the new site drew visitors from 42 states and from 41 countries.

Conclusion:

Attempts to improve patient education offered on our Neuro-Ophthalmology website were successful. Web traffic data provide a useful quantitative metric to monitor the impact of these efforts. Improved Internet educational content is an important strategy in empowering patients with better understanding of a neurological condition, likely leading to improved patient outcomes.

References:

1. Gremeaux V, Coudeyre E: The Internet and the therapeutic education of patients: A systematic review of the literature. *Ann Phys Rehabil Med.* 2010; 53:669-92.
2. Anderson AS, Klemm P: The internet: Friend or foe when providing patient education? *Clin J Oncol Nurs* 2008;12:55-63.

Key Words: Patient education, Internet

Financial Disclosure: The authors had no disclosures.

Poster 133

Development of Smartphone Technology to Monitor Disease Progression in Multiple Sclerosis

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Introduction:

In Multiple Sclerosis, the accurate assessment of disease progression is critical to delivering appropriately individualized care (Amato 2007). Smartphones offer the potential for rich, real-time data capture that may fundamentally shift traditional paradigms of clinical monitoring (Boulos 2011).

Methods:

We established a consortium to build smartphone technology that tracks clinical changes in MS. The consortium consists of clinicians and scientists with expertise in multiple sclerosis, cognitive psychology, computer science, ethics, and regulatory issues. We plan three consecutive studies: first, to develop a suite of tests for a smartphone platform with encrypted data back-up and field-test the device; second, to expand the range of data collected through the smartphone (including measures of contrast sensitivity and conjugacy of eye movements); third, to assess the utility of this method for monitoring and predicting clinical disease progression. The consortium has created a suite of 21 custom applications for the Android smartphone platform, including standardized MS questionnaires and specific tests of color vision, dexterity, and cognition. Users will be presented approximately 2-5 applications each day, requiring about 5-10 minutes to complete. Collected data will be encrypted and securely stored on a cloud server.

Results:

The prototype phone, with a suite of clinical tests, is complete. The first study is scheduled to commence in November 2011, and results from this study will guide further development of the smartphone system.

Conclusion:

The members of this consortium represent a wide range of expertise, and their input has been fundamental to this effort to test the hypothesis that high-density, longitudinal data captured via smartphone technology can enhance the ability to monitor clinical symptoms in MS. Potential benefits of this approach include more accurate predictions of disease progression, improved design of clinical trials, and better treatment decisions for patients with MS and other chronic neurological diseases.

References:

1. Amato, M. P. and E. Portaccio. Clinical outcome measures in multiple sclerosis. *Journal of the neurological sciences* 2007; 259: 118-122.
2. Boulos, M. N., S. Wheeler, et al. How smartphones are changing the face of mobile and participatory healthcare. *Biomedical engineering online* 2011; 10: 24.

Key Words: Biomarker, Color vision, Multiple Sclerosis, Emerging technology

Financial Disclosure: Dr. Ramachandran is an employee of Vertex Pharmaceuticals. The remaining authors had no disclosures.

Poster 134

Rodent Model of Perioperative Ischemic Optic Neuropathy

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Introduction:

Perioperative ischemic optic neuropathy (ION) is a rare condition with increased prevalence in spinal and cardiac surgery. The mechanisms of perioperative ION are not known although factors such as hypotension, hemodilution, perioperative fluid administration, and patient positioning, among others, have been present in many affected patients. An animal model is necessary to evaluate the mechanisms of perioperative ION in a controlled manner.

Methods:

Adult Wistar rats were divided into four groups of 4-6 each. In three groups, animals were head down tilted 70 degrees for 5 h. An added control group was supine only, for the same duration. One group was moderately hemodiluted to hematocrit 25-30%. The third study group was tilted but not hemodiluted. Animals were monitored with non-invasive tail blood pressure and pulse oximetry. Blood was removed from a cannulated internal jugular vein, and replaced using hydroxyethylstarch. Recovery was assessed two weeks later using electroretinography (ERG) and histological examination of 4-micron thick retinal paraffin embedded sections, and immunohistochemistry was performed on 7 micron thick cryosections. The a- and b-waves and oscillatory potentials were measured before and 28 d later. P2 was derived using the Hood-Birch model. The scotopic threshold response (STR) is specific to retinal ganglion cells (RGCs) in the rat.

Results:

There were no changes in the a- or b-waves or the P2. The STR decreased 50%. We found no TUNEL staining in the retinal ganglion cells and no histological changes in the retina or the optic nerve.

Conclusion:

Moderate hemodilution and steep head-down tilt induced functional alterations in the retinal ganglion cells which were not evident as neuronal injury on histological examination. The followup measurement period may be too early to demonstrate histological damage, but this model does appear to reflect retrograde functional changes in the RGCs, possibly due to ischemia in the optic nerve.

References: None

Key Words: Ischemic optic neuropathy, Animal model, electroretinography, optic nerve

Financial Disclosure: The authors had no disclosures.

Poster 135

Risk Factors for Ischemic Optic Neuropathy After Spinal Fusion Surgery

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Introduction:

Ischemic optic neuropathy (ION) is the most common cause of perioperative visual loss after spine surgery. Previous studies to identify risk factors have been limited by lack of intraoperative data, small numbers cases, or heterogeneous case mixes. Consequently, risk factors for perioperative ION in spine surgery remain unclear.

Methods:

We utilized a multi-center case control design to compare 80 adult patients with ION from the ASA Postoperative Visual Loss Registry to 315 adult controls without ION after spinal fusion surgery. Controls were randomly selected from 17 institutions, and matched to cases by year of surgery. Pre-existing medical conditions and perioperative data were compared using multivariate logistic regression.

Results:

The multivariate predictive model was built in stages starting with those variables with $p < 0.2$ in the univariate analysis, including for Stage 1: sex, obesity, diabetes, hypertension, atherosclerosis, pre-op systolic blood pressure, mean arterial blood pressure; Stage 2: Wilson frame; Stage 3: anesthesia duration and estimated blood loss, Stage 4: lowest intra-op hematocrit, systolic or mean arterial blood pressure $> 40\%$ below baseline for > 30 min, colloid as percent of non-blood replacement. In the final multivariate model, risk factors for ION after spinal fusion surgery included male sex (OR = odds ratio: OR 2.53, 95% CI 1.35-4.91, $p = 0.005$), obesity (OR 2.83, 95% CI 1.52-5.39, $p = 0.001$), Wilson frame use (OR 4.30, 95% CI 2.13-8.75, $p < 0.001$), anesthesia duration (OR per 1 hr = 1.39, 95% CI 1.22-1.58, $p < 0.001$), estimated blood loss (OR per 1 liter = 1.34, 95% CI 1.13-1.61, $p = 0.001$), and colloid as percent of non-blood replacement (OR per 5% = 0.67, 95% CI 0.52-0.82, $p < 0.001$).

Conclusion:

Obesity, male sex, Wilson frame use, longer anesthetic duration, greater estimated blood loss, and lower percent colloid administration were significantly and independently associated with developing ION after spinal fusion surgery.

References: None

Key Words: Ischemic optic neuropathy, Retrospective study, Spine surgery, Optic nerve

Financial Disclosure: The authors had no disclosures.

Poster 136

A Case of Sudden, Reversible Vision Loss Secondary to Allergic Fungal Sinusitis

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Introduction:

Non-invasive fungal sinusitis occurs in patients with intact immune systems and is secondary to allergic over-responsiveness to fungi. Patients may have a marked allergic diathesis, such as atopy and the presence of nasal polyps. Ocular symptoms as the initial manifestation are rare. We present a case of sudden, reversible, vision loss from Allergic Fungal Sinusitis caused by *Bipolaris* species.

Methods:

A twenty-one year old male presented with one week history of sudden right eye swelling, pain, and decreased vision. BCVA was 20/40 OD with no rAPD. Extra-ocular motility of the right eye was limited in all directions and showed proptosis and hypoglobus with no palpable mass. CT showed a vascular mass involving the right nasal sinuses. MRI was suggestive of chronic inflammatory changes compatible with highly inspissated secretions versus fungal infection.

Results:

The patient was started on systemic antifungal therapy. The following day vision deteriorated to 20/400 OD; a right rAPD was present. The patient underwent endoscopic sinus surgery revealing viscous fungal material with mucinous secretions. Histologic examination consisted of eosinophilic material mixed with inflammatory cells. GMS stain showed numerous branching septated fungi. Lactophenol Cotton Blue stain revealed the fungus to be *Bipolaris* species. The patient was started on high dose IV steroids. Visual acuity recovered to 20/20 OU, with full extra-ocular motility and resolution of symptoms. The patient was diagnosed with a highly debilitating allergic form of rhinosinusitis caused by fungal elements, namely *Bipolaris*.

Conclusion:

Historically mistaken for a paranasal sinus tumor, allergic fungal sinusitis (AFS) is now believed to be an allergic reaction to aerosolized environmental fungi in an immunocompetent host. Our case highlights the need for clinical suspicion of AFS when an immunocompetent patient presents with rapidly progressive vision loss and signs of inflammation unresponsive to antifungal treatment. Timely intervention may help prevent permanent visual loss.

References:

1. Ponikau JU, Sherris DA, Kern EB, et al. The diagnosis and incidence of allergic fungal sinusitis. *Mayo Clin Proc.* 1999;74:877-884.
2. Chang WJ, Shields CL, Shields JA, et al. Bilateral orbital involvement with massive allergic fungal sinusitis. *Arch Ophthalmol.* 1996;114:767-768.
3. Jacobson M, Galetta SL, Atlas SW, Curtis MT, Wulc AW. *Bipolaris*-induced orbital cellulitis. *J Clin Neuro Ophthalmol.* 1992;12:250-256.
4. DeJuan E, Green WR, Iloff NT. Allergic periorbital mucopyocele in children. *Am J Ophthalmol.* 1983;96:299-303.
5. Dunlop IS, Billson FA. Visual failure in allergic aspergillus sinusitis: case report. *Br J Ophthalmol.* 1988;72:127-130.

Key Words: Allergic Fungal Sinusitis, Rhinosinusitis, *Bipolaris*

Financial Disclosure: The authors had no disclosures.

Poster 137

A slide is worth a thousand scans: Diplopia with a cavernous sinus metastasis of a remote endometrial stromal tumor

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Introduction: Structural lesions involving the visual pathways often require tissue diagnosis to guide therapy. However, pathology is not always definitive, and a biopsy showing poorly differentiated cells poses significant difficulty in identifying the primary tumor. We report a case of metastatic disease causing cavernous sinus syndrome in which biopsy yielded poorly differentiated tissue. The patient reported a remote uterine tumor, and only after obtaining slides from 7 years prior was the diagnosis made.

Methods:

Single Case Report

Results: A 54 year old woman presented with right ear pressure and hearing loss for 14 months with right temporal headaches and diplopia for 2 weeks. Previous imaging had detected a mass in the right cavernous sinus. Medical history was unremarkable aside from hypertension and a remote history of uterine tumor, resected 7 years prior.

Head CT and brain MRI both showed a right sphenoid sinus mass with bony erosion. Trans-sphenoidal biopsy revealed a spindle cell sarcoma with staining positive for vimentin, CD34, SMA and negative for HMB-45, S-100, p63, EMA, PGR, desmin, CAM5.2, MSA and pan-keratin consistent with mesenchymal origin including fibrosarcoma, leiomyosarcoma and malignant glomangiopericytoma. Subsequently, pathology slides from the patient's prior uterine tumor were obtained from an outside hospital. They showed a spindle cell tumor with staining positive for DC10 and BCL2 and negative for desmin, WT-1, s-100 and cytokeratins including AE1/AE3, CAM 5.2, EMA, CK7 and CK19 with less than 5% positive staining for ER and PR with the pathologists favoring an endometrial stromal tumor. This prior pathology permitted definitive diagnosis. Clinically, the patient progressed to a complete cavernous sinus syndrome.

Conclusion: The patient presented with a clear sphenoid sinus mass with unusual pathology. The nature of the prior uterine resection was not known initially. Only after obtaining outside slides from 7 years prior permitted a definitive diagnosis of metastatic endometrial stromal tumor to the right sphenoid sinus and cavernous sinus resulting in cavernous sinus syndrome

References: None

Key Words: Cavernous sinus syndrome, metastasis, endometrial stromal tumor

Financial Disclosure: The authors had no disclosures.

Poster 138

Acute vision loss in intracranial hypertension secondary to choroidal neovascularization

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Introduction:

We observed a rare ocular complication of choroidal neovascularisation (CNV) in a patient with idiopathic intracranial hypertension (IIH). The mechanism and treatment of such cases remain controversial.

Methods:

A 43-year-old woman with poorly controlled IIH since 1999 complained of subacute vision loss, predominately centrally in the left eye. In addition, she had worsening intra-cranial noises and headaches

Visual acuities were 20/20 OD and counting fingers 1 foot OS. She had a left RAPD with decreased color vision OS. Visual fields were constricted bilaterally, worse on the left, with dense superior and inferior arcuate defects OD. There was bilateral papilledema, worse on the left. In addition, there were peripapillary hemorrhages, exudates and choroidal folds OS. She also had subretinal and intraretinal hemorrhage in the macula. OCT showed peripapillary and subretinal fluid OS. Fluorescein angiography showed CNV superior-temporally to the disc OS. Neuroimaging was negative for any intracranial process or venous thrombosis. Opening pressure on lumbar puncture was 36 cm of water with normal components

Results:

The patient underwent an uncomplicated ventriculoperitoneal shunt and intravitreal injection of bevacizumab OS. Vision improved substantially in both eyes.

Conclusion:

CNV associated with papilledema from IIH is rare. Approximately nineteen cases have been reported in literature. The cause of the CNV may be due to disruption of the Bruch's membrane junction at the border of the optic nerve with subsequent subretinal fluid accumulation and neovascularization. There is still on-going discussion regarding how to treat the CNV. Some patients have had substantial visual recovery with lowering the intracranial pressure and the use of anti-VEGF agents, as seen in our patient.

References:

1. Hoye VJ III, Berrocal AM, Hedges TR III, Amaro-Quireza ML. Optic coherence tomography demonstrates subretinal macular edema from papilledema. *Arch Ophthalmol*, 119(9):1287-129, 2001
2. Jamerson SC, Arunagiri G, Willis Brian, Lesy MJ. Intravitreal bevacizumab for the treatment of choroidal neovascularization secondary to pseudotumor cerebri. *Int Ophthalmol*, 29, 183-185, 2009
3. Sathornsumetee B, Webb A, Hill DL, et al. Subretinal hemorrhage from a peripapillary choroid neovascular membrane in papilledema caused by intracranial hypertension. *J Neuro-Ophthalmol*, 26, 197-199, 2006
4. Aslan O, Acaroglu G, Ozdamar. Regression of peripapillary neovascular membrane following lumbar peritoneal shunting in a case of idiopathic intracranial hypertension. *Neuro-ophthalmology* 33,195-198, 2009

Key Words: idiopathic intracranial hypertension, choroidal neovascularization

Financial Disclosure: The authors had no disclosures.

Poster 139

Congenital Mydriasis Associated with Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS)

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Introduction:

Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) is a rare, congenital disease affecting smooth muscle peristalsis in the gastrointestinal tract and urinary bladder. We report the second case of bilateral mydriasis associated with MMIHS and present the first report of pilocarpine testing which suggests MMIHS is caused by an underlying smooth muscle myopathy.

Methods:

Case report

Results:

A 23-year-old Caucasian female was referred to our institution 28 weeks into her second pregnancy after an abnormal fetal ultrasound. A high-resolution ultrasound at our facility found a large female fetus, bilateral hydronephrosis, and a markedly distended fetal urinary bladder. Fetal MRI confirmed the ultrasound findings and also demonstrated a small sigmoid and descending colon raising suspicion for MMIHS.

The patient was born at 35-weeks and appeared neurologically normal for her age except for prominent bilateral mydriasis. Inability to tolerate feeds persisted throughout her hospital course requiring nourishment with total parenteral nutrition (TPN). At 8-weeks of age exploratory laparotomy was performed with resection of non-viable small intestine although she remained dependent on TPN.

Exam at 3.5 months of age demonstrated intermittent fixation and follow OU. Motility, alignment, facial grimacing, and corneal sensation were normal with no nystagmus. Portable slit lamp examination was unremarkable except for bilateral 7 mm nonreactive pupils with hypotrophic appearing irides. Fundus examination was normal in both eyes. Pupillary testing OU with pilocarpine 0.125% and pilocarpine 1% failed to show any pupillary constriction. MRI of the brain and orbits was unremarkable and an electroencephalogram revealed no epileptiform activity.

Conclusion:

We present a case of MMIHS-associated congenital mydriasis. The lack of pupillary response to pilocarpine 1% would suggest that mydriasis in MMIHS represents either a primary or secondary smooth muscle myopathy and may have important implications in the understanding of the disease.

References: None

Key Words: megacystis microcolon intestinal hypoperistalsis syndrome, congenital mydriasis

Financial Disclosure: The authors had no disclosures.

Poster 140

Curved fingers and tight eyes: distal arthrogryposis and 'ophthalmodesmosis'

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Introduction:

The abnormalities underlying congenital ophthalmoplegias are being increasingly recognised as multi-factorial. We present a case where there may be an analogous process giving rise to both arthrogryposis and a pseudo-Brown syndrome.

Methods:

Case report; eye movement recording; muscle biopsy.

Results:

A male ambulance driver presented at age 53 years with intermittent diplopia, especially on looking up. He was found to have distal arthrogryposis and limitation of elevation and abduction of both eyes. His son was a similar phenotype.

He had mild bilateral ptosis with dipping of the eyes on adduction, limited abduction and reduced upgaze, especially in adduction. Hess Chart demonstrated limitation of elevation and adduction of both eyes with particular under-action of inferior oblique.

Forced duction test was positive. The vertical and horizontal saccades appeared clinically to be of normal velocity. Eye movement recording showed peak velocities of over 600 deg/sec (lower limit of normal = 200) for horizontal movements (amplitude of vertical movements was too low for recording).

A muscle biopsy of vastus lateralis showed changes in keeping with a mitochondrial cytopathy.

Conclusion:

Our patient's limb contractures are in keeping with Distal Arthrogryposis (DA). He has an autosomal dominant DA with ocular motility limitation (DA type 5). The ocular motility limitation may be under-recognized. The positive forced duction test and normal/ high saccadic speed is supportive of a mechanical limitation. We propose that lack of foetal eye movements may give rise to a picture of "tethering" of ocular movements analogous to the limb abnormalities – 'ophthalmodesmosis'. Biopsy evidence of a mitochondrial cytopathy has not been previously reported in DA Type 5. The underlying genetic abnormality is unknown.

References:

1. Hall JG, Reed SD and Greene G. The Distal arthrogryposes: delineation of new entities – review and nosologic discussion. *Am J Med Genet* 1982;11:185-239
2. Bamshad M, Van Heest AE, Pleasure D. Arthrogryposis: a review and update. *J Bone Joint Surgery* 2009; 91 Suppl 4: 40-46

Key Words: Distal arthrogryposis, Brown syndrome

Financial Disclosure: The authors had no disclosures.

Poster 141

Oculomotor palsy and contralateral hemianesthesia & hemiparesis secondary to rhombencephalitis presumably due to CNS infection with *Listeria monocytogenes*

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Introduction:

The differential diagnosis of subacute and progressive midbrain dysfunction accompanied by constitutional symptoms includes CNS neoplasms, infections, inflammatory and demyelinating disorders.

Methods:

A 64 year old woman was referred with diplopia, right ptosis, and ascending left hemibody numbness and weakness for three months, accompanied by malaise, fatigue, anorexia, and 35 lb. weight loss. She was diabetic, hypertensive, and smoked cigarettes. On exam, she had partial, pupil-involving right oculomotor palsy, dysarthria, left extremity weakness, and ataxia of left extremities, posture & gait. Brain CT scan was normal. Brain MRI revealed a right midbrain lesion, best seen on FLAIR, but not on diffusion-weighted study, with minimal peripheral linear enhancement on T1 post-contrast study. NMR spectroscopy and PET scan were non-diagnostic. CT scans of the chest, abdomen, and pelvis were negative. CSF study was unrevealing on three occasions. Multiple CSF and blood cultures for bacteria were negative. Extensive tests for autoimmune diseases, viruses, mycobacteriae, fungi, spirochetes, and mycoplasmae were negative. Biopsies for *T. whipplei* were negative. After a three-day course of IV corticosteroids, her condition worsened, and the MRI lesion enlarged.

Results:

Based on MRI appearance, a presumptive diagnosis of CNS Listeriosis was made, and she was given a 6-week course of IV ampicillin. Her oculomotor palsy resolved, and her other neurologic deficits improved. Five weeks after discharge, she had mild cognitive problems, minimal gait and postural ataxia, but was independent.

Conclusion:

Although all blood and CSF cultures were negative, we believe that our patient's illness was due to CNS infection by *Listeria monocytogenes*, based on lesion morphology & localization, and dramatic improvement with antibiotic therapy.

References:

1. Antal E-A, Loeberg E-M, Maehlen J. Neuropathological findings in 9 cases of *Listeria Monocytogenes* brain stem encephalitis. *Brain Pathol* 15:187-191, 2005.
2. Armstrong RW, Fung PC. Brainstem Encephalitis (Rhombencephalitis) due to *Listeria monocytogenes*: Case report and review. *Clin Infect Dis* 16(5):689-702,1993.
3. Drevets DA, Bronze MS. *Listeria monocytogenes*: epidemiology, human disease, and mechanisms of brain invasion. *FEMS Immunol Med Microbiol* 53:151-165, 2008.
4. Vasquez-Boland JA, Kuhn M, Berche P, et al. *Listeria* pathogenesis and molecular virulence determinants. *Clin Microbiol Rev* 14(3):584-640, 2001.

Key Words: Rhombencephalitis, *Listeria monocytogenes*, oculomotor nerve palsy

Financial Disclosure: The authors had no disclosures.

Poster 142

Orbital Inflammation as a Paraneoplastic Manifestation of non-CNS Lymphoma

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Introduction:

Orbital lymphoma may present with proptosis, diplopia, and/or vision loss. We present a case of recurrent orbital inflammation and diplopia in the setting of systemic lymphoma without orbital involvement.

Methods:

Clinical case report

Results:

A 55 year old man presented with subacute onset of binocular horizontal double vision, right eye tearing, and mild facial numbness. Orbital MRI showed enlargement of all 4 rectus muscles of the right eye with right optic nerve sheath enhancement extending into the cavernous sinus. He was prescribed prednisone for orbital pseudotumor after an extensive but unrevealing systemic assessment. His diplopia improved but then recurred upon steroid taper. He then developed left eye ptosis and motility deficit that developed over several days but resolved spontaneously over the next few weeks. Repeat head and body imaging (including FDG-PET) was unremarkable, large volume repeat LP was normal, and inflammatory serologic studies were normal except for mildly increased ACE level. Several months later, an apparent mass lesion in the right cavernous sinus, Meckel's cave and pterygopalatine fossa was seen on MRI scan. When diplopia when recurred 2 months later and the patient complained of new back pain, FDG-PET scan showed a small round lytic lesion in T12 vertebral body, and MRI indicated abnormal signal in bilateral cavernous sinus and skull base. Vertebral core biopsy showed large B-cell lymphoma and further studies showed a potential primary source in the testes. Combined R-CHOP chemotherapy and local radiotherapy induced apparent disease remission until several weeks ago when a new optic neuropathy was found.

Conclusion:

Given the absence of malignant cells in the CSF on multiple lumbar punctures, it is possible that the neuro-ophthalmic disorders arose from a paraneoplastic syndrome. Specific markers are being investigated at this time in the setting of unremarkable repeat cranial MRI.

References: None

Key Words: lymphoma, diplopia, orbital disorder, paraneoplastic syndrome

Financial Disclosure: The authors had no disclosures.

Poster 143

Progressive, Painful Vision Loss in a Young Female with Systemic Sarcoid due to Meningothelial Meningioma

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Introduction:

Meningioma of the optic nerve typically presents with optic nerve atrophy, optociliary shunt vessels, and painless vision loss. We present a 46-year-old Caucasian woman with systemic sarcoid (liver biopsy) with progressive, painful vision loss, choroidal folds, and a hyperemic optic nerve believed to be due to neurosarcoid. Biopsy of the lesion, however, revealed a meningothelial meningioma.

Methods:

A 46-year-old woman with history of systemic sarcoid and a right optic nerve lesion presented with further vision loss and eye pain on the right. The optic nerve lesion was diagnosed over 10 years ago and was presumed to be due to sarcoid. The optic nerve lesion, however, never responded to treatment with steroids nor methotrexate. She then presented to our institution due to worsening vision and pain.

Results:

The patient's vision was 20/80 with an afferent pupillary defect and decreased color vision on the right side. Ophthalmic exam on the left side was unremarkable. She also had restricted motility and mild proptosis on the right. Posterior segment exam revealed a hyperemic, swollen optic nerve with choroidal folds extending to the macula. Visual field testing showed constricted fields on the right. Repeat MRI revealed enlargement of the right optic nerve lesion. Suspicion of the original diagnosis due to lack of response to treatment in the past prompted a biopsy with minimal debulking. Biopsy revealed a meningothelial meningioma, not sarcoid. One week post-operatively, the patient's vision was 20/25 and her color vision and visual field returned to normal, although it subsequently declined over time.

Conclusion:

This case of a patient with systemic sarcoid and an optic nerve meningioma initially diagnosed as neurosarcoid illustrates the importance of performing a biopsy to establish the diagnosis, especially when not responsive to treatment. In addition, this case highlights meningiomas can also present atypically without the characteristic triad.

References:

1. Yanoff M, Duker J. Optic nerve sheath meningiomas. *Ophthalmology*; Mosby. 982-983, 2009.
2. Ortiz O, Schochet SS, Kotzan JM, Kostick D. Radiologic-pathologic correlation: meningioma of the optic nerve sheath. *Am J Neuroradiol*. 17:901-906, 1996.
3. Miller NR. Primary tumours of the optic nerve and its sheath. *Eye*. 18:1026-1037, 2004.
4. Saeed P, Rootman J, Nugent RA, White VA, Mackenzie IR, Koornneef L. Optic nerve sheath meningiomas. *Ophthalmology*. 110(10):2019-2030, 2003.

Key Words: Optic nerve meningioma, neurosarcoid

Financial Disclosure: The authors had no disclosures.

Poster 144

There is no such thing as too much saline.

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Introduction:

52 yo Jamaican female presented in January 2011 with “nausea and difficulty swallowing her saliva”. These symptoms progressed until patient was unable to eat. An MRI of the brain, February 2011, was read as normal. Her other workup included a negative GI work-up. Blood work normal. The symptoms continued to progress and the patient was admitted for dehydration multiple times from February to March 2011 and received saline at the ER. Her symptoms continued, and in March 2011 she was vomiting 2x daily. The patient was evaluated by an ophthalmologist who noted nystagmus. Neuro-ENT ordered an MRI 05/02/2011 and the patient was admitted. VA at distance with correction was 20/25 in the right eye, and 20/25+2 in the left. Her EOMs showed slowed saccades to left OU. The patient exhibited Brun’s nystagmus (vestibular nystagmus combined with gaze paretic nystagmus). The patient was found to have CN VII, IX, X, XII affected on left with “gaze-evoked nystagmus”, and gait difficulties.

Methods:

Case presentation

Results:

MRI showed a region of restricted diffusion, enhancement, and T2 hyperintensity within the posterior left medulla. This was most compatible with an active demyelinating plaque, and less likely subacute infarct or neoplasm, given its rapid increase in size. MR Spectroscopy was performed within the left dorsal medullary lesion and showed an elevated choline to creatinine ratio.

Conclusion:

Our differential diagnosis included multiple sclerosis, extrapontine myelinolysis, a post-viral infection, and sarcoidosis. Due to the patient’s long standing nausea and vomiting, along with her history of multiple visits to the ER for saline boluses, our initial diagnosis was extrapontine myelinolysis. The patient was placed on steroids and the clinical exam followed daily. Upon follow up with MRI, patient was determined to have a diagnosis of Multiple Sclerosis.

References:

1. Kumar, Central Pontine Myelinolysis, an update; Neurologic Research, volume 28, page 360-366, April 2006.

Key Words: Central pontine myelinolysis, MS, demyelinating plaque

Financial Disclosure: The authors had no disclosures.

Poster 145

Eosinophilic Granuloma: Evolution of Diagnosis and Treatment

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Introduction:

Eosinophilic granuloma is a rare disease affecting young children. When symptomatology and clinical course mimics an infectious process, diagnosis of this neoplasm can pose a unique dilemma. We present a case of localized eosinophilic granuloma of the orbit with extradural extension.

Methods:

Retrospective case report.

Results:

Previously healthy 27 month old boy with 6 weeks of unilateral periorbital edema, erythema and slight proptosis diagnosed and treated as orbital cellulitis by pediatrician is sent for neurosurgical evaluation. Parents note the swelling resolved with the antibiotics and time. On exam he is central, steady and maintained OU, has full extraocular movements, equal pupils without RAPD. On external exam the lids are without erythema or swelling, there is slight proptosis and subtle resistance to retropulsion OD. Outside MRI and CT scan demonstrate a right superior lateral orbital process which extends through the orbital roof but is extradural. An orbital biopsy was performed. On hematoxylin and eosin stained slides there are sheets of histiocytes with occasional multinucleated cells on a background of eosinophilic and lymphocytic inflammation. The Langerhans cells express Langerin (CD207), CD1A and CD68. S-100 stain highlights few dendritic cells.

Conclusion:

Pathologic diagnosis is consistent with Langerhans Cell Histiocytosis. The child underwent systemic oncologic evaluation followed by treatment with vinblastine and prednisone. Learning points of the case include that there is a relatively new stain called "Langerin" (CD207), thought to be specific for the Birbeck granules, and that the Histiocyte Society recommends systemic chemotherapy for patients with maxillofacial involvement.

References:

1. Minkov M. Multisystem Langerhans cell histiocytosis in children: current treatment and future directions. *Paediatr Drugs*, 13(2), 75-86, 2011.
2. Vosoghi H, Rodriguez-Galindo C, Wilson MW. Orbital involvement in langerhans cell histiocytosis. *Ophthal Plast Reconstr Surg*, 25(6):430-3, 2009.
3. Woo KI, Harris GJ. Eosinophilic granuloma of the orbit: understanding the paradox of aggressive destruction responsive to minimal intervention. *Ophthal Plast Reconstr Surg*, 19(6):429-39, 2003.
4. Song A, Johnson TE, Dubovy SR, Toledano S. Treatment of recurrent eosinophilic granuloma with systemic therapy. *Ophthal Plast Reconstr Surg*, 19(2):140-4, 2003.

Key Words: Eosinophilic Granuloma, Langerin Stain, Orbital Cellulitis, Histiocyte, Chemotherapy

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Poster 146

Primary CNS Non-Hodgkin B cell Lymphoma of L frontal lobe s/p chemo and XRT recurs as a smoldering uveitis and infiltrative optic neuropathy OS

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Introduction:

Primary CNS Lymphoma is an uncommon extra-nodal Non-Hodgkin Lymphoma (NHL). Up to 40% of NHL patients present with fever, weight loss, and sweats⁽¹⁾. Lymphadenopathy and extra-nodal findings are common. Ocular involvement can occur as a primary ocular, central nervous system, or isolated intraocular lymphoma⁽²⁾. Optic neuropathy can lead to rapid visual loss⁽²⁻⁵⁾.

Methods:

Case Report

Results:

70 year-old WF w/ CNS Non-Hodgkin B cell Lymphoma L frontal lobe s/p chemo/XRT c/o blurry vision OS (9/00) describing "ink blobs" and intermittent flashes. Optic neuropathy OS was diagnosed due to VA 20/60, Color 3/14, 0.9 log RAPD, and central scotoma. MRI was normal. Vitreous cells OS w/ disc leakage on FA were found, and prednisone was started for uveitis. A diagnostic vitrectomy showed rare lymphocytes and histiocytes. Flow cytometry was negative. Vision improved to 20/30 OS by 3/01 after subtenons triamcinolone. Recurrent uveitis OS continued to smolder for the next four years. MRI was again normal in 10/05 and 3/07. Vision OS next decreased abruptly to CF@1 foot in 5/08 w/ a 1.5 log RAPD. A superior hemiretinal artery/vein occlusion was managed by Retina. She returned to Neuro-op in 9/09 complaining that her OS went blind about 1 year ago. She was more concerned now about her OD which suddenly lost vision recently. Her vision was 20/70 and NLP. A temporal hemianopia was found OD.

Conclusion:

MRI 9/09 revealed circumferential thickening of the globe OS w/ enhancement of the left optic nerve, chiasm, and optic tract. This correlated with her temporal hemianopia OD. Enucleation of the blind, painful left eye confirmed the diagnosis of Non-Hodgkin B cell Lymphoma. The patient was treated with her second course of XRT. The radiologic changes in the L optic nerve, chiasm, and tract quickly resolved, however, her vision and temporal hemianopia have remained stable.

References:

1. Anderson, Chabner, Young, Berard, Garvin, Simon, DeVita. Malignant lymphoma. 1. The histology and staging of 473 patients at the National Cancer Institute. *Cancer*. 50(12):2699-2707, 1982.
2. Kim, Shah, Arora, Solanki. Isolated optic nerve infiltration in systemic lymphoma--a case report and review of literature. *Ophthal Plast Reconstr Surg*. 26(4):291-293, 2010.
3. Siatkowski, Lam, Schatz. Optic neuropathy in Hodgkin's disease. *Am J Ophthalmol*. 114:625-629, 1992.
4. Kettani, Lamari, Lahbil. Bilateral optic neuropathy and non-Hodgkin's lymphoma. *Bull Soc Belge Ophthalmol*. 300:35-39, 2006.
5. Kay. Optic neuropathy secondary to lymphoma. *J Clin Neuroophthalmol*. 6:31-34, 1986.

Key Words: Lymphoma, Uveitis, Non-Hodgkin Lymphoma, Infiltrative Optic Neuropathy

Financial Disclosure: The authors had no disclosures.

Poster 147

Leber Hereditary Optic Neuropathy (LHON): a novel mutation?

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Introduction:

Leber hereditary neuropathy is a maternally inherited mitochondrial disease with diagnosis is confirmed by genetic analysis (blood test): screening for three primary mutations in the mitochondrial ADN (location 11778 or less commonly at the 3460 or 14484), we describe a patient with LHON with a possible novel mutation

Methods:

Review of medical history of a patient with progressive optic neuropathy . Review of the literature.

Results:

33 years old patient complains of headaches and rapidly bilateral progressive vision loss. After 45 days of the onset of symptoms he presents VA counters fingers OU – bilateral relative cecentral scotomata – sluggish reactive pupil – optic nerve with focal blurring and congestion. VEP prolonged latency symmetrical – BEAP impaired. MRI normal. Toxic and nutritional compromise were ruled out. No family history of visual impairment was found.

The patient was treated previously in another center with steroids with no improvement of his clinical condition. Suspecting LHON, genetic examination was performed and a new possible mutation for Leber was found.

Patient progressed to temporal pallor and papille excavation; VA deteriorated to light perception despite of idebenone treatment.

Bilateral and rapidly progressive optic neuropathy conducted to LHON diagnosis. Genetic examination was normal for usual mitochondrial mutations in MT-NG6 – MT-ND4 y MT-ND1 genes, but sequencing of MT-ND1 gen identified a m.3547 A>G mutation which we couldn't find reported associated with LHON. However it has been found related, in a small group of patients in Colombia, with mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS). This mutation induces a change in proteins translation orientation changing in 81 position Isoleucine for Valine

Conclusion:

Initially patient was treated with corticosteroids suspecting optic neuritis with no improvement. Clinical manifestation oriented the diagnosis and genetic examination supported the LHON diagnosis nevertheless this mutation has not been reported in LHON patients but in MELAS ones. This is an interesting point of discussion because they can be overlapping syndromes although this patient has not manifestations of this disease

References:

1. Cornejo, Parra, Herrera, Gallego, Pineda, Bedoya, Ruiz Linares, Análisis molecular en 29 pacientes con diagnóstico presuntivo de mitocondriopatía: encefalopatía mitocondrial con acidosis láctica y aplopejía (MELAS) / Molecular analysis in mitochondrial encephalopathy with lactic acidosis and stroke (MELAS), Acta neurol. Colomb, 16(3):195-202, oct. 2000
2. Yen, Wang, Wei, Lebers Hereditary optic neuropathy: A multifactorial Disease, Retinal and Eye Research, 25, 381-396, 2006.
3. Finsterer, Inherited mitochondrial neuropathies, Journal of the Neurological Sciences 304 , 9–16, 2011

Key Words: LHON , possible novel mutation

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Poster 148

Neurocysticercosis and subsequent Multiple Sclerosis

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Introduction:

Both multiple sclerosis (MS) and neurocysticercosis (NC) are two entities in which clinical manifestations, immunoserologic assays and neuroimaging images are neither pathognomonic nor specific requiring for their diagnosis an accurate examination of the clinical history of patients and an meticulous and adequate follow-up

Methods:

Review of medical history of a patient with Neurocysticercosis and subsequent Multiple Sclerosis.
Review of the literature

Results:

Ten years-old patient presenting with a 4 months evolution of horizontal diplopia, with spontaneous partial improvement. He was initially given a viral diagnosis. V.A. 20/20 OU. Normal ocular examination. Right VI paresis. No neurological compromise. He provides C.T. and M.R.I. performed 1 month after the first symptoms, which are repeated after consult.

New studies evidence improvement of some lesions, but the emergence of new ones. L.P. is also conducted. Images compatible with Neurocysticercosis and + ELISA in serum, with negative L.P. for the parasite. He refers haven ingested wild boar meat previous to the commencement of clinical manifestations. No brain biopsy was performed. Systemic evaluation was negative for parasite compromise. Diagnosis of Neurocysticercosis is reached and treatment with Albendazol and corticosteroids begins.

No systemic nor neurologic compromise were found

In the course of treatment, there was partial improvement but the patient presented scalp and systemic paresthesia, as well as headaches. The evolution presented itself with relapses, because of which a new serie of treatment was done; it resulted in the resolution of the paresthesia but with persistence of the diplopia.

After 20 months from the beginning of clinical manifestations, loss of lower member strength appeared – V.E.P. with prolonged latency. Changes in the M.R.I. supported the diagnosis of M.S. beginning treatment with interferon

Conclusion:

Draws attention the dissociation between the minimum symptomatology and the major neurological compromise evidenced in the successive M.R.I. There are always difficulties while establishing a Neurocysticercosis diagnosis, particularly when encountering negative C.S.F. Another point of interest in the case is analyzing the possible correlation of Neurocysticercosis with subsequent M.S.

References:

1. Esclerosis múltiple y neurocisticercosis, Correspondencia: Dr. Alberto J. Dorta-Contreras. Laboratorio Central de Líquido Cefalorraquídeo (LABCEL). Apartado 10049. CP 11000. Ciudad de La Habana, Cuba. E-mail: adorta@infomed.sld.
2. J.J. Rodríguez-Uranga, A.J. Uclés-Sánchez, M.M. López-Muñoz, A. Serrano-Cabrera, *MULTIPLE SCLEROSIS AND NEUROCYSTICERCOSIS: A DIAGNOSTIC DILEMMA*, REV NEUROL 2004; 38 (9): 832-836, 2004.

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