

131 (Submitted)

Ocular microvascular function in Cerebral Small Vessel Disease

Hong Jiang^{1,2}, Delia Cabrera DeBuc¹, Aizhu Tao^{1,3}, Zhe Xu^{1,3}, Byron L. Lam¹, Jianhua Wang¹

¹Bascom Palmer Eye Institute, University of Miami, Miami, FL, USA, ²Department of Neurology, University of Miami, Miami, FL, USA, ³School of Ophthalmology and Optometry, Wenzhou Medical College, Wenzhou, China

Introduction:

Cerebral small vessel disease (CSVD) is a common devastating neurological disorder that presents as progressive cognitive decline and gait difficulty. The cause and treatment of CSVD remain unclear largely due to the difficulty in studying the small cerebral vessels in vivo. Ocular microvasculature is similar as the vasculature in central nervous system anatomically and physiologically. The purpose of this project was to demonstrate the feasibility of imaging ocular microvascular function in patients with CSVD.

Methods:

A retinal function imager (RFI, Optical Imaging Ltd, Rehovot, Israel) was used to image the retina for measuring velocity of blood flow and to image the conjunctiva for creating noninvasive capillary-perfusion maps (nCPMs). Eight CSVD patients (average age 62.9 ± 6.7 , 6 females and 2 males) were imaged to measure retinal blood flow velocity. Seven of them were imaged to create conjunctival nCPMs. Custom software was used to segment the nCPMs and fractal analysis was used to determine the microvascular density and complexity.

Results:

The average retinal venous velocity of CSVD patients was 2.4 ± 0.8 mm/s, which appeared lower than the published normality dataset with the same age range (~ 3.5 mm/s) (Burgansky-Eliash et al 2013). The nCPMs showed the conjunctival capillary network was loose and vessel tortuosity was obviously evident in 6 of 7 patients whose nCPMs were obtained. The fractal dimension of conjunctival nCPMs from 7 of these CSVD patients was 1.70 ± 0.05 .

Conclusions:

This study demonstrated the feasibility of measuring retinal venous blood flow velocities and obtaining conjunctival nCPMs for fractal analysis in patients with CSVD. CSVD patients appeared to have decreased retinal venous velocity, which may indicate hypoperfusion. Possible conjunctival vasculopathies such as tortuosity and loose capillary network may warrant further studies.

References:

- Burgansky-Eliash, Z., et al. "The correlation between retinal blood flow velocity measured by the retinal function imager and various physiological parameters." Ophthalmic Surg.Lasers Imaging Retina 44.1 (2013): 51-58.

Keywords: Miscellaneous, Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

132 (Submitted)

Asymptomatic Optic Neuritis in Neuromyelitis Optica and Multiple Sclerosis – a comparison.

Su Ann Lim¹

Tan Tock Seng Hospital, Singapore, Singapore

Introduction:

Chronic asymptomatic retrobulbar optic neuritis although rare has been reported in Multiple Sclerosis (MS)^{1,2,3}. We report two cases of asymptomatic chronic optic neuritis in MS and two cases in patients with Neuromyelitis Optica (NMO). Their clinical presentations as well as outcomes will be described and discussed.

Methods:

Retrospective case series

Results:

Two known MS patients and one with NMO had visual field abnormalities discovered during their routine follow-up visits. These were repeatable and progressive within a 2-3 week period. All were treated with pulsed steroid, two recovered vision while one did not. Another patient was found to have perception of light vision in one eye and optic atrophy at the time of diagnosis of NMO. No treatment was undertaken and vision has remained poor.

Conclusions:

Chronic asymptomatic retrobulbar optic neuritis is well documented in MS but can also occur in NMO. Vigilant follow-up is required to diagnose such cases. Other causes such as a compressive etiology should be excluded. Treatment with pulsed steroid may result in visual recovery and should be attempted. More studies should be undertaken to better understand the pathophysiology in these cases.

References:

- 1)Ashworth B. Chronic Retrobulbar and Chiasmal Neuritis. Brit J Ophthalmol 51, 698-702;1967.
- 2)Read SJ, Harrison JD, Pender MP. Progressive Visual Loss: An Unusual Presentation of Multiple Sclerosis. J Clin Neuroscience 3(3);264-6;1996.
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Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

133 (Submitted)

Bilateral Disc Swelling in Renal Failure; Hypertensive Optic Neuropathy or Papilloedema?

Su Ann Lim¹

Tan Tock Seng Hospital, Singapore, Singapore

Introduction:

Both hypertensive optic neuropathy and papilloedema present with bilateral disc swelling, normal visual acuity and colour vision in the acute stage. We present a patient with chronic renal failure, elevated blood pressure and bilateral disc swelling. Although initially diagnosed with malignant hypertension and hypertensive optic neuropathy, neuro-ophthalmological evaluation indicated papilloedema and lumbar puncture confirmed idiopathic intracranial hypertension (IIH). We will show photos to differentiate the clinical features of these two life- and sight-threatening conditions. We also review literature on the etiology of disc swelling in patients with renal failure and postulate on the mechanism of IIH in patients with renal failure.

Methods:

Retrospective case report

Results:

A 29 year old female with no past medical history presented to the emergency department complaining of mild bilateral blurring of vision. On examination, visual acuity, colour vision and confrontational visual fields were normal. Her blood pressure however was elevated at 237/112 mmHg and her creatinine was elevated at 1115umol/L. Humphrey visual field however revealed bilateral generalized depression and fundal examination revealed bilateral disc swelling. On detailed questioning, she had no headaches, double vision or tinnitus to suggest increased intracranial pressure. Despite this, increased intracranial pressure was suspected due to clinical signs which will be discussed. Magnetic resonance imaging including venography was normal and lumbar puncture revealed an opening pressure greater than 50 cm water with a diagnosis of IIH.

Conclusions:

Patients in chronic renal failure can present with disc swelling due to several causes, namely hypertensive optic neuropathy, anterior ischaemic optic neuropathy as well as IIH. It is important to recognize and differentiate these conditions so that appropriate treatment can be instituted. In addition, further studies should be undertaken to understand the pathophysiology of this condition.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

134 (Submitted)**Orbital Ultrasonographic Findings in Tumors to the Extraocular Muscles**

Giovanni Greaves¹, Kym Livingston¹, Grant Liu¹, Nicholas Volpe², Sonul Mehta¹, Madhura Tamhankar¹

¹*Departments of Ophthalmology and Neurology, University of Pennsylvania, Philadelphia, PA, USA,*

²*Department of Ophthalmology, Feinberg School of Medicine, Chicago, IL, USA*

Introduction:

The ultrasound characteristics of extraocular muscle (EOM) enlargement can help to differentiate thyroid orbitopathy from other causes such as neoplasm and inflammation¹. There are few reports in the literature describing ultrasonographic features in patients who present with neoplastic EOM enlargement¹⁻³. Our objective was to characterize the nature of ocular misalignment and

describe the orbital ultrasonographic findings in patients presenting with tumors to the EOMs.

Methods:

A retrospective chart review of all patients who underwent orbital ultrasonography for EOM enlargement from 2001-2013 was conducted. The details of clinical presentation and ultrasonographic features of EOM enlargement in patients with neoplastic causes were recorded. The reflectivity of the enlarged muscle was compared to ultrasonographic findings in patients with thyroid eye disease and myositis.

Results:

Amongst eight patients identified as having tumors to the EOMs, six of the eight patients had low reflectivity of the enlarged muscle when compared with its contralateral counterpart. The enlargement was 2-3 times the normal width of the muscle. Six of these patients had a past history of cancer while in two patients the EOM enlargement was the presenting feature of an underlying neoplasm. The most common symptoms were diplopia, eye redness, proptosis, swelling, restricted gaze and tearing. Seven of the eight patients exhibited some form of strabismus. Of this seven, four had paretic deviation, two had restrictive and one had a combined restrictive and paretic cause. Seven of the eight patients had one EOM involved whereas one patient had two EOMs involved.

Conclusions:

Patients with tumors to the EOM often have an underlying history of cancer. Our results demonstrate that these patients have a low reflective enlargement of the involved muscle and paretic patterns of deviations are more prevalent as compared to patients with thyroid eye disease. This enlargement is on the order of two to three times the thickness of its contralateral counterpart.

References:

- 1. Dick AD, Nangia V, Atta H. Standardised echography in the differential diagnosis of extraocular muscle enlargement. *Eye (Lond)*, 6(6):610-7, 1992.
- 2. DiBernardo C, Pacheco EM, Hughes JR, Iliff WJ, Byrne SF. Echographic evaluation and findings in metastatic melanoma to extraocular muscles. *Ophthalmology*, 103(11):1794-7, 1996.
- 3. Wiggins RE Jr, Byrne SF. Metastatic tumor to the extraocular muscles: report of 5 cases. *J AAPOS*, 16(5):489-91, 2012.

Keywords: Orbital and Eyelid Disorders, Neuro-Imaging (MRI, CT, etc), YES

Financial Disclosures: The authors had no disclosures.

137 (Submitted)

A Round-About Diagnosis: How a Metastasis could do anything.

Anna Gushchin¹, Talmadge Cooper

Stanford University/ Byers Eye Institute, Palo Alto, CA, USA

Introduction:

A patient presented to the acute care clinic for second opinion regarding vision loss in the right eye to no light perception.

Methods:

Case presentation of a 64 year old Russian female who presented with acute vision loss followed by sequential ipsilateral CNIII palsy associated with galactorrhea and diabetes insipidus which led to a diagnosis of a metastatic breast cancer to the sellar region.

Results:

Age appropriate workup in a patient with acute vision loss with a lack of a positive review of systems but elevated serum markers for giant cell arteritis led to a delayed diagnosis of a metastasis to the pituitary region which was found only once the tumor extended along the optic nerve to cause CNIII dysfunction. After undergoing endonasal debulking and gamma knife surgery, she is doing well and maintaining vision in the left eye.

Conclusions:

When unable to elicit a review of systems, always look at other factors in patient's presentation: such as polydipsia/polyuria during clinic visits, biopsy of a "benign" breast lesion at outside facility, all may have clues for a systemic diagnosis to explain the unclear ocular findings.

References:

- 1. Chan JW. Acute monocular visual loss in carcinomatous hypertrophic pachymeningitis mimicking giant cell arteritis. *Rheumatol Int.* 2006 May;26(7):683-4.
- 2. Dmuchowska, D.A., Krasnicki, P., Obuchowska, I., Kochanowicz, J., Syta-Krzyzanowska, A., Mariak, Z. Ophthalmic manifestation of skull base metastasis from breast cancer. *Medical Science Monitor.* 2012;18(11).
- 3. Uzum, A.K., Kartal, I., Tamer, G., Mert, M., Colak, N., Aral, F. Pituitary Metastasis of Breast Carcinoma. *Neurosurgery Quarterly.* 2013;23(1).

Keywords: Systemic Disease, Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), YES

Financial Disclosures: The authors had no disclosures.

139 (Submitted)**A Novel Superonasal Transconjunctival Approach To Optic Nerve Sheath Decompression Without The Disinsertion Of Extraocular Muscles**

Kevin E. Lai¹, Kenneth C. Lao², R. Michael Siatkowski¹, Bradley K. Farris¹

¹Dean McGee Eye Institute, Oklahoma City, OK, USA, ²Scott and White Eye Institute, Temple, TX, USA

Introduction:

We wish to introduce a novel superonasal transconjunctival approach to optic nerve sheath decompression (STONEd) that does not require the disinsertion and reinsertion of any extraocular muscles.

Methods:

We will present data from approximately 15 consecutive patients undergoing optic nerve sheath decompression without disinsertion of extraocular muscles at our institution since January 1, 2013.

Results:

We will describe the rate of stability or improvement of visual acuity and visual fields, as well as any postoperative complications, at the 1-week and 6-week post-operative visits. We will compare operative times and intra-operative and post-operative complications with a previously reported retrospective cohort that used surgical techniques requiring disinsertion and reinsertion of extraocular muscles. Video of this technique will be presented.

Conclusions:

To our knowledge, this is the first report of a superonasal transconjunctival approach to optic nerve sheath decompression that does not require the disinsertion of extraocular muscles. STONeD has the potential advantages of reduced intra-operative time, fewer post-operative complications, and a flatter learning curve for fellowship training compared to previously reported techniques. In addition, there are no differences in visual outcome using this procedure compared to other techniques.

Keywords: CSF (Intracranial Hypertension, Intracranial Hypotension, etc), Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), YES

Financial Disclosures: The authors had no disclosures.

140 (Submitted)**Comparing the Classic 2 Repetitions Per Second (2RPS) Visually Evoked Potential (VEP) with a New Metric using the Standard 60-Second Arc Check Size Driven by a Binary M-Sequence in a Group of Multiple Sclerosis Patients and Controls**

Marlen and Mirna Lucero¹, Darrel Conger¹, Amy Conger¹, Shin Beh¹, Benjamin Frohman¹, Laura Balcer², Peter Calabresi³, Elliot Frohman¹, Terea Frohman¹

¹UTSouthwestern Medical Center, Dallas, TX, USA, ²NYU, New York, NY, USA, ³Johns Hopkins, Baltimore, MD, USA

Introduction:

Newer display technologies like liquid-crystal-on-silicon (LCOS) are rapidly supplanting the antiquated CRT methods, mandating a critical analysis of the altered VEP responses. Newer stimulators optimized for multi-focal stimulation currently only generate qualitative results. Additionally these newer stimulators can be tightly calibrated allowing for multi-center studies. Utilizing the binary m-sequence which when applied to a conventional standard checkerboard VEP allows for a new test with shorter testing times, higher signal-to-noise ratios and a single quantitative result. We aim to compare the classic 2 repetitions-per-second (2RPS) visually evoked potential (VEP) with a newer conventional VEP test using the standard 60-second arc check-size generated by a binary m-sequence to test its viability and specificity in a group of multiple sclerosis (MS) patients and controls.

Methods:

Patients with a history of MS and retinal nerve fiber damage, as measured by ocular coherence tomography (OCT), and a control group were tested using two separate methods. All tests were performed using a VERIS system with a FMS3 LCOS stimulator and analyzed using VERIS Science 6.4 software. A standard 10-20-electrode pattern was used. Each subject was tested using a VEP and then m-sequence VEP (m-seqVEP) protocols.

Results:

Ten MS patients with prior optic neuritis were compared with a ten healthy control subjects. The

classic VEP results in both groups were comparable with previously published studies. The m-seqVEP test, however, had a quantifiable result on more subjects and ROC analysis showed a higher degree of specificity.

Conclusions:

Using the m-sequence paradigm, in conjunction with a standard checkerboard framework, may represent a more pragmatic and innovative solution, capable of achieving the necessary utility for reorganizing VEP testing. For clinical and research purposes, the combination of the standard checkerboard with m-sequence stimulator may provide greater sensitivity and specificity in more patients and with greater specificity.

Keywords: Nerve Fiber Layer and Retinal Testing (OCT, ERG Etc), Disorders of Vision Processing, YES

Financial Disclosures: The authors had no disclosures.

142 (Submitted)

Effects of Stimulating Melanopsin-Containing Retinal Ganglion Cells in Migraine Patients using Multifocal Objective Pupillometry

Christian J Lueck^{1,2}, Eman N Ali³, Ted Maddess³, Corinne F Carle³

¹*Department of Neurology, The Canberra Hospital, Canberra, ACT, Australia,* ²*Australian National University Medical School, Canberra, ACT, Australia,* ³*Department of Neuroscience, John Curtin School of Medical Research, Australian National University, Canberra, ACT, Australia*

Introduction:

Recent evidence has linked intrinsically-photosensitive retinal ganglion cells (ipRGCs) to photosensitivity in migraine. It is possible that multifocal pupillographic objective perimetry (mfPOP) might exacerbate migraine and the response to mfPOP might be different in migraineurs. This study was designed to establish the effects of stimulating ipRGCs using mfPOP on migraine severity parameters and pupillary response characteristics

Methods:

A randomized case-control crossover study tested migraineurs and normal controls using mfPOP utilising a blue protocol (BP) to stimulate ipRGCs and a yellow protocol (YP) to stimulate cone photoreceptors. Migraine diaries were obtained a week prior to, and a week after, each testing. Responses were analysed according to response time-to-peak and standardised amplitude (AmpStd). The percentage area under the receiver operator characteristic (%AUC) was used to predict migraine status.

Results:

38 migraineurs (41.97 ± 16.02 years, 23 females) and 24 normal controls (39.17 ± 14.84 years, 14 females) were enrolled. There was no significant difference in the mean number of migraine attacks/subject in the weeks prior to, or following, testing with either protocol. The AmpStds (in dB) were lower for migraineurs than controls: 9.04 ± 11.2 (mean ± SE) vs. 9.48 ± 10.4 for BP, and 10.74 ± 4.96 vs. 11.4 ± 5.23 for YP, though these differences did not reach statistical significance. A migraine attack occurring in the 2 weeks prior to testing had a significant independent effect in lowering AmpStd while a history of triptan use increased AmpStd. Time-to-peak was shorter in YP, probably related to differences in the stimuli. The %AUC was highest for AmpStd (77.2% for YP and 84.6% for BP).

Conclusions:

Stimulating ipRGCs did not affect migraine severity. Pupillary response characteristics were influenced by recent attacks of a migraine and a history of triptan use.

Keywords: Pupil, Miscellaneous, YES

Financial Disclosures: Christian Lueck, Eman Ali: None. Ted Maddess received personal compensation from EyeCo for consulting services and received share options from Seeing Machines Pty. Corinne Carle and Ted Maddess are authors on a patent application that is under license to Seeing Machines Inc., which produces the pupil perimeter used in this research.

143 (Submitted)**Retinal vessel diameter assessment in papilledema by semi-automated analysis of SLO images: feasibility and preliminary results**

Heather E Moss¹, Justin Wanek¹, Gillian Treadwell², Sherryl DeLeon³, Mahnaz Shahidi¹

¹University of Illinois at Chicago Department of Ophthalmology and Visual Sciences, Chicago, IL, USA, ²University of Illinois at Chicago Department of Medicine, Chicago, IL, USA, ³University of Illinois at Chicago College of Medicine, Chicago, IL, USA

Introduction:

Retinal blood vessel diameter changes are promising indirect markers for intra-cerebral and intra-optic nerve mechanical forces in idiopathic intracranial hypertension (IIH) because they are a consequence of compression of the ophthalmic artery and vein caused by the swollen optic nerve and elevated intracranial pressure. We report feasibility and preliminary results of retinal vessel diameter measurements using semi-automated image analysis of scanning laser ophthalmoscopic (SLO) images in eyes with pseudopapilledema and papilledema before and after treatment.

Methods:

A semi-automated customized image analysis software was used to measure the diameters of superior and inferior retinal arteries and veins on 21 SLO images of elevated optic discs. Measurements obtained by two observers were compared with each other and with manual measurements of the same vessels by the same observers. Retinal vein and artery diameters were compared between 6 papilledema and 5 pseudopapilledema subjects. Retinal vein and artery diameter changes were studied for 4 papilledema subjects following treatment for and resolution of papilledema.

Results:

Inter-rater reliability was 0.97 (Pearson correlation, r) for semi-automated measurements and 0.90 for manual measurements. Correlation between manual and semi-automated measurements was 0.85. Vein diameter for papilledema subjects was larger than for pseudopapilledema subjects ($p=0.045$, Mann-Whitney). Papilledema subjects had a decrease in vein diameter following treatment for and resolution of papilledema ($p=0.043$, Wilcoxon signed rank). Artery diameters were neither different between papilledema and pseudopapilledema groups, nor did they change following treatment.

Conclusions:

A novel methodology for semi-automated measurement of retinal artery and vein diameters from SLO images of elevated optic nerves is reliable and feasible. Vein diameters were larger in papilledema subjects before treatment compared with pseudopapilledema subjects. Vein diameters decreased in

papilledema subjects following treatment. Further study is needed to determine the clinical utility of retinal vein diameter measurements as a marker for diagnosis and treatment of papilledema.

References:

- Hayreh SS, Pathogenesis of oedema of the optic disc (papilloedema): A preliminary report, The British Journal of Ophthalmology, 48, 522, 1964.
- Lee SY, Shin DH, Spoor TC, Chaesik K, McCarty B, Kim D, Bilateral retinal venous caliber decrease following unilateral optic nerve sheath decompression, Ophthalmic surgery, 26, 25, 1995.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), CSF (Intracranial Hypertension, Intracranial Hypotension, etc), YES

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145 (Submitted)

Huge anterior cranial fossa Neurenteric Cyst With Unusual Ocular Presenting symptoms : A Case Report

Vannakorn Pruksakorn¹, Supharat Jariyakosol

Department of Ophthalmology, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand

Introduction:

To describe a case of anterior cranial fossa neurenteric cyst in adult patient

Methods:

A 47-year-old healthy Thai male presented with esotropia for 1 year and gradually decreased vision in the past 6 months. He denied ocular pain, nausea, vomiting and headache. His visual acuity was counting finger 2 feet in the right eye and hand motion in the left eye. Eye examinations revealed temporal pallor of both optic discs, decreased corneal sensation and marked limitation of abduction in the left eye. The Krimsky test showed esotropia 90 PD, The findings were otherwise unremarkable. Blood hormonal test results were within normal limit. Orbital MRI depicted a large lobulated cystic mass, 7.1* 8.0* 7.5 cm. involves sellar, suprasellar, anterior cranial fossa region and bilateral small size optic nerves. Mass effect on inferior frontal lobe, medial temporal lobe, pons, midbrain, optic nerves, optic chiasm, optic tract, hypothalamus, as well as cavernous sinuses is observed. Aspiration of the cyst was done by endoscopic transphenoidal approach and incisional biopsy was done at aspirated cyst wall.

Results:

Pathologic examination showed fragments of fibrous cyst wall, lined with non-keratinized stratified squamous epithelium. In occasional areas, ciliated simple columnar epithelium is noted in top of the squamous epithelium, compatible with endodermal cyst.

Conclusions:

Neuroenteric cysts are rare, congenital, benign lesions lined by mucin-secreting columnar epithelium reminiscent of intestinal tract. These cysts tend to be found in the spine, rare in intracranial location. Intracranial lesion typically found in posterior fossa. We report the large anterior fossa neurenteric cyst case presented with bilateral optic atrophy, unilateral fifth (ophthalmic branch) and sixth cranial nerve

palsy without other systemic symptoms. The unusual presentation may cause misdiagnosis and improper initial investigation. Therefore, we should keep in mind of this rare disease with unusual presentation.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Cranial Nerves (Paresis etc), YES

Financial Disclosures: The authors had no disclosures.

146 (Submitted)

Retinal Ganglion Cell Layer Thinning within One Month of Presentation for Non-arteritic Anterior Ischemic Optic Neuropathy and Optic Neuritis

Mark Kupersmith¹, Mona Garvin², Jui-Kai Wang², Mary Durbin³

¹INN/Roosevelt Hospital and NYEEI, New York, NY, USA, ²Department of Electrical and Computer Engineering, The University of Iowa, Iowa City, IA, USA, ³Zeiss-Meditec, Inc, Dublin, CA, USA, ⁴Department of Ophthalmology, Iowa University School of Medicine and Center for Prevention and Treatment of Visual Loss, Veterans Administration Iowa City, IA, USA

Introduction:

High definition optical coherence tomography (HD-OCT) reveals retinal ganglion cell layer (GCL) and retinal nerve fiber layer (RNFL) thinning in chronic optic nerve injury. However, in acute optic nerve injury, the optic disc and axons are often swollen, preventing RNFL thickness evaluation of early neuron loss. Identifying early signs of nerve loss is essential for prompting treatment and testing therapies. We investigated whether GCL thinning occurs before RNFL loss in eyes with acute optic neuropathy, and if early GCL loss correlates with permanent deficits.

Methods:

We prospectively studied eyes with non-arteritic anterior ischemic optic neuropathy (NAION; n=38) and optic neuritis (ON; n=29), acutely and at 1 month, using standard automated perimetry and HD-OCT. Using 2 methods, 3-D layer segmentation (method 1) and by HD-OCT proprietary (method 2), we computed the thickness of macula combined GCL and inner-plexiform-layer (GCL+IPL) and peripapillary RNFL.

Results:

At presentation, the mean macula GCL+IPL values, $80 \mu\text{m} \pm 8.1$ for NAION and $83 \mu\text{m} \pm 8.9$ for ON eyes, did not differ from unaffected fellow eyes ($83 \mu\text{m} \pm 6.4$ and $82 \mu\text{m} \pm 7.0$), using method 1 while method 2 failed in 30% of eyes. At 1month, only 4 NAION eyes and 3 ON eyes had RNFL loss, while 29 NAION eyes and 20 ON eyes had GCL+IPL thinning, with on average losses of $-17.2 \mu\text{m}$ and $-9.0 \mu\text{m}$ ($p=0.001$) respectively. The amount of GCL $\pm$ thinning correlated with the visual field mean deviation ($r=0.61$, $p=0.001$) for NAION eyes.

Conclusions:

Conclusion: GCL thinning develops rapidly, prior to RNFL loss, and appears to be a biomarker of early structural loss in NAION and ON.

Keywords: Nerve Fiber Layer and Retinal Testing (OCT, ERG Etc), Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), YES

Financial Disclosures: Mary Durbin is an employee of Zeiss-Meditec, Inc. Randy Kardon has been a

consultant to Novartis steering committee member for OCTiMS multi center study

150 (Submitted)

Oculomotor Nerve Palsy In A Patient With Pancreatic Neoplasm

Tarek A Shazly¹, Gabrielle R Bonhomme

University of Pittsburgh Medical Center, Pittsburgh, PA, USA

Introduction:

Intraductal Papillary Mucinous Neoplasm (IPMN) are potentially malignant neoplasms of the pancreas. They are managed with surgical resection versus close radiological monitoring in select cases.

Methods:

Case report and review of the literature.

Results:

We present a case of a 59-year-old male with past medical history significant for hypertension, non-insulin dependent diabetes, hyperlipidemia and prior transient ischemic attack who sustained progressive double vision, right ptosis, and severe retrobulbar pain for 3 weeks. Physical examination revealed near total, pupil-involving third nerve palsy with intact 2nd, 4th, 5th and 6th cranial nerve function. CT scan of the head and MRI of the brain obtained at an outside hospital were initially read as normal. The diagnosis of right microvascular 3rd cranial nerve palsy was made, which historically may be monitor expectantly for several months without neuroimaging. Given the patient's conjunctival chemosis and asymmetric IOP, cerebral angiography was conducted to exclude dural arteriovenous malformation, and was negative. Upon careful review of fat-suppressed orbital MRI, an intraconal, avidly enhancing, linear lesion along the inferior division of 3rd nerve as well as the labyrinthine segment of the left facial nerve can be seen. Right maxillary and ethmoidal sinusitis is also evident. Cerebrospinal fluid analysis revealed slightly elevated glucose and protein with normal cytology and negative cultures. CT scanning of his chest abdomen and pelvis ordered to rule out lymphoma revealed a 5 mm hypodensity in the body of the pancreas that likely represent an IPMN. Magnetic Resonance Cholangiopancreatography revealed 2 lobulated cystic lesions measuring 8 x 4 mm and 4 x 3 mm in the body of the pancreas. Periportal and portacaval lymph nodes were also detectable.

Conclusions:

Careful review of orbital imaging may be necessary in seemingly classic microvascular cranial neuropathy. Failure to recognize subtle MRI findings of this patient could have missed his life-threatening pancreatic pathology.

Keywords: Posterior Afferent Visual Pathway (Post-Chiasmal), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

151 (Submitted)

Optic Neuropathy due to Intracranial Dolichoectasia– A Case Series

Jennifer J Lowe¹, William L Hills²

Introduction:

Dolichoectasia, also known as dilatative arteriopathy, comes from the greek words *dolichos* (elongated) and *ectasia* (dilated). Several proposed mechanisms of injury causing cranial neuropathies due to dolichoectasia include: direct compression of the nerve, impaired circulation by direct compression of smaller feeder vessels, obliteration of nutrient vessels derived from the diseased artery, and emboli. Visual pathway compression by internal carotid artery and vertebrobasilar dolichoectasia has been reported. We describe three patients with compressive optic neuropathy due to supraclinoid ICA dolichoectasia, and a fourth patient with transient blood pressure spikes and associated left homonymous hemianopia thought to be due to VBD.

Methods:

Chart review and descriptive case reports of four patients referred to neuro-ophthalmology for visual field abnormalities found to have intracranial dolichoectasia. Commonalities reported and neuro-imaging presented.

Results:

Four patients presented in their 7th-8th decade with a history of hypertension and unexplained visual field abnormalities. Three patients were found to have decreased color vision, as well as unilateral relative afferent papillary defect, increased cup to disc ratio, optic nerve pallor, visual field defect and MR evidence of optic nerve compression by a dolichoectatic supraclinoid internal carotid artery. The third patient reported episodic left homonymous hemianopsia with transiently increased blood pressure found to have right optic tract compression by a dolichoectatic posterior cerebral artery.

Conclusions:

All four cases describe compressive optic neuropathy. Intracranial dolichoectasia can be a cause of optic neuropathy and must be considered when evaluating patients with a history of hypertension and/or atherosclerosis. The use of high resolution MR imaging can play an important role in diagnosis and treatment. We report the only case to our knowledge of blood pressure associated homonymous hemianopsia due to vertebrobasilar dolichoectasia.

References:

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- 5. Campos et al, Oligosymptomatic and giant basilar artery dolichoectasia discovered after a stroke: case report, *Arquivos de Neuro-Psiquiatria*, 65-2, 2007.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Neuro-Imaging (MRI, CT, etc), YES

Financial Disclosures: The authors had no disclosures.

153 (Submitted)

Torsional Nystagmus in Syringobulbia: An association often listed, but rarely described

Gregory E Stein¹, Jeffrey G Odel², Linus D Sun³, Christopher G Filippi⁴

¹*Ophthalmology, New York, NY, USA*, ²*Ophthalmology, New York, NY, USA*, ³*Neurology, New York, NY, USA*, ⁴*Radiology New York, NY, USA*

Introduction:

Syringobulbia is characterized by a syrinx in the brainstem, typically the medulla. Torsional nystagmus is a common sign in syringobulbia [1]. It tops the list of causes of torsional nystagmus in two works on Neuro-ophthalmology, yet the association receives little attention [2,3]. It is characterized by rhythmical slow followed by fast corrective movements about the visual axis [4]. Initially it may be present in extreme lateral gaze, but over time it may present in all directions [1]. Primarily torsional nystagmus is usually due to diseases affecting the central vestibular connections [3] such as brainstem stroke, tumor, vascular malformations, multiple sclerosis and trauma [4]. We report a patient whose oscillopsia from torsional nystagmus led to discovery of syringobulbia.

Methods:

A clinical case presentation includes a video and MRI

Results:

A 50 year-old female treated for herpetic keratitis complained of oscillopsia. She had a car accident 10 years prior with injury to her neck and back. On exam she had torsional nystagmus in all fields of gaze. The torsional nystagmus was characterized by the upper poles of the patient's eyes beating toward her left shoulder. The frequency and amplitude of the nystagmus, as well as the oscillopsia, increased with levoversion. The patient had paresthesias and decreased sensation on the left arm. Imaging revealed a syrinx extending from the T8 level to the cervicomedullary junction and obex. T2 prolongation extended into the posterior aspect of left medulla.

Conclusions:

This case demonstrates torsional nystagmus in syringobulbia. This confirms the finding that torsional nystagmus beats toward the affected side of the medulla [1, 5] and contradicts opposing findings[4]. The patient suffered from hypoesthesia, but ultimately it was nystagmus that led to imaging and the diagnosis of syringobulbia. This case correlates nystagmus with abnormalities in the brainstem and demonstrates how an ophthalmic exam can uncover systemic disease.

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Keywords: Misc Motility Disorders (Nystagmus etc), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

154 (Submitted)

Hemifield Slide Phenomenon As a Result of Heteronymous Hemianopia

Jason H Peragallo¹, Omer Y Bialer¹, Stacy L Pineles², Nancy J Newman^{1,3,4}

¹Emory University/Ophthalmology, Atlanta, GA, USA, ²Jules Stein Eye Institute, University of California Los Angeles/Ophthalmology, Los Angeles, CA, USA, ³Emory University/Neurology, Atlanta, GA, USA, ⁴Emory University/Neurological Surgery Atlanta, GA, USA

Introduction:

Patients with heteronymous hemianopia may experience diplopia due to loss of corresponding retinal points and dissociation of the uniocular visual fields, so-called hemifield slide phenomenon. Concomitant strabismus may complicate hemifield slide. Literature regarding strabismus surgery and restoration of single binocular vision in patients with hemifield slide is limited.

Methods:

In this case series, the medical records of patients with diplopia in the setting of heteronymous hemianopic visual field defects were identified and retrospectively reviewed.

Results:

Three patients were identified. One patient had binasal visual field defects and two patients had bitemporal visual field defects resulting in hemifield slide phenomenon. The two patients with bitemporal defects underwent strabismus surgery for their diplopia and underlying ocular deviation. One patient who underwent strabismus surgery demonstrated a missing strip of central vision or a duplication of central vision dependent on her adjustable suture placement. Two patients, one with binasal defects and one with bitemporal defects, experienced resolution of their diplopia after expansion of their visual fields.

Conclusions:

Hemifield slide phenomenon can occur with any heteronymous hemifield defect. Because of the absence of corresponding retinal points, diplopia may persist despite apparent ocular alignment with strabismus surgery. Expansion of visual field defects may restore corresponding retinal points, allow fusional mechanisms to function, and relieve diplopia. Strabismus surgery to restore binocular visual function can be performed in hemifield slide patients. Patients with improving visual fields may have a better sensory prognosis.

Keywords: Disorders of Vision Processing, Misc Motility Disorders (Nystagmus etc), YES

Financial Disclosures: The authors had no disclosures.

155 (Submitted)

Successful Outcomes in Surgical Treatment of Third Nerve Palsy

Jason H Peragallo¹, Beau B Bruce^{1,2,3}, Amy K Hutchinson¹, Phoebe D Lenhart¹, Valérie Biousse^{1,2}, Nancy J Newman^{1,2,4}, Scott R Lambert¹

¹Emory University/Ophthalmology, Atlanta, GA, USA, ²Emory University/Neurology, Atlanta, GA, USA, ³Emory University, Rollins School of Public Health/Epidemiology, Atlanta, GA, USA, ⁴Emory University/Neurological Surgery Atlanta, GA, USA

Introduction:

Strabismus from third nerve palsy (3NP) is difficult to treat. Our goal was to explore factors associated with successful surgical outcomes in 3NP.

Methods:

Institutional records of all adult patients (≥ 18 years) from 1988-2012 with 3NP who underwent strabismus surgery or botulinum toxin injections were retrospectively reviewed. Success was defined as absence of diplopia, vertical deviation ≤ 2 PD, and horizontal deviation ≤ 10 PD.

Results:

56 patients from four surgeons were included. 30 (53.6%) were female; mean age was 47 (range 20-80). 44 (79%) had unilateral 3NP; 9 (16%) had other ocular motor nerve palsies. 3NP were complete in 24 (43%). Underlying etiology was idiopathic/microvascular in 5 (9%), traumatic in 13 (23%), neoplastic in 12 (21%), aneurysmal in 10 (18%), and other central nervous system related in 16 (29%). Trauma was associated with more frequent aberrant regeneration than other etiologies: 9/13 (69%) vs. 4/43 (9%) ($p < 0.01$). 10 patients (18%) had > 1 surgery. Surgical success was achieved in 28/56 (50%). Success rate was unaffected by etiology, degree of palsy, pupillary involvement, presence of aberrant regeneration, or number of other cranial nerves involved. However, adjustable sutures were used in 27 patients (48%), and there was a trend toward higher success rates when adjustable vs. nonadjustable sutures were used (63% vs. 38%, $p = 0.06$).

Conclusions:

Unlike with 6th nerve palsies, the etiology and degree of 3NP does not appear to affect the ultimate success rates or number of procedures performed. Strabismus surgeries for 3NP with adjustable sutures may be associated with better outcomes.

Keywords: Cranial Nerves (Paresis etc), Misc Motility Disorders (Nystagmus etc), YES

Financial Disclosures: The authors had no disclosures.

156 (Submitted)

Visual Outcomes in Pediatric Optic Neuritis

Michael J Wan¹, Olumuyiwa Adebona¹, Leslie Benson², Mark Gorman², Gena Heidary¹

¹Boston Children's Hospital and Harvard Medical School / Department of Ophthalmology, Boston, MA, USA, ²Boston Children's Hospital and Harvard Medical School / Department of Neurology, Boston, MA, USA

Introduction:

Despite well-established differences between optic neuritis in children and adults, pediatric treatment protocols and visual outcome data is often derived from adult studies. The purpose of this study was to describe clinical features and visual outcomes of a large cohort of pediatric patients with optic neuritis.

Methods:

Retrospective, single-center study of all patients with first-episode optic neuritis and at least 3 months of follow-up over a 10-year period (2002-2012).

Results:

Of the 59 pediatric patients with presented with first episode optic neuritis, 47 had at least 3 months follow-up and 37 had at least 1 year of follow-up. Mean age was 12.3 years old, 74% were female, 45% had bilateral involvement and 47% developed an underlying diagnosis (36% MS, 6% ADEM, 4% NMO). A large majority of the patients (94%) received systemic treatment (85% steroids, 9% multimodal). At 3 months, 64% of the patients were 20/20 or better and 89% were 20/40 or better. At 1 year, 78% of the patients were 20/20 or better and 86% were 20/40 or better. The only factor associated with a poor visual outcome at 1 year (defined as a visual acuity of <20/40) was a visual acuity of less than 20/20 at the 3-month visit ($p = 0.03$). Other factors such as bilateral disease, optic nerve edema, treatment and visual acuity at presentation were not significantly associated with a poor visual outcome at 1 year.

Conclusions:

In this large cohort of pediatric patients with optic neuritis, the majority of patients regained normal vision and visual outcomes overall were quite similar to the Optic Neuritis Treatment Trial (ONTT). Future studies of pediatric optic neuritis should focus on long-term visual outcomes and a randomized trial is needed to evaluate the risks and benefits of treatment.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

157 (Submitted)**Persistent Intracranial Hypertension After Successful Drainage Of Colloid Cyst Of The Third Ventricle In A Pregnant Obese Woman**

Asher Y. Neren¹, Roman Shinder, Valerie I. Elmalem

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

Introduction:

To present a unique case of persistent papilledema in a 27 week pregnant woman subsequent to third ventriculostomy for drainage of a symptomatic colloid cyst.

Methods:

A 25 year-old obese woman, 27 weeks pregnant, presented with left eye visual loss and diminished peripheral vision. Six weeks prior, she saw an outside ophthalmologist for diplopia and was found to have bilateral optic disc edema. MRI Brain and Orbits revealed a colloid cyst of the third ventricle with obstructive hydrocephalus. She underwent urgent neurosurgical endoscopic third ventriculostomy with colloid cyst drainage. One week after surgery, there was persistent CSF egress from her incision site and staples were replaced with water-tight sutures. In the ensuing 2 weeks, she developed

progressive visual loss. On presentation, visual acuity was 20/20 OD and 20/50 OS, with severely constricted visual fields OU. Fundusoscopic examination showed grade IV papilledema OU. MRI/MRA showed improved hydrocephalus, and MRV excluded venous sinus thrombosis. Lumbar puncture opening pressure was 30 cm H₂O with normal CSF contents. On oral acetazolamide, she developed nausea, vomiting, dehydration, and extremity paresthesias.

Results:

Given persistent papilledema with severely constricted visual fields and poor tolerance for conservative measures, left optic nerve sheath fenestration (ONSF) was performed with improved visual function. In her second trimester of pregnancy, she was not an ideal candidate for a CSF shunting procedure.

Conclusions:

This is the first case of persistent papilledema after successful drainage of a colloid cyst of the third ventricle in a young pregnant obese woman. It is proposed that she had both pseudotumor cerebri syndrome and obstructive hydrocephalus from a colloid cyst of the third ventricle. This case reinforced the dilemma of management of papilledema with visual loss in pregnant women.

References:

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Keywords: CSF (Intracranial Hypertension, Intracranial Hypotension, etc), Neuro-Imaging (MRI, CT, etc), YES

Financial Disclosures: The authors had no disclosures.

158 (Submitted)

Correlation of Thickness in Optic Disc Optical Coherence Tomography and Clinical Grading System of Papilledema

Bokkwan Jun¹, Neil R. Miller

Neuro-ophthalmology, Wilmer Eye Institute, Baltimore, MD, USA

Introduction:

To demonstrate the correlation between the nerve fiber layer thickness in optic disc optical coherence tomography (OCT) and clinical grading system of papilledema (modified Frisén scale, MFS)

Methods:

Optic disc OCT for retinal nerve fiber layer (RNFL) thickness and optic disc photographs were

obtained in pseudotumor cerebri (PTC) cases with papilledema documented as clinical grading system. We analyzed the average thickness in each quadrant and in total and demonstrated them with MFS and also calculated Spearman rank correlation of RNFL and MFS.

Results:

The data were obtained from 53 cases of PTC. In grade 0, the average thickness is 85.3 (61-114) μm (temporal/superior/nasal/inferior = 59.7/100.45/63.95/116.05 μm). In grade I, the average thickness is 97.31 (79-127) μm (temporal/superior/nasal/inferior = 68.88/98.38/70.06/148.94 μm). In grade II, the average thickness is 185.25 (136-264) μm (temporal/superior/nasal/inferior = 125.75/195.75/134.75/269 μm). In grade III, the average thickness is 287.33 (259-390) μm (temporal/superior/nasal/inferior = 234.22/304.89/236.67/374.33 μm). In grade IV, the average thickness is greater than 500 μm and in grade V, the average thickness is greater than 600 μm . RNFL thickness and MFS correlates well ($R=0.95$ in overall, 0.8 between grade 0 and I, 0.67 between grade I and II, 0.99 between grade II and III, 0.97 between grade III and IV, 1 between grade IV and V). Among the four quadrants, RNFL thickness in inferior quadrant correlates better ($R=0.93$) than other quadrants ($R=0.78$ in temporal, 0.73 in superior, 0.80 in nasal).

Conclusions:

For higher grades (III, IV, V) of papilledema, MFS itself may be sufficient to evaluate the disc swelling. For lower grades (0, I, II) optic disc OCT may be useful as an adjunct method to confirm and differentiate the severity of disc swelling. Especially the change or difference of RNFL thickness in inferior quadrant may imply progress of disc swelling better than other quadrants.

Keywords: Nerve Fiber Layer and Retinal Testing (OCT, ERG Etc), CSF (Intracranial Hypertension, Intracranial Hypotension, etc), YES

Financial Disclosures: The authors had no disclosures.

159 (Submitted)

Vitreopapillary Traction with Idiopathic Peripapillary Choroidal Neovascularization Mistaken for Optic Disc Swelling in Young Woman

Bokkwan Jun¹, Vivek Patel

Neuro-ophthalmology, Wilmer Eye Institute, Baltimore, MD, USA

Introduction:

To report a rare case of significant vitreopapillary traction with idiopathic peripapillary choroidal neovascularization (CNV) in young woman.

Methods:

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

Results:

A 24-year-old healthy woman presented with sudden decrease of central vision in the right eye and was referred by a local ophthalmologist with unilateral optic disc swelling of the right eye. On examination of the right eye, visual acuity was 20/70 at distance, J10 at near, Humphrey visual field test showed cecocentral scotoma, color perception was 5/10 on Hardy Rand and Rittler and trace relative afferent pupillary defect was observed. MRI brain and orbit with and without gadolinium were unremarkable for demyelination or enhancement. Fundus exam showed vitreopapillary traction and

peripapillary subretinal hemorrhage in the right eye. Optic disc optical coherence tomography showed significant increase of thickness, elevation of disc and vitreous traction and fluorescein angiography demonstrated hyperfluorescent lesion consistent with CNV and focal leakage of optic disc in the right eye.

Conclusions:

We present an uncommon case of vitreopapillary traction with idiopathic peripapillary CNV which was initially mistaken as optic disc swelling in young woman.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Retina, YES

Financial Disclosures: The authors had no disclosures.

160 (Submitted)**Evaluation of Retinal Nerve Fiber Layer in patients with idiopathic Optic Perineuritis using Optical Coherence Tomography**

Soolienah Rhiu¹, Jaeho Jung², Heeyoung Choi²

¹*Dongtan Sacred Heart Hospital, Hallym University College of Medicine, Hwaseong, South Korea,*

²*Pusan national university yangsan hospital, Pusan national university College of Medicine, Busan, South Korea*

Introduction:

The aim of this study is to assess the effect of optic perineuritis (OPN) on retinal nerve fiber layer (RNFL), and the ability of optical coherence tomography(OCT) to evaluate the retinal nerve fiber loss after idiopathic optic perineuritis.

Methods:

The diagnosis of idiopathic OPN was made in patients who had an acute optic neuropathy and typical optic nerve sheath enhancement in fat suppression and contrast enhancement orbital magnetic resonance imaging (MRI), normal laboratory findings. Subjects were underwent by Cirrus Spectral Domain-OCT (Carl Zeiss Meditec, Inc., Dublin, CA) and the OCT parameters, optic nerve head and RNFL thickness, was calculated automatically by the equipment's software at initial visit and 12 months follow-up.

Results:

4 patients were studied in this study. All patient showed that RNFL, especially temporal sector RNFL (Papillomacular bundle), was significant thinner in affected eye when compared to normal value and the other sound eye at 12 months after acute OPN.

Conclusions:

Our study suggests that retinal nerve fiber loss was observed in idiopathic optic nerve sheath inflammation and OCT was good technique for axonal loss and disease severity indicator in OPN

Keywords: Orbital and Eyelid Disorders, Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), YES

Financial Disclosures: The authors had no disclosures.

161 (Submitted)

The Impact of Vitamin D Status on Recovery from Optic Neuritis

Fiona Costello^{1, 3, 4}, Jodie Burton^{1, 2, 4}, Jessie Trufyn^{1, 4}

¹University of Calgary, Department of Clinical Neurosciences, Calgary, AB, Canada, ²University of Calgary, Department of Community Health, Calgary, AB, Canada, ³University of Calgary, Department of Surgery, Calgary, AB, Canada, ⁴Hotchkiss Brain Institute Calgary, AB, Canada

Introduction:

Optic neuritis (ON) is a common manifestation of multiple sclerosis (MS), and serves as a model to study the effects of inflammatory and degenerative activity in this central nervous system (CNS) disorder. Vitamin D insufficiency is a risk factor for MS and ameliorates inflammation. The objective of this study is to study the relationship between vitamin D status and ON recovery, by using optical coherence tomography (OCT) measures of retinal integrity. The hypothesis is that vitamin D sufficiency (25(OH)D > 80 nmol/L) is associated with better OCT outcomes. Primary outcomes include retinal nerve fiber layer (RNFL) thickness, inter-eye differences (IED) in RNFL thickness and ganglion cell layer (GCL) thickness at 6 months and at baseline between vitamin D sufficient and insufficient groups.

Methods:

In this prospective cohort study, patients with acute ON are undergoing OCT, full field visual evoked potential (VEP), and ophthalmic testing at baseline, 3-months, 6-months, and 12-months. Serum 25(OH)D testing is performed at baseline and 6-months.

Results:

To date, 49/50 patients have been enrolled (36 females). Sixty-eight percent of patients have been vitamin D insufficient at baseline, which has been associated with more optic disc edema and increased mean RNFL thickness (131 vs 106 μ m, $p=0.14$), macular volume (10.2 vs 9.8 mm³, $p=0.04$) and IED in RNFL in acute ON. At month 6, IED in GCL thickness has been greater in vitamin D insufficient patients (13 vs 8 μ m). Regardless of baseline RNFL thickness or vitamin D level, men have had significantly lower 6 month RNFL values (70 vs 81 μ m, $p=0.03$) and greater IED in RNFL and GCL measures (20 vs 8 μ m for both, $p=0.012$ and $p=0.008$ respectively) versus women.

Conclusions:

Vitamin D insufficiency is associated with worse optic disc edema in acute ON. Male gender is an independent risk factor for worse recovery at 6-months. Ganglion layer measures at 6-months in sufficient patients suggests a possible neuroprotective role for vitamin D.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Nerve Fiber Layer and Retinal Testing (OCT, ERG Etc), YES

Financial Disclosures: The authors had no disclosures.

162 (Submitted)

Validity and Acceptance of Color Vision Testing on Smartphones

Omar Ozgur¹, Mark Vieyra², Trisha Emborgo³, Rebecca Farmer Huselid³, Rudrani Banik¹

¹New York Eye and Ear Infirmary, New York, NY, USA, ²Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel, ³Hunter College, The City University of New York, New York, NY, USA

Introduction:

Color vision testing (CVT) is an important part of the neuro-ophthalmic examination. The validity of CVT may be affected by the quality of the displayed image, background illuminance, image distance, and visual acuity. iPhone® and Android® smartphones provide diagnostic testing applications including CVT. There is minimal evidence to support the validity of these non-standardized smartphone applications. The purpose is to assess the validity of smartphone CVT by comparing results using the Eye Handbook (EHB) CVT application with Ishihara color plates (ICP).

Methods:

An IRB-approved prospective, randomized study was performed of 193 patients with near visual acuity of $\geq 20/60$ at 14 inches. The study group included patients with ocular pathology. The control group included patients with no known pathology. CVT was performed with both ICP and EHB under a standardized background illuminance, randomized by order of testing and phone model. OD was the study eye. The testing was scored by number of correct plates out of eleven test plates for each modality. A paired-samples t-test was performed.

Results:

In the control group (n=80), mean score for ICP was 10.91 $\pm$ 0.284 correct plates and for EHB was 10.94 $\pm$ 0.291, with a difference of 0.0250 $\pm$ 0.0274, $p = 0.4176$, SD = 0.274, 95% CI -0.036 to 0.086. In the study group (n=113), mean score on ICP was 10.08 $\pm$ 2.109 correct plates and for EHB was 10.29 $\pm$ 2.073, with a difference of 0.212 $\pm$ 0.784, $p = 0.0048$, SD = 0.784, 95% CI 0.066 to 0.359. Using ICP as the "gold standard" for CVT, the sensitivity of EHB was 89% and specificity 99%. For all subjects, 60% preferred EHB, 12% preferred ICP, and 29% had no preference.

Conclusions:

In patients with ocular pathology, there was a statistically significant difference in CVT results comparing EHB with ICP. In control patients, this difference was not statistically significant. The majority of subjects preferred EHB to ICP testing.

References:

- Tools:

- Eye Handbook Color Plate application (Copyright © 2009 Cloud Nine Development LLC).
<http://www.eyehandbook.com/>

- Articles:

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Keywords: Disorders of Vision Processing, Miscellaneous, YES

Financial Disclosures: The authors had no disclosures.

163 (Submitted)

Using Resting State Functional Magnetic Resonance Imaging to Track Recovery After Optic Neuritis

Fiona Costello^{3,4,5}, Fil Cortese¹, Bradley Goodyear^{1, 2, 3, 5}, Jessie Trufyn³

¹University of Calgary, Department of Computers and Electrical Engineering, Calgary, AB, Canada,

²University of Calgary, Department of Radiology, Calgary, AB, Canada, ³University of Calgary, Department of Clinical Neurosciences, Calgary, AB, Canada, ⁴University of Calgary, Department of Surgery Calgary, AB, Canada, ⁵Hotchkiss Brain Institute Calgary, AB, Canada

Introduction:

Optic neuritis (ON) is a common cause of vision loss in MS. Resting-state functional magnetic resonance imaging (fMRI) functional connectivity detects temporal correlations in spontaneous blood oxygen level-dependent (BOLD) signal oscillations. This study investigates how ON affects the functional connectivity of brain regions with primary visual cortex (V1).

Methods:

20 patients (mean age = 34; 2 males) with ON as a clinically isolated syndrome (CIS) or associated with MS and 12 healthy controls underwent 3 imaging sessions at baseline, 6-months and 12-months. Image acquisition was conducted using a 3T MR scanner and consisted of anatomical images for co-registration of fMRI data; task-related fMRI scan to localize primary visual cortex (V1); and two resting-state fMRI scans.

Results:

At baseline, controls have stronger functional connections between V1 and areas involved in processing vision, touch (somatosensory association cortex), movement (premotor cortex), sound (primary auditory cortex) and awareness (dorsal posterior cingulate cortex) than ON patients. Patients show increased functional connectivity between V1 and frontal and anterior temporal cortices 12 months post-ON with eyes open. With eyes open and eyes closed, MS patients show a unique pattern of functional connectivity as compared with CIS patients. Both controls and MS patients show an increase in functional connectivity between premotor, dorsolateral prefrontal, posterior cingulate cortices and V1 compared to CIS patients. In contrast, controls and CIS patients show an increase in functional connectivity between anterior prefrontal, anterior cingulate, secondary visual areas and V1 as compared to MS patients.

Conclusions:

Functional connections between V1 and other brain areas are altered in the presence of ON, specifically in areas involved with complex visual processing. An increase in functional connectivity 12 months after ON suggests compensatory mechanisms or cortical plasticity may be involved. ON patients with MS can be distinguished from CIS patients by comparing the resting-state functional connectivity between V1 and secondary visual areas.

Keywords: Neuro-Imaging (MRI, CT, etc), Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), YES

Financial Disclosures: The authors had no disclosures.

164 (Submitted)

Brainstem Pathways of the Subjective Visual Vertical Tilt: Analysis of the Topology of Brainstem Lesions

Sun-Young Oh^{1,2}, Tae-Ho Yang¹, Ji-Yun Park³

¹*Department of Neurology, Chonbuk National University College of Medicine, Jeonju, Korea,*

²*Research Institute of Clinical Medicine, Chonbuk National University, Biomedical Research Institute of Chonbuk National University Hospital, Jeonju, Korea,* ³*Department of Neurology, Presbyterian Medical Center, Jeonju, Korea*

Introduction:

The eye-head coordination in roll plane is driven by crossed graviceptive projections running from otoliths and semicircular canals via the medial longitudinal fasciculus (MLF) to the contralateral interstitial nucleus of Cajal (INC). Unilateral damage of these fibers regularly causes the ocular tilt reaction (OTR) which consist of ocular torsion, skew deviation and head tilt, and the tilt of subjective visual vertical (SVV). The aim of this study was to analyze the topology of ischemic brainstem lesions generating pathological tilt of the SVV.

Methods:

We examined 82 patients with isolated acute unilateral brainstem infarction for SVV tilt of otolith-ocular dysfunction. We analyzed by means of MRI-based voxel-wise lesion-behavior mapping, and constructed the probabilistic lesion maps.

Results:

Fifty percent of patients with acute brainstem lesion showed abnormal tilts of SVV, of which 76% patients showed ipsilesional tilt and 24% were contralesional. Patients with abnormal SVV tilt toward contralesional side involved the areas including medial vestibular nuclei in the rostral medulla, medial longitudinal fasciculus (MLF), central ventral tegmental tract (CVTT), superior cerebellar peduncle, and rostral interstitial medial longitudinal fasciculus (riMLF), and interstitial nucleus of Cajal (INC). And, patients with SVV tilt to ipsilesional have lesions at the medial and inferior vestibular nuclei at the caudal medulla, inferior cerebellar peduncle, and ipsiversive vestibule-thalamic tract (IVTT).

Conclusions:

In this study, we found that brainstem lesions can cause ipsi- or contralesional SVV tilt depending on which tracts are involved, and it may be normal in cases with sparing the ascending vestibular tracts.

Keywords: Misc Motility Disorders (Nystagmus etc), Neuro-Imaging (MRI, CT, etc), YES

Financial Disclosures: The authors had no disclosures.

165 (Submitted)

Homonymous visual field defect showing negative MRI: Crossed Cerebellar Diaschisis

Ji-Yun Park², Sun-Young Oh¹, Tae-Ho Yang¹

¹*Department of Neurology, Chonbuk National University Medical School and Hospital, Jeonju, Korea,*

²*Department of Neurology, Presbyterian Medical Center, Jeonju, Korea*

Introduction:

Homonymous hemianopia (HH) arises from a lesion interrupting the retrochiasmal visual pathways anywhere from the optic tract to the visual cortex. These lesions can usually be seen by structural imaging studies. Crossed cerebellar diaschisis (CCD) is a cerebellar suppression of the cerebral

blood flow due to contralateral supratentorial lesions. CCD has usually been reported in patients with frontal or parietal lobe lesions, but rarely reported in the occipital visual cortex.

Methods:

We describe a 35-year-old man suffering from homonymous hemianopia after head trauma with negative magnetic resonance imaging (MRI) findings.

Results:

We report a young man who presented with homonymous hemianopia and mild ataxia after head trauma but without lesions on brain magnetic resonance imaging (MRI). Diffuse tensor tractography showed a symmetric optic tract and radiation. However, brain 18FDG-PET (fludeoxyglucose-positron emission tomography) revealed crossed hypometabolism of the occipital cortex and cerebellum, suggestive of CCD.

Conclusions:

These findings suggest that CCD can cause HH after brain injury and that HH associated with CCD is more related to cortical hypometabolism of the visual cortex rather than prominent structural lesions or fiber tract dysfunction on the visual pathway.

Keywords: Posterior Afferent Visual Pathway (Post-Chiasmal), Neuro-Imaging (MRI, CT, etc), YES

Financial Disclosures: The authors had no disclosures.

166 (Submitted)**Strabismus Caused by Infantile Orbital Myofibromatosis Originating From An Extraocular Muscle**

Neda Nikpoor¹, Craig McKeown, Kara M. Cavuoto

Bascom Palmer Eye Institute, Miami, FL, USA

Introduction:

Myofibromatosis is the most common fibrous tumor of infancy. Of the two forms, solitary and multicentric, the solitary form is the more common. Skin, bone, viscera, and soft tissue involvement is typical, but lesions rarely involve the orbit. Spontaneous regression can occur within two years and recurrence is rare in multicentric disease.

Methods:

We present an atypical case of a child with orbital myofibroma resulting in strabismus requiring surgical intervention despite spontaneous regression.

Results:

A 2-year-old girl with systemic myofibromatosis and a history of excision of multiple non-ocular myofibromas presented with right upper lid ptosis, 20 prism diopters (Δ) of right hypertropia, and unilateral downgaze restriction of the right eye. Echography and magnetic resonance imaging confirmed a lesion of the right superior/levator complex, with characteristics favoring a diagnosis of right orbital myofibroma. Serial echography demonstrated spontaneous regression of the lesion with normalization of the involved extraocular muscles. However, the right hypertropia persisted and increased in magnitude to 35 Δ . The patient subsequently underwent right superior rectus recession with no residual hypertropia in primary gaze one year after surgery.

Conclusions:

Notably, this patient had a remnant motility deficit requiring strabismus surgery despite the spontaneous regression of the myofibroma. To our knowledge, there are no reported cases of spontaneously regressing orbital myofibromas in a person with systemic myofibromatosis. Of the cases of orbital myofibroma, most originated from the inferior orbit whereas our patient's lesion originated from the superior orbit. Furthermore, we were unable to find any other published cases of an infantile myofibroma originating from the extraocular muscles.

Keywords: Misc Motility Disorders (Nystagmus etc), Orbital and Eyelid Disorders, YES

Financial Disclosures: The authors had no disclosures.

169 (Submitted)**Compressive Optic Neuropathy Secondary To Sphenoid Sinus Aspergillosis**

Xiu Ling Tan¹, Kong Yong Goh^{1,2}

¹National Healthcare Group Eye Institute, Tan Tock Seng Hospital, Department of Ophthalmology, Singapore, Singapore, ²Mount Elizabeth Novena Specialist Centre, Dr GOH Eye Clinic, Singapore, Singapore

Introduction:

Compressive optic neuropathy as an initial presentation of fungal sinusitis is a rare but important occurrence. We report a case of compressive optic neuropathy in an immunocompetent patient secondary to sphenoid sinus aspergillosis and an ONODI cell.

Methods:

Clinical presentation, radiological and histopathological data are reviewed and discussed.

Results:

A 63-year-old Chinese man presented with acute painless blurring of vision of his right eye. Significant past medical history included diabetes mellitus. He had bitemporal headache but was otherwise constitutionally well. Best-corrected visual acuity was 20/400 and 20/20 in his right and left eyes respectively. A right relative afferent pupillary defect was present, associated with reduced colour vision and diminished light perception and red desaturation. Fundal examination revealed a right temporal optic disc pallor. There were no signs of orbital apex or cavernous sinus syndromes. Computed tomography of the brain and orbits revealed an enhancing irregular soft tissue mass in the right sphenoid sinus, associated with bony erosion of the roof and posterior superior orbit wall. Magnetic resonance imaging demonstrated that the right optic nerve was inseparable from the soft tissue mass at the orbital apex and canal. The patient underwent emergency endoscopic bilateral sphenoidotomy. The right optic nerve was seen within an ONODI cell. The right sphenoid sinus mucosa was thickened with yellow green pus seen. A fungal ball was removed. Fungal cultures grew *Aspergillus fumigatus*. Histopathology of the biopsied mucosa confirmed invasive sinus aspergillosis. Postoperatively, the right eye vision improved to 20/40, with resolution of the headache. The patient completed a course of systemic voriconazole and methylprednisolone.

Conclusions:

Fungal sphenoid sinusitis is difficult to diagnose and treat due to its nonspecific symptoms. Hence ophthalmologists should have a high index of suspicion. An early diagnosis with timely aggressive surgical resection and appropriate anti fungal treatment is important to prevent permanent visual loss.

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Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Neuro-Imaging (MRI, CT, etc), YES

Financial Disclosures: The authors had no disclosures.

170 (Submitted)

A Unifying Neurological Mechanism for Infantile Nystagmus

Michael C Brodsky¹, Lou F Dell'Osso¹

¹*Departments of Ophthalmology and Neurology, Mayo Clinic , Rochester, MN, USA, ²Daroff-Dell'Osso Ocular Motility Laboratory and Department of Neurology, Case Western University, Cleveland, OH, USA*

Introduction:

Why do infants with congenitally poor vision in both eyes develop infantile nystagmus? And what neuroanatomical pathways mediate this binocular oscillation? Since infantile nystagmus usually begins between 2 and 3 months of age when subcortical optokinetic systems normally cease to function, could persistent activation of this subcortical optokinetic system lead to infantile nystagmus?

Methods:

Investigation into the evolutionary underpinnings of optokinetic nystagmus to test the hypothesis that delayed or deranged development of high contrast foveal acuity permits subcortical optokinetic pathways , which are normally operative in the first two months of human life, to generate an antagonistic signal during attempted fixation or pursuit movements.

Results:

Lateral-eyed afoveate animals utilize the subcortical accessory optic system to generate accurate responses to full-field optokinetic input. Humans have developed a cortical foveal pursuit system that suppresses the perception of this full-field optokinetic motion during active pursuit. When humans rotate their eyes to pursue a moving target, however, the visual world sweeps across their retinas to create a contraversive optokinetic stimulus. When foveal vision is slow to develop in infancy, this phylogenetically old optokinetic system continues to be ontogenetically expressed.

Conclusions:

Infantile nystagmus signals a breakdown in the armistice between subcortical and cortical optokinetic pathways during early visual development. Neuroanatomically, infantile nystagmus arises from a discordance between foveal smooth pursuit modulated by the visual cortex and the afoveate full-field optokinetic system modulated by the accessory optic system and its central cerebellar connections. Delayed or deranged development of central foveal vision unleashes the fundamental antagonism between subcortical optokinetic pathways and cortical pursuit, causing infantile nystagmus to be

expressed. Because the accessory optic system functions as a subcortical pursuit system in afoveate animals, its reactivation in humans explains the paradox that infantile nystagmus is inherent to the pursuit system, yet pursuit gain remains normal.

Keywords: Misc Motility Disorders (Nystagmus etc), Disorders of Vision Processing, YES

Financial Disclosures: The authors had no disclosures.

171 (Submitted)

Reproducibility of Circumpapillary Retinal Nerve Fiber Layer Measurements Using Hand-held Optical Coherence Tomography in Sedated Children with Optic Pathway Gliomas

Robert A. Avery¹, Avital Cnaan¹, Joel S. Schuman², Chieh-Li Chen², Natalie C. Glau¹, Roger J. Packer¹, Graham E. Quinn³, Hiroshi Ishikawa²

¹Children's National Medical Center, Washington, DC, USA, ²University of Pittsburgh Medical Center, Pittsburgh, PA, USA, ³Children's Hospital of Philadelphia, Philadelphia, PA, USA

Introduction:

Hand-held optical coherence tomography (HH-OCT) measures of circumpapillary retinal nerve fiber layer (cpRNFL) thickness have a close relationship to vision loss (visual acuity and or visual field) in children with optic pathway gliomas (OPGs). The intravisit reproducibility of cpRNFL measurements using HH-OCT in sedated children was investigated.

Methods:

HH-OCT (Bioptigen) measures of cpRNFL were acquired in children with and without OPGs undergoing sedation for a clinically indicated MRI. A 6 x 6 mm volume (isotropic 300x300 or non-isotropic 1000x100 samplings) was acquired over the optic nerve. Children with two or more acceptable scans (same sampling volume and eye) acquired during a single imaging session were included. Subjects could contribute scans from one or both eyes and sampling types. Automated software segmented the volume and determined the cpRNFL thickness (3.4 mm circle centered on the geometric center of the optic nerve head, which was manually delineated). The coefficient of variation (CV) for the global average and anatomic quadrant cpRNFL thickness was calculated.

Results:

375 eligible HH-OCT scans (N=270 isotropic volumes; N=105 non-isotropic volumes) from 101 subject eyes (33 unique subjects) were analyzed (age range 1.75-8.9 years). Seventy-seven subject eyes had OPGs, of which 17 had experienced vision loss. The isotropic volumes (median 4 scans per eye) demonstrated the best CV for the global average (4.3%), followed by the superior (5.5%), inferior (6.6%), temporal (8.3%) and nasal (8.3%) quadrants. For non-isotropic images (median 2 scans per eye), the global average (2.9%) demonstrated the best CV, followed by inferior (4.7%), superior (5.0%), nasal (5.6%), and temporal (6.2%) quadrants. Subject eyes with vision loss had an increased CV ($p < 0.01$) compared to those with normal vision.

Conclusions:

cpRNFL measures acquired with HH-OCT during sedation demonstrate good reproducibility. HH-OCT has the potential to monitor progressive optic neuropathies in young children who have difficulty cooperating with traditional OCT devices.

Keywords: Nerve Fiber Layer and Retinal Testing (OCT, ERG Etc), Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), YES

Financial Disclosures: The authors had no disclosures.

172 (Submitted)

Congenital Ocular Motor Apraxia with Wheel-Rolling Ocular Torsion-A Diagnostic Sign of Joubert

Michael C Brodsky⁴, Paraskevi-Eleni Papanagnu¹, Lindsay L Klaehn², Genie M Bang³

¹Mayo Clinic Department of Ophthalmology , Rochester, MN, USA, ²Mayo Clinic Department of Ophthalmology , Rochester , MN, USA, ³University of Illinois Department of Ophthalmology , Chicago, IL, USA, ⁴Mayo Clinic Department of Ophthalmology Rochester, MN, USA

Introduction:

Joubert syndrome is a multisystem disorder that is associated with a constellation of cyclic ocular motor disturbances.^{1,2} We describe the association of congenital ocular motor apraxia with tonic alternating torsional deviations of the eyes as a diagnostic sign of Joubert syndrome.

Methods:

Retrospective review of clinical, neuroimaging, video-oculography and genetic studies in two children with congenital ocular motor apraxia and Joubert syndrome

Results:

In both children, retinal examination showed a tonic alternating cyclodeviation with a periodicity of 10 to 15 seconds. Heidelberg infrared videoimaging documented tonic conjugate wheel-rolling torsional rotations of both eyes (ranging in amplitude from 30 to 45 degrees in each direction). Video-oculography showed a periodic alternating skew deviation. Magnetic resonance imaging showed a pathognomonic "molar tooth" sign with hypoplasia of the cerebellar vermis and elongated non-decussating superior cerebellar peduncles. Genetic testing confirmed sequence variants in the *CC2D2A* gene in our first patient, and in the *C5orf42* gene in our second patient.

Conclusions:

In patients with congenital ocular motor apraxia, the unique finding of tonic alternating ocular torsional deviations on retinal examination establishes the diagnosis of Joubert syndrome. It is unclear whether the tonic wheel-rolling torsional eye movements resulted from cerebellar malformations of Joubert syndrome or from the non-decussation of brainstem pathways that are known to accompany this condition. This finding does not appear to be mutation specific.

References:

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- 2. Couch SM, Brodie SE, Leavitt JA, Brodsky MC. Something to sink your teeth into. Survey of Ophthalmology 2011;56:544-9.

Keywords: Misc Motility Disorders (Nystagmus etc), Disorders of Vision Processing, YES

Financial Disclosures: The authors had no disclosures.

174 (Submitted)

Localizing Value of Ipsilateral Horner's Syndrome and Fourth Nerve Palsy.

Dolores Riberi Ayerza¹, Roberto Ebner

British Hospital, Buenos Aires, Argentina

Introduction:

To describe the association of ipsilateral Horner's Syndrome and IVth nerve palsy in a patient with a right cavernous sinus tumor.

Methods:

Single case report

Results:

A 54-year-old female presented with vertical diplopia and neck pain. Mild ptosis and miosis were observed in the right eye. Her examination was normal except for 2mm blepharoptosis and miosis OD with anisocoria greater in the dark. An ice-pack-test was negative. Topical Apraclonidine 0.5% test demonstrated midriasis and lid retraction OD after 20 minutes confirming the presence of Horner's syndrome. Parks three-step-test was suggestive of right superior oblique muscle paresis with right hypertropia of 6 prism diopters most consistent with IVth nerve paresis. MRI of the brain and orbits with and without contrast revealed a mass in the right cavernous sinus most consistent with meningioma.

Conclusions:

The presence of contralateral IVth nerve palsy with ipsilateral Horner's syndrome has been previously documented in ventral midbrain lesions. However, our case suggests that clinicians should be aware that cavernous sinus lesions may also lead to a similar clinical presentation when ipsilateral IV th nerve palsy and Horner's Syndrome are present.

Keywords: Pupil, Cranial Nerves (Paresis etc), YES

Financial Disclosures: The authors had no disclosures.

175 (Submitted)

Sixth Nerve Palsy + Ipsilateral Horner's Syndrome = Parkinson's Syndrome.

Dolores Riberi Ayerza¹, Roberto Ebner

British Hospital, Buenos Aires, Argentina

Introduction:

We present five patients with VI th nerve palsy and ipsilateral Horner's Syndrome (HS), an association orienting diagnosis towards the cavernous sinus.

Methods:

Consecutive case series of five patients with horizontal diplopia due to VIth nerve paresis and HS with ipsilateral cavernous sinus lesion.

Results:

All 5 patients had abducens palsy with horizontal diplopia in primary position (3 patients) or evident in lateral gaze only (2 patients) and ipsilateral HS. Apraclonidine 0.5% evidenced sympathetic denervation in all patients in this series. All 5 cases had neuroimages (MRI in 3 cases, Computerized Tomography-CT in one case and Cerebral Angiography-CA in one case) demonstrating cavernous sinus lesions; 2 meningiomas, 1 carotid-cavernous aneurism, 1 foreign body (bullet) and 1 squamous cell carcinoma.

Conclusions:

The association of VI th nerve palsy with ipsilateral HS was described by D. Parkinson and named after him (PS). It has a great localizing value being the site of the lesion; the cavernous sinus. The use of Apraclonidine 0.5% has great diagnostic value for HS but does not provide information about the level of the sympathetic pathway lesion. In our series helped to demonstrate the presence of sympathetic denervation. The isolated VI th nerve palsy has no localizing value and neither for the HS alone. The association of ipsilateral HS and abducens palsy (PS) in our series was accompanied by neuroimages (MRI, CT and CA) showing cavernous sinus lesions of different origin in all cases. Neuroimaging of the cavernous sinus is recommended in the evaluation of Parkinson's Syndrome, selecting the adequate image according to the nature of the suspected causing factor.

Keywords: Pupil, Cranial Nerves (Paresis etc), YES

Financial Disclosures: The authors had no disclosures.

176 (Submitted)**A Longitudinal Analysis of Stroke Risk Following Non-Arteritic Ischemic Optic Neuropathy**

Hilary M Grabe¹, Lindsey B DeLott^{1,2}, Taylor Blachley¹, Wayne T Cornblath^{1,2}, Joshua D Stein¹

¹University of Michigan / Department of Ophthalmology and Visual Sciences, Ann Arbor, MI, USA,

²University of Michigan / Department of Neurology, Ann Arbor, MI, USA

Introduction:

Non-arteritic ischemic optic neuropathy (NAION) is the most common acute optic neuropathy in patients > 50 years old.¹ The association between NAION and stroke is uncertain. Several small studies have shown no increase in stroke following NAION,^{2,3} while others have shown an increased stroke risk following NAION.^{4,5} Our goal was to determine if patients with NAION are at an increased long term risk of ischemic stroke and if that risk is independent of other known stroke risk factors.

Methods:

We used claims data from the OptumInsight Clinformatics Data Mart which contains records of all beneficiaries in a large managed care network in the United States who had some form of eyecare from 2001-2012. Inclusion criteria were ≥ 50 years old with continuous enrollment in the plan for ≥ 2 years. Incident NAION and ischemic stroke were identified by International Classification of Diseases-9 (ICD-9) codes. We excluded those with pre-existing stroke or NAION during their first 2 years in the plan, giant cell arteritis or optic neuritis. Unadjusted and adjusted Cox proportional hazards modeling were used to determine whether the development of NAION affected the risk of subsequent development of ischemic stroke. Models were adjusted for sociodemographic factors, medical, and ocular comorbidities.

Results:

Among the 1,930,194 enrollees who met the inclusion criteria, 2531 developed NAION (0.1%) and

50,699 (2.6%) experienced an incident ischemic stroke. After adjustment for covariates, individuals with NAION had a 162% increased hazard of developing ischemic stroke (adjusted HR 2.62, [95% CI 2.12-3.24]) relative to those who did not have a NAION.

Conclusions:

After accounting for comorbid medical conditions and other confounding factors, patients in our study with NAION had an increased risk of ischemic stroke. We recommend aggressive risk factor modification and counseling about signs and symptoms for ischemic stroke in patients with NAION.

References:

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Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

177 (Submitted)

Pseudoisochromatic plate testing: are we measuring color vision or something else?

Jiawei Zhao¹, Sarita Dave¹, Jiangxia Wang^{1,2}, Prem S. Subramanian¹

¹*Johns Hopkins University School of Medicine, Baltimore , MD, USA*, ²*Johns Hopkins Bloomberg School of Public Health , Baltimore , MD, USA*

Introduction:

Color vision testing with pseudoisochromatic (PIP) plates [Hardy-Rand-Ritter (HRR), Ishihara] is a typical part of the neuro-ophthalmologic examination, and abnormal responses are often used as evidence of optic neuropathy. However, color disc arrangement tests such as the Farnsworth D-15 are the gold standard for detecting and classifying color vision deficits. The aim of this study was to determine if apparent color vision deficits on HRR and Ishihara plate testing represent truly aberrant color vision or indicate a decrement of other psychovisual parameters.

Methods:

Forty-seven subjects (age 18-65) were recruited prospectively from an outpatient clinic. The study group included patients diagnosed with optic neuropathy (n=23), retinal dystrophy (n=3) and dry eye syndrome (n=1), and controls (n=20). Individuals with Va<20/200 or with congenital color blindness

were excluded. All subjects underwent a comprehensive eye examination including visual acuity, color-vision and contrast sensitivity testing. Color vision was assessed using HRR and Ishihara PIP plates and Farnsworth D-15 hue discrimination test. Contrast sensitivity was measured using Pelli-Robson contrast sensitivity charts.

Results:

HRR score and contrast sensitivity (CS) were correlated (Kendall's correlation=0.31, $p<0.001$), with no relationship between HRR score and Farnsworth D-15 score (Kendall's correlation=0.06, $p=0.250$). On multivariate analysis, CS ($\beta=8.36$, $p<0.001$) and visual acuity ($\beta=1.92$, $p=0.025$) both showed association with HRR scores. Ishihara score weakly correlated with CS (Kendall's correlation=0.16, $p=0.016$) but did not correlate with Farnsworth D-15 score (Kendall's correlation=0.04, $p=0.363$). HRR score had a stronger relationship with CS than did the Ishihara score ($p=0.014$).

Conclusions:

Both Ishihara and HRR PIP testing appear to measure contrast sensitivity in a visual acuity-dependent manner; they do not accurately assess color vision. HRR is a more sensitive measure of contrast sensitivity deficits than is the Ishihara PIP set.

Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Miscellaneous, YES

Financial Disclosures: The authors had no disclosures.

178 (Submitted)**Hiding In Plain Sight: Uncommon Presentations Of Digoxin Ocular Toxicity**

Dov B. Sebro¹, Jeffrey G. Odel, Lora R. Dagi Glass, Stephen H. Tsang

Edward S. Harkness Eye Institute, Columbia University Medical Center/Department of Ophthalmology, New York, NY, USA

Introduction:

Visual symptoms of digoxin toxicity include photopsias, frosted vision, chromatopsias, and vision loss, which can occur even at therapeutic levels. Full field electroretinogram (ERG) easily supports the diagnosis and should be used even in atypical presentations.

Methods:

Case reports of two patients with digoxin-related visual loss, in which the diagnosis was not considered by their physicians because of variations in clinical presentation.

Results:

Case 1: An 81 YO male with macular degeneration OU, on digoxin for congestive heart failure, complained of difficulty reading for 2 months. His retinologist, noting no change in his exam, referred him for outside neuro-ophthalmology consultation that attributed the symptoms to ARMD. He was seen in second opinion and his vision was 20/100 OU with Humphrey visual fields 24-2 showing generalized depression OU. ERG showed profound delays in cone and rod implicit times and decreased amplitudes. Serum digoxin level was 2.2 (therapeutic range 0.5-2.0). 2 weeks after stopping digoxin, the patient reported he could read again. ERG showed improved responses. Case 2: A 74 YO female on digoxin for atrial fibrillation presented with recurrent periods of decreased vision for several months. After outside neuro-ophthalmologic consultation was inconclusive, she presented for second opinion, confused about her medications. Her vision was CF at 5 feet OD and 20/400 OS. Humphrey visual fields 24-2 showed generalized depression OU. ERG showed profound delays in

both cone and rod implicit times and decreased amplitudes. Serum digoxin level was 2.3. 3 weeks after stopping digoxin the patient's visual acuity improved to 20/30 OU. Repeat ERG showed improving rod and cone function.

Conclusions:

ERG can indicate a retinal origin of visual loss in digoxin toxicity even with prior macular problems, as the whole retina is affected and the majority of photoreceptors lie outside the macula. Spontaneous improvement is possible because of variations in compliance and metabolism.

References:

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- 5. White PD. An Important Toxic Effect of Digitalis Overdosage on the Vision. *N Engl J Med.* 272:904-5; 1965.

Keywords: Nerve Fiber Layer and Retinal Testing (OCT, ERG Etc), Retina, YES

Financial Disclosures: The authors had no disclosures.

179 (Submitted)

Risk of Glioma Development in Subjects with Neurofibromatosis Type 1 without Glioma on Initial MRI

Marc H. Levin¹, Gregory T. Armstrong², Julian H. Broad², Robert Zimmerman³, Larissa T. Bilaniuk³, Tamara Feygin³, Grant T. Liu¹, Michael J. Fisher²

¹*Children's Hospital of Philadelphia, the Perelman School of Medicine at the University of Pennsylvania, Division of Ophthalmology, Philadelphia, PA, USA,* ²*Children's Hospital of Philadelphia, the Perelman School of Medicine at the University of Pennsylvania, Department of Pediatrics and Division of Oncology, Philadelphia, PA, USA,* ³*Children's Hospital of Philadelphia, the Perelman School of Medicine at the University of Pennsylvania, Department of Radiology, Philadelphia, PA, USA*

Introduction:

Vision loss occurs in nearly one-half of neurofibromatosis type 1 (NF-1) children with an optic pathway glioma (OPG), and is the main indication for systemic chemotherapy in this population. The purpose of this study was to determine whether, in children with NF-1 without an OPG on initial magnetic resonance imaging (MRI), other potentially concerning radiographic features predict later OPG development and associated decreased vision.

Methods:

Children with NF-1 who underwent orbital MRI at a national referral center between 1992 and 2005, and had ≥ 1 year of subsequent visual acuity (VA) data, were identified retrospectively. Three pediatric neuroradiologists independently reviewed the earliest available MRI to provide a consensus assessment. Eyes with optic nerve tortuosity (ONT) were identified using validated operational criteria.¹ Subjects with OPG on initial MRI were excluded. In parallel, the presence of nerve tortuosity, enhancement, or thickening was established based on clinical MRI reports. The value of these radiographic characteristics in predicting long-term OPG development and associated diminished VA (defined as ≥ 0.2 logMAR below age-matched normative value at last testing) was evaluated per eye using uni- and multivariate analyses.

Results:

133 evaluable subjects (264 eyes) with adequate imaging and neuro-ophthalmic follow up were identified. Median age at earliest MRI was 3.5 years; vision was followed 7.1 (mean) $\pm$ 0.3 years after the original MRI. Consensus review established 11 eyes (8.3%) of seven subjects with ONT. Sixteen subjects (12%) later developed a definite OPG, only one of whom experienced related vision loss. Eyes with potentially concerning imaging findings did not develop OPG or associated vision loss more frequently than controls.

Conclusions:

In patients with NF-1 without an OPG on initial imaging, potentially concerning radiographic features do not appear to substantially elevate the risk of vision loss. The excellent prognosis in this population supports less aggressive surveillance in asymptomatic patients without an OPG.

References:

- 1. Armstrong, Localio, Feygin, Bilaniuk, Phillips, et al. Defining optic nerve tortuosity. AJNR Am J Neuroradiol. 28:666-671. 2007.

Keywords: Neuro-Imaging (MRI, CT, etc), Systemic Disease, YES

Financial Disclosures: The authors had no disclosures.

180 (Submitted)

A Retrospective Study of Chronic Relapsing Inflammatory Optic Neuropathy (CRION)

Padmaja Sudhakar¹, Sachin Kedar^{1,2}, Joseph Berger¹

¹University of Kentucky, Dept of Neurology, Lexington, KY, USA, ²University of Kentucky, Dept of Ophthalmology, Lexington, KY, USA

Introduction:

CRION is a poorly understood disease leading to recurrent inflammation of the optic nerve. Very little is known about etio-pathogenesis, course and outcome of this rare entity. We aim to review the demographics, clinical course, diagnostic work-up, treatment and visual outcome in patients with CRION seen at our institute.

Methods:

Clinic charts of patients with CRION who met the following criteria were reviewed- One episode of optic neuritis (ON) and at least 1 relapse- Absence of known causes of ON including neuromyelitis optica (NMO)- Absence of demyelinating disease (white matter lesions on MRI brain and additional neurological deficits).- Demonstrable response to immunosuppressive treatment with relapse on

withdrawal or dose reduction.

Results:

Thirteen patients (9 females, 4 males; age range: 15 to 69 years) with CRION seen between July 2010 and June 2013 were included. Twelve patients had bilateral ON (11 sequential; 1 simultaneous) and 1 unilateral. Disease activity ranged from 8 months to 34 years. Relapse rates ranged from 1 to 30. Final visual acuity (VA) was better than 20/40 in 64% eyes; 20% had severe visual loss (20/200 or worse) including 3 with no light perception. 43% eyes had severe visual field loss (>-15dB) while 60% eyes demonstrated retinal nerve fiber thickness (RNFLT) <75µm, which was independent of relapse rate. CSF abnormalities included mononuclear pleocytosis (1 patient) and elevated protein (1 patient). Non-specific serological marker elevation included anti-thyroid antibodies (1 patient) and ANA (2 patients). There was no predictable response to any of the treatment modalities- steroids (12 patients), methotrexate (2 patients), intravenous immunoglobulin (2 patients), combination (1 patients) and no therapy (4 patients).

Conclusions:

CRION appears to be a heterogeneous autoimmune disease that results in significant retinal nerve fiber loss and poor visual outcomes in more than a third of affected eyes.

References:

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Keywords: Anterior Afferent Visual Pathway (Optic Neuropathy and Chiasm), Miscellaneous, YES

Financial Disclosures: The authors had no disclosures.

181 (Submitted)

Focal venous hypertension as a pathophysiologic mechanism for port-wine stains, the Sturge-Weber syndrome, and related disorders: proof of concept with novel hypothesis for underlying etiological cause.

Cameron F. Parsa¹

Introduction:

To provide an in-depth re-examination of assumed causes of port-wine stains, and the Sturge-Weber, Cobb, Klippel-Trénaunay, and related syndromes to support an alternative unifying pathophysiologic mechanism of venous dysplasia producing focal venous hypertension with attendant tissue responses; to provide proof of concept with new patient data; to propose a novel etiological hypothesis for the venous dysplasia in these syndromes and find supportive evidence.

Methods:

Data from twenty patients with port-wine stains and corneal pachymetry readings was collected prospectively by the author in an institutional referral-based practice. The literature was searched using Medline, and articles and textbooks were obtained from the bibliographies of these publications.

Results:

Newly obtained dermatologic, corneal pachymetry, fundus ophthalmoscopic, ocular and orbital venous Doppler ultrasonography, and magnetic resonance imaging findings in patients with the Sturge-Weber syndrome or isolated port-wine stains, along with published data, reveal diffusely thickened tissues and neural atrophy in all areas associated with venous congestion.

Conclusions:

Contrary to traditional understanding, signs and symptoms in the Sturge-Weber and related syndromes, including both congenital and acquired port-wine stains are shown to arise from effects of localized primary venous dysplasia or acquired venous obstruction rather than neural dysfunction, differentiating these syndromes from actual phacomatoses. Effects of focal venous hypertension are transmitted to nearby areas via compensatory collateral venous channels in the above conditions, as in the Parkes Weber syndrome. A novel underlying etiology—prenatal venous thromboocclusion—may be responsible for the absence of veins with persistence and enlargement of collateral circulatory pathways with data in the literature backing this offshoot hypothesis.

Keywords: Systemic Disease, Miscellaneous, YES

Financial Disclosures: The authors had no disclosures.

182 (Submitted)

Acute Optic Neuropathy Is Not Associated with Posttraumatic Stress Disorder contrary to Rhegmatogenous Retinal Detachmen

Ruth Huna-Baron¹, Anat Abudi², Anna Liashets-Peer¹, Michel Kinori¹, Joseph Moisseiev¹, Joseph Zohar², Ido Didi Fabian¹

¹*The Goldschleger Eye Institute The Sheba Medical Center and The Sackler Faculty of Medicine, Tel Aviv University, Ramat-Gan, Israel,* ²*The Division of Psycnter and The Sackler Faculty of Medicine, Tel Aviv University , Ramat-Gan, Israel*

Introduction:

Posttraumatic stress disorder (PTSD) is an anxiety disorder that may develop as a consequence of ocular insult. The burden of PTSD can be high and cause functioning disability. We studied the prevalence and risk factors of PTSD in an acute optic neuropathy (AON) cohort, and compared these

findings with those reported on patients following surgery for rhegmatogenous retinal detachment (RRD).

Methods:

A prospective consecutive observational study of 171 AON patients and 366 patients who underwent surgery for RRD. Study patients were screened for the existence of PTSD symptoms via telephone survey, and those positively identified as having PTSD underwent a structured psychiatric interview.

Results:

None of the 171 AON patients was diagnosed with PTSD, as opposed to 9 out of 366 patients (2.5%) in RRD patients ($P = .063$). No significant differences were found in visual acuity at presentation and last follow-up visit between the two groups (logMAR 0.73 ± 0.80 for the RRD group and logMAR 0.36 ± 0.45 for the AON group, $P \geq .29$). All of the RRD patients underwent surgery compared to none of the AON patients ($P < .001$), and 58 (34%) of the AON patients were administered steroids during the acute phase compared to none of the RRD patients ($P < .001$). Past history of trauma emerged as being a risk factor for the development of PTSD ($P = .004$). There were no statistical group differences in the 25-Item National Eye Institute Visual Function Questionnaire scores.

Conclusions:

PTSD tended to be associated with RRD but not with AON. It may result from the smaller number of AON patients in our study or from the third AON patients treated with corticosteroids in the acute phase. There are data suggesting that glucocorticoids given exogenously in proximity to the traumatic event may serve to prevent the development of PTSD.

References:

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Fabry Disease with Anterior Ischemic Optic Neuropathy

An-Guor Wang^{1,3}, Chiao-Ying Liang², May-Yung Yen^{1,3}

¹Taipei Veterans General Hospital, Department of Ophthalmology, Taipei, Taiwan, ²Taichung Veterans General Hospital, Department of Ophthalmology, Taichung, Taiwan, ³National Yang-Ming University School of Medicine, Taipei, Taiwan

Introduction:

Fabry disease is a X-linked inherited lysosomal storage disease due to the absence or reduction of α -galactosidase A activity. Anterior ischemic optic neuropathy (AION) has rarely been reported in Fabry disease. Herein we report a case with bilateral sequential AION.

Methods:

case report

Results:

A 14-year-old boy suffered from sudden loss of vision OS 3 days after mountain climbing. BCVA was 6/6 OD and HM/20cm OS. Slitlamp revealed faint nebula of superficial cornea and subtle lens opacity OU. There was a markedly pale swelling of the left optic disc, accompanied with tortuosity of retinal vessels OU. FAG revealed markedly delayed filling of left optic disc and choroid. Visual field was full OD and totally obscured OS. CT scan revealed an enlarged left intraconal optic nerve with contrast enhancement. He received methylprednisolone pulse therapy, but his vision remained CF/10cm. Another attack occurred in the right eye 7 months later. BCVA dropped from 6/5 to CF/30cm OD within 24 hours after a scheduled MRI examination. FAG revealed filling defect in right optic disc. Steroid pulse therapy was given immediately with systemic urokinase, coenzyme Q10 and L-arginine. Two days later, his vision did not improve and then hyperbaric oxygen (HBO) was used. His vision recovered to 6/7.5 OD on the next morning after HBO therapy.

Conclusions:

Fabry disease should be suspected by the characteristic corneal verticillata, spoke-like cataract, and conjunctival/retinal vessel tortuosities. The diagnosis could be confirmed with enzyme level measurement or genetic analysis. Enzyme replacement therapy with recombinant human α -galactosidase A has been shown safe and effective. Hyperbaric oxygen therapy could be beneficial in Fabry patient with ischemic vasculopathy.

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