### POSTER PRESENTATIONS

#### 39th Annual Meeting

February 9–14, 2013
Snowbird Ski Resort • Snowbird, Utah

**North American Neuro-Ophthalmology Society**

**Tuesday, February 12, 2013 • 6:00 p.m. – 9:30 p.m.**

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

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

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

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

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*Poster 8 has been withdrawn.*
Redefining Wolfram Syndrome in the Molecular Era

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Introduction:
Wolfram syndrome is a progressive neurodegenerative disorder that was historically defined by a cluster of common clinical manifestations: diabetes insipidus, diabetes mellitus, optic atrophy and deafness (DIDMOAD). In the premolecular era, Wolfram syndrome was thought to be relatively rare with an estimated prevalence of 1 in 770,000 in the United Kingdom. Although genetically heterogeneous, the majority of affected patients harbor pathogenic WFS1 mutations inherited as an autosomal-recessive trait. As part of our ongoing epidemiological study of inherited optic neuropathies in the North of England, it became apparent that Wolfram syndrome is a more complex disease than originally considered with direct implications for patient screening and genetic counselling.

Methods:
This study included twenty probands with suspected inherited optic atrophy in whom previous molecular investigations had failed to identify pathogenic mitochondrial DNA mutations, and mutations within the OPA1 and OPA3 genes. The entire WFS1 coding region was sequenced using genomic DNA samples extracted from peripheral blood leukocytes.

Results:
Pathogenic WFS1 mutations were identified in five probands. Three probands harbored compound heterozygous autosomal-recessive WFS1 mutations. In two probands, single heterozygous autosomal-dominant WFS1 mutations were found. A wide clinical spectrum of varying disease severity was observed: (i) optic atrophy and early-onset sensorineural deafness (n = 2), (ii) optic atrophy and late-onset diabetes mellitus (n = 1), and (iii) a progressive neurodegenerative phenotype (n = 2), with only one patient fulfilling the DIDMOAD criteria. Based on our epidemiological data, the minimum prevalence of this disorder is estimated at 1 in 440,000 in the North of England.

Conclusions:
The greater availability of WFS1 testing has expanded the clinical phenotypes associated with Wolfram syndrome. Interestingly, WFS1 mutations can exhibit an autosomal-dominant mode of inheritance. Genetic screening should therefore be considered in suspected cases of inherited optic atrophy, especially if associated with sensorineural deafness or diabetes mellitus.

References:

Keywords: Wolfram syndrome, DIDMOAD, Autosomal dominant optic atrophy, Mitochondrial disorders

Financial Disclosures: The authors had no disclosures.
Poster 2*

Management and Outcomes of Idiopathic Intracranial Hypertension with Moderate-Severe Visual Field Loss: Pilot Data for the Surgical Idiopathic Intracranial Hypertension Treatment Trial

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Introduction:
The management of idiopathic intracranial hypertension (IIH) remains controversial. The Idiopathic Intracranial Hypertension Treatment Trial (IIHTT) is investigating medical intervention (weight loss +/- acetazolamide) for newly-diagnosed IIH patients with baseline mean deviation (MD) between -2 dB – -7.00 dB on automated perimetry with Humphrey Field Analyzer (HFA). We collected pilot data on management and outcomes of IIH patients with moderate-severe visual field loss at presentation, in preparation for the Surgical IIH Treatment Trial.

Methods:
This was a multi-center, retrospective study. Data were requested from 40 Neuro-Ophthalmology Research Disease Investigator Consortium (NORDIC) sites. Patients with newly-diagnosed IIH satisfying modified Dandy criteria seen between January 2009 and September 2012 with baseline MD worse than -7.00 dB were included. Data were gathered regarding treatment approach – medical (weight loss +/- acetazolamide) or surgical (optic nerve sheath fenestration (ONSF) or cerebrospinal fluid (CSF) shunting). HFA outcomes at 3-12 months were analyzed.

Results:
Of over 400 patients meeting inclusion criteria, 107 had pre- and post-intervention HFA data available. Of 53 patients treated medically, baseline average MD was -11.90 +/- 8.21 dB. Final MD at 3-12 months was -7.76 +/- 8.39 dB, with a mean change of + 4.14 dB. Of 27 patients treated with ONSF, baseline MD was -16.49 +/- 9.92 dB. Final MD was -13.51 +/- 10.81 dB, with a mean change of +2.98 dB. Of 27 patients treated with CSF shunting, baseline MD was -17.67 +/- 9.43 dB. Final MD was -9.96 +/- 10.07 dB with a mean change of +7.71 dB. Eyes worsening ≥ -4.00 dB from baseline or unable to perform follow-up perimetry in the medical, ONSF, and CSF shunting groups were 4.7%, 14.8%, and 3.7%, respectively.

Conclusions:
Although limited by their retrospective nature, these data suggest that medical and surgical interventions can be effective for moderate-severe visual field loss in IIH, with a suggestion that CSF shunting might be more effective.

Keywords: Idiopathic Intracranial Hypertension, Medical Treatment, Optic Nerve Sheath Fenestration, Cerebrospinal Fluid Shunting, Acetazolamide

Financial Disclosures: The authors had no disclosures.
Correlation Between Clinical Parameters And Diffusion-Weighted Magnetic Resonance Imaging In Idiopathic Intracranial Hypertension

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Introduction:
Idiopathic intracranial hypertension (IIH) is a neurological disorder characterized by increased intracranial pressure without an identifiable etiology and is an important cause of papilledema and visual loss [1, 2]. Although many patients retain good visual function, permanent visual loss can occur and there are few clinical parameters that are predictive of final visual outcome [1]. Orbital imaging offers an alternative and quantitative method to assess papilledema as well as other structural changes in the optic nerve characteristic of IIH [3-5]. Diffusion-weighted magnetic resonance imaging (DW-MRI) hyperintensity of the optic nerve head (ONH) has been reported [6], but the degree to which this finding correlates with visual parameters and outcomes remains unknown.

Methods:
A retrospective chart review of patients with securely diagnosed IIH by clinical examination and visual field analysis who underwent orbital imaging within four weeks of diagnosis was conducted. DW-MRI images were assessed for each eye by a masked neuroradiologist for the degree of diffusion restriction in the ONH. We compared DW-MRI grading with visual field mean deviation and papilledema grade by Kendall’s Tau Rank correlation analysis and t-tests.

Results:
Forty-two patients (41 female, 1 male) with a mean age of 31.2±10.0 years were included in the study. A statistically significant difference (p=0.0195) was found between papilledema grade and patients with severe DW-MRI findings (n=16; 3.75±1.25) versus non-severe (n=26; 2.79±1.24). No other significant differences were found between papilledema grade, visual field mean deviation or visual acuity, and DW-MRI findings.

Conclusions:
We found a significant correlation between ONH diffusion restriction and severity of papilledema in patients with IIH but not with visual field loss or other visual parameters. This finding might provide insight into the pathophysiology of papilledema in IIH. It is possible that DWI-ONH changes could predict future visual outcome in such patients and this analysis is ongoing.

References:

Keywords: Idiopathic intracranial hypertension, Papilledema, Diffusion-weighted magnetic resonance imaging (DW-MRI)

Financial Disclosures: The authors had no disclosures.
**Contrast Sensitivity Visual Acuity Defects in the Earliest Stages of Parkinsonism**

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**Introduction:**
To demonstrate that contrast sensitivity defects are present in patients with Parkinsonism in its earliest stages. Testing for such deficiencies may prove a sensitive screening tool for this disease. Diminished contrast sensitivity visual acuity is recognized in patients with Parkinson’s disease (PD), presumably due to loss of retinal dopamine. However, to date no study has detailed the presence or degree of loss correlated with the stages of Parkinsonism, particularly the earliest stages.

**Methods:**
25 early-stage Parkinson’s disease subjects Stage I-II H&Y (15 Stage I,10 Stage II) versus 25 controls were studied. Participants completed questionnaires about their vision, eye pathology, current medications, and four symptoms typically seen in premotoric state of PD. Tests included UPDRS, directive and contrast sensitivity visual acuities (SLOAN) at 100%, 2.5%, and 1.25%, intraocular pressure, Schirmer’s test for tear film formation, HRR color vision tests, confrontational visual fields, extraocular movements, pupillary functions, dilated funduscopic exam, and spectral Optical Coherence Tomography (OCT) to quantitatively assess inner retinal layers. Visual acuity <20/50 and known co-morbid ophthalmologic pathologies were exclusionary. The statistical methods used were the student’s t-test and analysis of variance.

**Results:**
Univariate analysis demonstrates a highly statistical difference between patients with early PD and controls. PD patients showed statistically poorer contrast acuity at 2.5% and 1.25% visual performance (P<0.05). No significant difference existed in ganglion cell layers between patients and controls. Further multivariate analysis comparing participants will be discussed, including the relative frequency of contrast sensitivity defects against acknowledged premotoric symptoms.

**Conclusions:**
In early PD patients with normal directive visual acuity, we found significantly decreased contrast sensitivity visual acuity. Contrast sensitivity testing in the office setting (SLOAN Wall Chart) can objectively detect early subclinical PD-related visual impairments.

**Keywords:** Parkinsonism, Contrast Sensitivity Acuity, Dopamine, Retina, Optical Coherence Tomography

**Financial Disclosures:** The authors had no disclosures.
**Visual Function and Freedom from Disease Activity in a Phase 3 Trial for Relapsing Multiple Sclerosis**

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**Introduction:**
In the phase 3 trial of natalizumab for relapsing MS (AFFIRM), low-contrast letter acuity (LCA) showed decreased probability of sustained visual loss in patients receiving active treatment. Freedom from clinical and radiological disease activity was also seen in 37% of treated patients over 2 years in AFFIRM. Given the importance of vision to overall functioning and quality of life in MS, we determined how freedom from disease activity might be associated with visual loss.

**Methods:**
Freedom from clinical disease activity was defined as no relapse and no sustained disability progression by the Expanded Disability Status Scale (EDSS); freedom from radiological disease activity was defined as no gadolinium-enhancing (Gd+) lesions and no new or enlarging T2-hyperintense lesions on MRI. Overall freedom from disease activity was defined as no activity on clinical and radiological measures over 2 years. Changes from baseline in LCA (2.5% and 1.25% contrast) and high-contrast visual acuity (VA) over 2 years were compared between groups with and without disease activity using analysis of covariance (ANCOVA), adjusted for baseline visual function score.

**Results:**
In the AFFIRM study cohort (n=942 patients), mean changes from baseline for LCA and VA were generally greater and in the direction of improved function over time in patients who were free of disease activity compared with those who were not. The overall disease activity free group was 41% less likely to have worsening of 1.25% LCA (P=0.007), and 78% less likely to have VA worsening (P=0.04) over 2 years, adjusting for baseline visual function score. Less LCA worsening was associated with freedom from clinical disease activity (P=0.0004), while less VA loss correlated with freedom from radiological disease activity (P=0.007).

**Conclusions:**
Patients free of MS disease activity are less likely to have VA worsening and have a greater chance of visual improvement. LCA is an important clinical trial outcome measure.

**References:**


**Keywords:** Visual function, Multiple sclerosis, Low-contrast letter acuity, Natalizumab

**Financial Disclosures:** Dr. Balcer has received speaking and consulting honoraria from Bayer, Biogen Idec, and Novartis, and is a member of a clinical trial advisory board for Biogen Idec. Dr. Galetta has received speaking and consulting honoraria from Biogen Idec, Novartis, and Teva. Drs. Zhang and Arnold are employees of Biogen Idec.
Dimensions of the Optic Nerve Head Neural Canal Using Enhanced Depth Imaging Optical Coherence Tomography in Non-Arteritic Ischemic Optic Neuropathy Compared to Normal Subjects

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Introduction:
An anatomically crowded scleral canal has been linked to the development of non-arteritic anterior ischemic optic neuropathy (NAION). Previous authors have described the superficial characteristics of a “disc at risk” in NAION. We sought to investigate the deeper structures of the optic nerve head neural canal using enhanced depth imaging optical coherence tomography (EDI OCT) in NAION patients.

Methods:
We conducted a prospective study of patients with NAION and normal controls. Complete ophthalmologic examination including axial length (AL) was performed on all patients. Serial horizontal and vertical EDI OCT of the optic nerve head were performed for both eyes of each subject. Horizontal and vertical diameters of: the optic disc, Bruch’s membrane opening (BMO), lamina cribrosa (LC), and narrowest neural canal opening (NCO) were measured and compared.

Results:
Data from 30 patients with NAION and 45 normal subjects were analyzed. The measurement of neural canal dimensions showed excellent measurement reproducibility (intraclass correlation coefficient: 0.921-0.972). Mean horizontal and vertical diameters of the optic disc, BMO, and narrowest NCO were smaller in NAION patients compared to control subjects, but their differences did not reach statistical significance (p > 0.1). Mean horizontal and vertical diameters of the LC were significantly smaller in the NAION group (1614 +/- 163 μm and 1693 +/- 210 μm, respectively) compared to those in the control group (1854 +/- 218 μm and 1932 +/- 248, respectively), p <0.001, controlling for age, gender, and AL . The unaffected fellow eyes of NAION patients had neural canals of similar size to those of NAION eyes.

Conclusions:
Based on EDI OCT imaging of the optic nerve head neural canal, the LC is smaller in NAION eyes compared to controls. Crowding at the level of the LC, where branches of the short posterior ciliary arteries enter the optic nerve, may play a role in the pathophysiology of NAION.

Keywords: Non-arteritic anterior ischemic optic neuropathy, Enhanced depth imaging (EDI), Optical coherence tomography, Lamina cribrosa, Disc at risk

Financial Disclosures: Jeffrey M. Liebmann received research support from Carl Zeiss Meditec, Inc., Heidelberg EngineeringGmbHOptovue, Inc., and Topcon Medical Systems, Inc. No other authors had disclosures.
Eye Movement Perimetry: Evaluation of Saccadic Latency, Saccadic Amplitude, and Visual Threshold to Peripheral Visual Stimuli in Young Compared With Older Adults

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Introduction:
Previous studies have evaluated for age-related differences in saccadic latency and amplitude to peripheral visual stimuli, but have not evaluated for differences using stimuli close to visual threshold. Several studies have suggested that eye movements could be used to evaluate for visual field loss, but have not evaluated the relationship between saccadic latency, amplitude, and visual threshold. We aimed to determine saccadic latency and amplitude, as well as visual threshold, to peripheral visual stimuli in young adults compared with older adults using a novel automated perimetry technique.

Methods:
We tested 20 normal subjects aged 18-30 years old (“young”) and 21 normal subjects at least 60 years old (“older”) for detection of briefly-flashed peripheral visual stimuli of differing sizes in 8 locations along the horizontal meridian (±4°, ±12°, ±20°, and ±28°). Subjects were instructed to look toward the stimulus and eye movements were recorded with an EyeLink 1000 infrared camera with temporal resolution of 1000 Hz. Limiting our analysis to the stimuli in the nasal hemifield (-4°, -12°, -20°, and -28°), we evaluated for group-level differences in saccadic latency, amplitude, and visual threshold for each stimulus position.

Results:
Saccadic latency increased as stimulus size approached visual threshold in both groups. Older subjects had increased saccadic latencies, decreased saccadic amplitudes, and higher visual thresholds. Repeated-measures ANOVA on ranks demonstrated significant differences (p <0.05) between the groups in saccadic latency (at all positions), amplitude (at all positions), and visual threshold (at -12°, -20°, and -28°). In both groups, saccadic latency and visual threshold were positively correlated, whereas amplitude was negatively correlated with saccadic latency and visual threshold.

Conclusions:
Older adults have increased latency and decreased amplitude of saccades, as well as higher visual thresholds, to peripheral visual stimuli compared with younger adults. Saccadic latency and amplitude are correlated with visual threshold.

References:

Keywords: Visual Fields, Saccadic Eye Movements

Financial Disclosures: Funding was received from Veterans Affairs Rehabilitation R&D Merit Review Grant.
Advanced MRI of Optic Nerve Drusen: Preliminary Findings

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Introduction:
Optic nerve head drusen (ONHD) are thought to be related to axonal metabolism and degeneration.[1] The pathogenesis of vision loss from ONHD is not well understood. We have developed a multi-shot, high resolution, rapid, artifacts minimized, diffusion tensor imaging (DTI) magnetic resonance imaging (MRI) acquisition.[2-4] We postulate that ONHD cause significant changes in diffusion indices and anticipate the sensitivity to detect differences between healthy volunteers and ONHD patients.

Methods:
Four patients with ONHD and visual field defects underwent exam, optical coherence tomography (OCT), and standardized automated perimetry (SAP). We performed 3T-MRI of 4 patients diagnosed with ONHD and 18 healthy controls. Optic nerves were segmented and mean cross-sectional area (CSA) reported. DTI was performed in two ONHD patients and quantitative diffusion indices of fractional anisotropy (FA), mean (MD), parallel and perpendicular diffusivities were calculated. Non-parametric rank sum tests were performed.

Results:
All ONHD patients had slow visual field loss. Average age was 30 (SD 18.32), vision was 20/20 in all eyes except one (0.01 logMAR, SD 0.035). SAP mean defect was 16.73 DB (SD 8.37). OCT of optic nerves measured 55.75 microns (SD 10.36). SAP loss varied from nasal defects to only a central island of vision. CSA in ONHD patients was less than healthy controls (11.4 ± 3.6 mm², 17.1 ± 4.1mm², p = 0.03). In ONHD patients, FA was lower, while MD, parallel- and perpendicular-diffusivity were elevated.

Conclusions:
Advanced MRI tools quantified ONHD pathology. In this small cohort, the nerve appears atrophic and the diffusion indices, abnormal. FA reflects a loss of axonal density. The higher MD values indicate inflammatory changes in the ONHD optic nerves. FA and MD findings suggest that a radiologic biomarker for axonal transport may exist and serve as a potent predictor and provide quantitative data to support a diagnosis of optic nerve axonal vision loss.

References:

Keywords: Optic nerve drusen, MRI, Diffusion Tensor Imaging, Atrophy

Financial Disclosures: The authors had no disclosures.
Clinical Features of OPA1-Related Optic Neuropathy: A Retrospective Case Series

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Introduction:
Dominant optic atrophy (DOA) is the most common hereditary optic neuropathy. The most common form of DOA, DOA type 1 (Kjer’s disease) accounts for 40-60% of cases and is associated with mutation of the OPA1 gene. In the largest similar study to date, clinical data were analyzed for mutation positive patients for OPA1, the OPA3 gene, and Leber's hereditary optic neuropathy1. In the present study, we evaluated clinical information for patients referred to a major tertiary care center for OPA1 testing for suspected DOA.

Methods:
62 samples from patients were prospectively collected, between April 2006 and June 2011, for PCR-based sequencing of the OPA1 gene. Retrospective clinical data were available for 54 (31 male; 23 female) of these individuals. Missense and small deletions/insertions were compared with previously reported mutations, and objective analyses were used to determine the likelihood of pathogenicity for specific DNA changes. Multiplex Ligation-dependent Probe Amplification (MLPA) analysis was performed on a subset of patients. The clinical features of the mutation-positive and mutation-negative individuals were compared.

Results:
62 patients underwent diagnostic OPA1 sequencing, 18 (29.0%) were found to have potentially disease-causing mutations by sequence analysis; 3 of these mutations are novel. MLPA analysis identified 3 patients with large-scale deletions unrecognized by sequencing. A majority (28/41; 68.3%) of referred cases were sent based in part on the appearance of the optic nerves. Comparison of the clinical data showed that automated visual field testing was the most sensitive measure of disease status in a subset of individuals.

Conclusions:
Our results show that OPA1 mutations are a common cause of inherited optic neuropathy in the tertiary referral setting. We believe this is the first time that both sequence analysis and MLPA testing have been used in conjunction with robust clinical data to compare features between a large number of mutation-positive and mutation-negative individuals.

References:

Keywords: Dominant optic atrophy, Kjer's disease, OPA1 gene, Hereditary optic neuropathy

Financial Disclosures: The authors had no disclosures.
The Utility of Orbital Ultrasound in the Diagnosis of Papilledema

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Introduction:
The distinction between pseudopapilledema (i.e. drusen or anomalous discs) and papilledema by ophthalmoscopic examination is not always readily apparent and there is no gold standard test for distinguishing between these two entities. We evaluated the utility of ultrasound (i.e. sensitivity and specificity) in distinguishing papilledema from pseudopapilledema in a large sample of adult patients.

Methods:
This was a retrospective chart review of patients with elevated optic discs who were evaluated by neuro-ophthalmologists and underwent orbital ultrasound by a single ultrasonographer over a 7 year period. Correlation was made between the final diagnosis of papilledema versus pseudopapilledema and ultrasound findings of presence or absence of dilated retrobulbar optic nerve sheaths and positive changes on the 30-degree test (1).

Results:
Amongst 56 patients enrolled, the average age was 40.4 years (range 18-79, SD± 14.1) with 21.4% males (n=12) and 78.6% females (n=44). The average follow up was 7.5 months (range 1.1-40.4, SD= 8.0). 35.7% of patients (n=20) had papilledema and 64.3% (n=36) had pseudopapilledema. Ultrasonography showing increased optic nerve width (>3.3 mm) and a positive 30-degree test was found to be 85.0 % (n=17/20) sensitive for papilledema and 83.3% specific (n=30/36), with negative findings highly correlated with a final diagnosis of pseudopapilledema. Of 30 controls none had positive findings on ultrasonography.

Conclusions:
Orbital ultrasound findings of increased optic nerve width and positive 30-degree tests are highly sensitive for the diagnosis of papilledema and also highly specific in a large sample of patients with significant neuro-ophthalmologic follow up. Although ultrasound does not replace clinical assessment, our study is consistent with small case series that suggest ultrasonography is a sensitive and specific tool (2) that can aid in distinguishing between papilledema and pseudopapilledema.

References:


Keywords: Papilledema, Orbital ultrasound

Financial Disclosures: The authors had no disclosures.
**Poster 12**

**Longitudinally Extensive Optic Neuritis Distinguishes Neuromyelitis Optica from Multiple Sclerosis**

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**Introduction:**
The anterior visual pathway is morphologically and physiologically part of the central nervous system, and its response to injury is similar. Demyelinating and inflammatory processes may affect the AVP and are typically disruptive of the local blood brain barrier, as is frequently noted in optic neuritis. One report showed that enhancement length did not distinguish optic neuritis between neuromyelitis optica (NMO) and multiple sclerosis (MS) when multiple cases were averaged. However, the specificity for longitudinally extensive enhancement of the AVP to differentiate optic neuritis among NMO, MS, and sarcoidosis has not been previously reported. Radiographic analysis can help distinguish abnormal processes affecting the optic nerves and anterior visual pathways (AVP). We hypothesize that length of pathologic AVP contrast enhancement can be a predictor of etiologic diagnosis in optic neuritis.

**Methods:**
A University Institutional Review Board approved this retrospective study. Patients presenting with optic neuritis secondary to multiple sclerosis, neuromyelitis optica, or sarcoidosis who had a neuro-ophthalmological exam and MRI within four weeks of onset were selected. Total length of gadolinium enhancement involving the afferent visual system was measured by an investigator blinded to the clinical diagnosis. Presence of pain, papillitis, and visual field metrics were also recorded.

**Results:**
Thirty-five patients were selected (25 MS, 5 NMO, 5 sarcoid). P values were <0.05 for bilateral involvement, chiasm involvement, and total AVP enhancement measuring >60mm for NMO when compared to MS. Length of enhancement did not distinguish sarcoid from NMO or MS. Neither pain, disc swelling, nor baseline HVF metrics differentiated NMO, MS, or sarcoid.

**Conclusions:**
Besides the presence of bilateral and chiasmal involvement, already described as characteristic of NMO, we here demonstrate that afferent visual system enhancement length greater than 60mm, a term we propose as ‘longitudinally extensive optic neuritis’, helps distinguish NMO from MS.

**References:**


**Keywords:** Optic neuritis, Multiple sclerosis, Neuromyelitis Optica, Magnetic Resonance Imaging, Sarcoid

**Financial Disclosures:** The authors had no disclosures.
Thyroid Eye Disease - Pilot Study

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Introduction:
Thyroid eye disease (TED) is a poorly understood and difficult disease process to manage. The ultimate goal of TED research is to be able to perform clinical efficacy studies of therapeutic strategies for active TED. Currently, however, there is no objective, generally accepted assessment tool that defines active disease. The purpose of this study was to identify objective ancillary tests and clinical measures that correlate with thyroid eye disease activity.

Methods:
Fifty-two patients with thyroid eye disease were recruited from three different clinics from two states. Patients were provided with their typical clinical examination and appropriate tests, external photos, serum immunology (TSH, TSI, free T4), and orbital imaging (CT or MRI). Based on the clinical examination, the VISA and clinical assessment score (CAS) were calculated. Additionally, two questionnaires (TEDQOL and VFQ-25) were administered.

Results:
Of the fifty-two patients recruited, 14 (27%) were considered “active.” Results indicated that CAS, VISA, optic neuropathy, the VFQ-25, and the “function” subset of the TEDQOL were significantly correlated with disease activity. Regarding the objective assessment tools (serum immunology and MRI), only TSI significantly correlated with activity of disease.

Conclusions:
The results of this study, therefore, suggest that TSI may be a useful objective assessment tool to determine the status of disease activity in TED. Future studies will need to examine whether these levels have any relation to the anticipated course of the disease or patients’ response to therapeutic modalities.

Keywords: Thyroid eye disease, TSI

Financial Disclosures: The authors had no disclosures.
Non-Mydriatic Ocular Fundus Photography Among Headache Patients in an Emergency Department

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Introduction:
Examination of the ocular fundus is one aspect of the physical examination that is helpful in screening for “red flags,” such as papilledema, that suggest a more ominous cause of headache. However, ocular funduscopic examination is rarely and poorly performed by non-ophthalmology personnel, especially in acute care settings such as the ED. Our objective was to determine the frequency of and the predictive factors for abnormal ocular fundus findings among emergency department (ED) headache patients.

Methods:
Cross-sectional study of prospectively enrolled adult patients presenting to our ED with a chief complaint of headache. Ocular fundus photographs were obtained using a non-mydriatic fundus camera that does not require pupillary dilation. Demographic and neuro-imaging information were collected. Two neuro-ophthalmologists independently reviewed photographs for findings relevant to acute care. The results were analyzed using univariate statistics and logistic regression modeling.

Results:
We included 497 patients (median age: 40 years, 73% women), among whom 42 (8.5%, 95%CI: 6-11%) had ocular fundus abnormalities. Of these 42 patients, 12 had disc edema, 9 had optic nerve pallor, 6 had grade III/IV hypertensive retinopathy, and 15 had isolated retinal hemorrhages. Body mass index ≥35 (odds ratio [OR]: 2.3, p=0.02), younger age (OR: 0.7 per 10-year increase, p=0.02), and higher mean arterial blood pressure (OR: 1.3 per 10 mmHg increase, p=0.003) were predictive of abnormal fundus photography. Patients with an abnormal fundus had a higher percentage of hospital admission (21% vs. 10%, p=0.04). Among the 34 patients with abnormal ocular fundi who had brain imaging, 14 (41%) had normal imaging.

Conclusions:
Ocular fundus abnormalities were found in 8.5% of headache patients presenting to our ED. Predictors of abnormal funduscopic findings included higher body mass index, younger age, and higher blood pressure. Our study confirms the importance of funduscopic examination in headache patients, particularly in the ED, and reaffirms the utility of non-mydriatic fundus photography in this setting.

Keywords: Non-mydriatic fundus photography, Headache, Papilledema, Optic disc pallor, Hypertensive retinopathy

Financial Disclosures: This study was supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York, and by the NIH/NEI core grant P30-EY06360.
Replacing the Direct Ophthalmoscope: Diagnostic Accuracy and use of Non-Mydriatic Ocular Fundus Photography by Emergency Department Physicians (Phase II of the FOTO-ED study)

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Introduction:
During the first phase of the FOTO-ED Study, 13% of patients had an ocular fundus finding, such as papilledema, relevant to their emergency department (ED) management. All of these findings were missed by ED physicians, who only examined 14% of enrolled patients by direct ophthalmoscopy. Our aim was to evaluate the sensitivity of non-mydriatic ocular fundus photography, an alternative to direct ophthalmoscopy, for relevant ocular fundus findings when photographs were read real-time by ED physicians during routine clinical care.

Methods:
354 patients presenting to our ED with headache, focal neurologic deficit, visual change, or diastolic blood pressure ≥120 had non-mydriatic fundus photography obtained (Kowa nonmyd-alpha-D). Photographs were placed on the electronic medical record for review by ED physicians. Identification of relevant ocular fundus findings (defined a priori as optic disc edema, grade III/IV hypertensive retinopathy, isolated intraocular hemorrhages, optic disc pallor, and retinal vascular occlusion) on photographs by ED physicians was compared to a reference standard of review by neuro-ophthalmologists.

Results:
ED physicians reviewed the photographs of 239 patients (68%). 35 patients (10%) had relevant findings identified by neuro-ophthalmologist review (6 disc edema, 6 grade III/IV hypertensive retinopathy, 7 isolated hemorrhages, 15 optic disc pallor, and 1 retinal vascular occlusion). 16 of 35 relevant findings (46%) were identified by the ED physicians, eight (24%) occurred in patients whose photographs were not reviewed by ED physicians, and the remaining were missed on photographs reviewed but recorded as “likely normal” or “normal”. The ED physicians reported that the photographs were helpful for 125 patients (35%).

Conclusions:
ED physicians used non-mydriatic fundus photographs more frequently than they perform direct ophthalmoscopy, and their detection of relevant abnormalities was improved. Ocular fundus photography often assisted with clinical decisions even when normal. Non-mydriatic ocular fundus photography offers a promising alternative to direct ophthalmoscopy.

Keywords: Non-mydriatic fundus photography, Direct ophthalmoscopy, Emergency department, Diagnostic test evaluation

Financial Disclosures: This study was supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York, and by the NIH/NEI core grant P30-EY06360 (Department of Ophthalmology). Dr. Newman received the Research to Prevent Blindness Lew R. Wasserman Merit Award. Dr. Bruce received research support from the NIH/NEI (K23-EY019341).
Saccades and Memory: Baseline Correlations of the King-Devick and SCAT2 Tests in Professional Hockey Players

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Introduction:
The King-Devick (K-D) test, a vision-based measure based on the speed of rapid-number-naming, has been proposed as a sideline tool to detect sports-related concussion. Requiring <1 minute to administer, the K-D test captures impaired eye movements and saccades, findings that correlate with suboptimal brain function. K-D time scores are significantly higher (worse) in athletes immediately following concussion, consistent with involvement of widely distributed visual pathways. The purpose of this study was to determine the relation of K-D scores to the more cognitively-based components of the Sport Concussion Assessment Tool (SCAT2) in a professional hockey team cohort during pre-season baseline testing. We also examined changes in K-D and modified SCAT2 scores (without balance testing due to skates) for two athletes who developed concussion and had rinkside testing.

Methods:
The modified SCAT2 and the K-D test, a brief measure of rapid number naming, were administered to members of a professional hockey team during the 2011-12 pre-season. Athletes with concussion also underwent rinkside testing.

Results:
Among 27 athletes tested, lower (worse) scores for the SCAT2 Standardized Assessment of Concussion (SAC) Immediate Memory Score (P<0.001) and the overall SAC score (P=0.01) were associated with greater (worse) times required to complete the K-D test at baseline, accounting for age. In two players tested rinkside immediately following concussion, K-D test scores worsened from baseline by 4.2 and 6.4 seconds. These athletes had no differences found for SCAT2 SAC components, but reported symptoms of concussion.

Conclusions:
In this preliminary study of professional athletes, K-D test scores, which capture saccadic eye movements and attention, were associated with reductions in immediate memory at a pre-season baseline. Both working memory and saccades share closely related anatomical structures, including the dorsolateral prefrontal cortex (DLPFC). A rapid sideline test of saccades is likely to be an important clinical tool to assess the athlete with suspected concussion.

References:

Keywords: Saccades, Eye movements, Concussion, Memory, Sports

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Introduction: Ocular myasthenia gravis (OMG) is a common presentation to the neuro-ophthalmology clinic. Previous studies have reported that generalized myasthenia gravis (GMG) develops in 50% or more of patients presenting with OMG, usually within 2 years. We sought to calculate the conversion rate of OMG to GMG at our neuro-ophthalmology clinic and identify any factors influencing generalization.

Methods: We conducted a retrospective chart review of myasthenia gravis patients between 1993 and 2012. Inclusion criteria required patients ≥18 years of age, symptoms of diplopia, ptosis or both, and at least 2 years of follow up after diagnosis. Exclusion criteria included systemic symptoms at presentation, history of thyroid eye disease, or strabismus surgery. OMG was defined as presence of diplopia and/or ptosis, and positive acetylcholine receptor Ab, single fiber electromyography (EMG), Tensilon test, or response to treatment. GMG was defined as the development of systemic symptoms including dysphagia, dysarthria, dyspnea or weakness.

Results: 247 patients were coded as myasthenia gravis, among which 60 patients presented with OMG and met inclusion criteria; 78% were males, and the average age was 61 years. Thymus abnormalities were present in 13%. Treatment consisted of pyridostigmine alone (48%), prednisone alone or in combination (37%), mycophenolate mofetil alone or in combination (10%). Overall conversion to GMG was 28% (n=17), of which 71% occurred within 2 years, 77% within 3 years, and 23% after 3 years. Of the 17 patients with GMG conversion, 24% were on immunosuppressants, while of the 43 patients without GMG conversion, 49% were on immunosuppressants (statistically insignificant trend toward lower conversion with immunosuppressants, p= 0.06, Fisher exact).

Conclusions: The conversion rate of our treated cohort was lower than previously reported overall rates. Our cohort also included a significant percentage of patients converting to GMG after 3 years.

Keywords: Conversion rate, Ocular myasthenia gravis, Generalized myasthenia gravis

Financial Disclosures: The authors had no disclosures.
Pattern-Reversal Visual Evoked Potential, Visual Field and Nerve Fiber Layer Thickness in Neuromyelitis Optica

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Introduction:
To describe pattern-reversal visual evoked potentials (VEP) in patients with neuromyelitis optica (NMO), and its correlation to visual-field examination (VF) and retinal nerve fiber layer thickness (RNFLt).

Methods:
25 patients with NMO (17 patients had experienced bilateral and 8 unilateral optic neuritis (ON): n=42 ON, and 8 no-ON eyes) were included (4 males, 41.7 ± 13.9 years). Evaluation included best-corrected visual acuity (BCVA), VEP (according to the ISCEV standard), 24-2 VF and optical coherence tomography (OCT) for RNFLt measurement.

Results:
In ON eyes, BCVA ranged from 0.0 logMAR (20/20) to no light perception. A significant correlation was observed between BCVA and RNFLt (r=-0.58; P<0.001). In 50% of ON eyes VEP could not be performed due to low vision, and no-ON eye showed normal VEP amplitude and latency. The remaining 21 ON eyes showed: normal amplitude and latency (29%), prolonged latency with normal amplitude (32%), or reduced amplitude with normal or prolonged latency (35%). Significant correlation between VEP amplitude and the RNFLt was observed (r=-0.414; P=0.026), but no correlation was found between VEP latency and RNFLt (r=-0.279; P=0.136). A significant correlation was also observed between visual field MD and the RNFLt (r=-0.615; P<0.001).

Conclusions:
There is no typical pattern for VEP results in NMO ON, neither for amplitude nor latency. Visual function impairment correlates to RNFLt decrease in NMO ON, as determined for visual acuity, visual field mean sensitivity and VEP amplitude.

Keywords: Neuromyelitis optica, Optic neuritis, Visually evoked potentials, Optical coherence tomography, Visual field test

Financial Disclosures: The authors had no disclosures.
T and B Lymphocytes Abnormalities in Optic Neuritis in China

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Introduction:
To study the role of T and B lymphocytes mediated immunity reaction in pathogenesis of optic neuritis(ON) in China.

Methods:
We examined and compared the expression of the T cell subsets (CD3+, CD4+Th, CD8+Ts) and B cells on peripheral blood mononuclear cells using flow cytometry in 85 patients with optic neuritis (ON) and 30 patients with non-inflammatory neurological system disease (NINSD) in Department of Neurology of Beijing Tongren Hospital in China from May 1st to Sep.30th, 2012. Sub-group analysis was done between neuromyelitis spectrum ON(NMO-S-ON) and multiple sclerosis related ON(MS-ON).

Results:
Total T or B cells abnormalities were seen in 61 cases (71.7%) of ON group and 8 cases (73.3%) of NINSD group. The differences were no significant (P=0.869), even in non-immunity treatment ON patients (P=0.691). The positivity rate of CD3+(67.8%±10.4%) and CD4+Th(38.1%±9.4%) were statistically lower in ON group than those of NINSD patients (CD3+=73.5%±5.9%, CD4+Th=51.0%±9.1%) (P<0.01) while the CD8+Ts(26.1%±8.2%) and B cells(19.5%±8.7%) were higher than NINSD(CD8+Ts =21.6%±6.5%, B cells=14.5%±4.9%) (P<0.01). Further analysis showed a tendency of CD8+Ts cell was higher in NMO-S-ON group than in MS-ON group (37.7%±9.4% vs 23.8±5.8%, P=0.162).

Conclusions:
The increased CD8+Ts and B cell may play an important role in pathogenesis of optic neuritis in China, and is possibly more correlated with the subgroup of NMO spectrum ON.

References:

Keywords: Optic neuritis, T lymphocytes, B lymphocytes, NMO-S-ON, MS-ON

Financial Disclosures: The authors had no disclosures.
Ganglion Cell Layer and Retinal Nerve Fiber Layer Analyses are Both Predictive of Visual Field Recovery Following Resection of Pituitary Adenomas

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Introduction:
Following resection of pituitary adenomas, varying degrees of visual recovery occur ranging from no improvement to complete recovery. Several factors have been identified that are predictive of visual recovery, including pre-surgical retinal nerve fiber layer (RNFL) thickness obtained from optical coherence tomography (OCT). Because decisions on treatment are more influenced by visual function close to and including fixation, ganglion cell layer (GCL) analysis by OCT that encompasses the central six to eight degrees may provide more meaningful clinical information than the RNFL. This study examines the pre-surgical RNFL and GCL, and their ability to predict visual field recovery.

Methods:
A retrospective review of all patients who underwent pituitary adenoma resection and had SD-OCT imaging between 2008 and 2012 identified 32 eyes of 16 patients. Exclusion criteria included prior pituitary adenoma resection, coincident glaucoma, or age younger than 18. Pre-treatment SD-OCT RNFL and GCL, visual acuity, and Goldmann visual fields were analyzed. Post-treatment visual acuity and Goldmann visual fields were obtained, and correlations between recovery and pre-operative RNFL and GCL measurements were analyzed.

Results:
Both the pre-operative RNFL and GCL were significantly reduced in patients with incomplete recovery of the visual field compared to patients with complete recovery (p <0.0001). Patients with more severe residual visual field deficits, as defined by incomplete recovery of the I4e, had significantly thinner RNFL and GCL compared to patients with mild residual visual field defects within the dimmer I2e isopter, but with full recovery of the I4e isopter (p = 0.002 and p=0.02 for RNFL and GCL, respectively).

Conclusions:
Both the pre-treatment RNFL and GCL obtained from SD-OCT are highly predictive of visual outcomes following resection of pituitary adenoma. Pre-operative RNFL and GCL thinning reduces the likelihood of visual field and acuity recovery.

References:


Keywords: Pituitary Adenoma, Bitemporal Hemianopia, Optical Coherence Tomography, Ganglion Cell Layer

Financial Disclosures: The authors had no disclosures.
Longitudinally Extensive Optic Neuritis in Pediatric Patients

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Introduction:
Optic neuritis (ON) may herald the onset of multiple sclerosis (MS), neuromyelitis optica (NMO), or other autoimmune conditions. Extensive optic nerve lesions on MRI may indicate a diagnosis other than MS with worse prognosis, as is the case for extensive spinal cord lesions, which predict NMO. We introduce a definition of longitudinally extensive ON and report its frequency in children with first events of ON ultimately diagnosed with a neuro-immunological disease.

Methods:
This is a retrospective study of patients who first presented with ON and ultimately received neuro-immunological diagnoses at a pediatric MS clinic since its inception in 2006. Those with interpretable brain or orbit MRI scans within 3 months of symptom onset were included. Lesion length, determined by T2 hyperintensity or contrast enhancement, was graded by consensus of two blinded neuro-radiologists as absent, focal or longitudinally extensive (at least two contiguous segments of the optic nerve).

Results:
Twenty-five patients met criteria. Of these 9 (36%) had longitudinally extensive optic neuritis (LEON). One third of MS cases (7 of 20) had LEON. Extensive lesions were not associated with non-MS (NMO, neurosarcoidosis) vs. MS diagnosis (RR 1.18, 95% CI: 0.24, 5.8 p=0.83). No statistically significant association between age of onset and LEON was observed (OR=0.89 per 1 year increase in age, 95% CI: 0.73, 1.09; p=0.25).

Conclusions:
In our cohort, LEON was present in one third of patients with first demyelinating events of ON and was not associated with rare, non-MS neuro-immunological diagnoses. Future studies may clarify whether lesion length associates with visual outcomes in pediatric ON and whether genetic or environmental risk factors affect lesion size and severity.

Keywords: Optic neuritis, Pediatric neuro-ophthalmology, MRI, Multiple sclerosis, Neuromyelitis optica

Financial Disclosures: The authors had no disclosures.
Visualization of Wilbrand's Knee

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Introduction:
In 1904 and 1915, German ophthalmologist Hermann Wilbrand reported that inferonasal crossing fibers within the chiasm curved anteriorly into the contralateral optic nerve. This anatomic bend, known as Wilbrand's knee, is classically cited as the explanation for the contralateral superotemporal visual field defect that may appear when a lesion affects the optic nerve at its junction with the chiasm (the so-called "junctional scotoma"). More recent reports, however, have called into question the existence of Wilbrand's knee or suggested that it may simply be an artifact of monocular enucleation.

Methods:
We studied three post-mortem optic chiasms using a novel optical imaging method, scattering anisotropic imaging (SAI), that allows discrimination of fibers traveling in different orientations and provides information for tractography. After diffusion tensor imaging (DTI) of the fixed post-mortem tissue (7 T x 15 hrs), the chiasm was sectioned into 25 micron slices and mounted on slides. None of the individuals had pre-mortem visual or optic nerve pathology.

Results:
SAI showed that, in the most anterior aspect of the chiasm, crossing fibers followed a U-shape path toward the opposite eye. After curving forward for a distance roughly half the width of the optic nerve, these crossing fibers turned back towards the optic tract. These crossing fibers appear to be organized into thin sheets. Neither fibers that cross more posteriorly nor crossing fibers on the superior aspect of the chiasm demonstrated this forward arcing trajectory.

Conclusions:
This examination of three normal human optic chiasms provides support for the existence of a bend in the path of crossing fibers within the chiasm, consistent with the classical description of Wilbrand's knee. This pattern was only observed for the subset of fibers that cross near the anterior and inferior aspect of the chiasm.

Keywords: Optic chiasm, Wilbrand's knee, Anatomy

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Transient Ocular Motor Nerve Palsies Associated With Cranial Nerve Schwannomas

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Introduction:
Schwannomas, or neuromas, of the cranial nerves are rare, most commonly affecting the vestibular nerve (acoustic neuroma) or trigeminal nerve. Schwannomas of the oculomotor and trochlear nerves are more rare but may occur. Although schwannomas of the abducens nerve itself are extremely rare, the abducens nerve may be affected by schwannomas of the trigeminal nerve. It has been recognized that patients with schwannomas may be either symptomatic or asymptomatic. A few isolated case reports, however, have suggested that symptoms from schwannomas may be transient or recurring.

Methods:
Eight cases of recurrent or transient ocular motor nerve deficits associated with focal, nodular cranial nerve enhancement are described. Clinical features and serial MRI findings were documented during both symptomatic and asymptomatic periods.

Results:
Three cases involved the oculomotor nerve (III), mimicking ophthalmoplegic migraine; four cases involved the trochlear nerve (IV); and two cases involved the trigeminal nerve (V) within Meckel’s cave, leading to palsies of the abducens nerve (VI). In each case, nodular cranial nerve enhancement persisted during both symptomatic and asymptomatic periods that was radiologically most consistent with cranial nerve schwannoma. Only the oculomotor nerve (III) cases were associated with pain.

Conclusions:
This case series suggests that symptoms from schwannomas may be transient or recurring. In each of our cases, the location of the lesions and their persistent nodular enhancement, even when asymptomatic, were most consistent with schwannoma of the oculomotor (III), trochlear (IV) or trigeminal (V) cranial nerves. Normally, the clinical course of schwannoma is one of slow progression but, as our cases demonstrate, it may also be one of remissions and relapses. It may be that these clinical episodes of acute, but transient, ocular motor cranial nerve deficits associated with these relatively small cranial nerve lesions represent episodic inflammation or ischemia – a form of mild “apoplexy” of the schwannoma.

Keywords: Schwannoma, Ophthalmoplegic migraine, Diplopia

Financial Disclosures: The authors had no disclosures.
**Poster 24**

**Pattern of Optic Atrophy in Glaucoma Versus Retrobulbar Optic Neuritis**

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**Introduction:**

Despite different optic nerve appearance, both glaucoma and resolved retrobulbar optic neuritis (ON) have been associated with thinning of the retinal nerve fiber layer and ganglion cell layer by histopathology and optical coherence tomography (OCT). This study aims to characterize the pattern of retinal thinning in these two conditions.

**Methods:**

An OCT retinal analysis software was used to segment and map the macular nerve fiber layer (NFL) and the ganglion-cell/inner-plexiform (GCL/IPL) layer in eyes with optic atrophy due to previous inactive ON (>6 months), glaucoma, and controls. Age, race, visual function (acuity/fields), refractive error, intraocular pressure (IOP), and optic nerve cup/disc and/or pallor were documented. Eyes with glaucoma and those with ON were matched by visual function. Data analyzed included the peripapillary retinal NFL (RNFL), macular NFL, GCL/IPL and outer macular thickness (using ETDRS map).

**Results:**

Twenty-one ON eyes (10 White/11 Black patients) were matched to 21 glaucoma eyes (13 White, 8 Black patients) and 42 controls. With the exception of the outer retinal layers, all OCT values tested (peripapillary RNFL, macular NFL and GCL/IPL), were significantly thinner in both glaucoma and ON compared to controls. For an equivalent average peripapillary RNFL, glaucoma eyes had thicker temporal and nasal RNFL than ON eyes (temporal:51±15μm vs. 41±19μm; nasal:59±17μm vs. 48±24μm, respectively, p<0.05). For the macular map, glaucoma eyes had thicker average inner and outer GCL/IPL thickness than ON eyes (Inner:556±119μm vs. 461±105μm, and Outer:405±66μm vs. 367±67μm, respectively, p<0.04 for both). The ratio of NFL to GCL/IPL was higher in glaucoma compared to ON.

**Conclusions:**

GCL/IPL was atrophied in both glaucoma and ON. Glaucoma eyes had a relatively preserved GCL/IPL thickness compared to ON. This might be related to the mechanism of neuronal injury in these two diseases. Further studies with larger numbers and correlations to histopathology are warranted.

**Keywords:** Optic atrophy, Optical coherence tomography, Retinal segmentation

**Financial Disclosures:** The authors had no disclosures.
Retinal Ganglion Cells Exhibit Spontaneous Hyperactivity and Abnormally Sustained Light-Evoked Responses in in a Mouse Model of Traumatic Brain Injury (TBI)

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Introduction:
We describe striking abnormalities in retinal ganglion cell physiology in an animal model of traumatic brain injury (TBI) that may help explain subtle but persistent abnormalities of vision reported by many TBI victims. These abnormalities precede decreased pattern ERG and morphologic changes we have previously reported in this model.

Methods:
C57BL/6J mice were secured in one side of a two-sided hyperbaric chamber in which the two sub-chambers were separated by a membrane. Air pressure was increased in the opposite chamber until the membrane ruptured, creating an acute, high-speed “blast” wave that impacted the subject. At specified intervals following the blast, in parallel with but preceding decreases in retinal ganglion cell number that we have previously reported, we made multielectrode recordings of ganglion cells in vitro. For each cell, we computed its average spontaneous discharge rate and measured responses to both simple (full field flashes) and complex (random checkerboard) stimuli, comparing with those in retinas from non-blasted control mice.

Results:
One week following blast injury, both spontaneous activity and light-evoked responses bursting were normal. By one month following injury, sustained spontaneous hyperactivity had developed, and a substantial portion of light-evoked responses were prolonged, sometimes outlasting the stimulus. Computed receptive fields were enlarged relative to non-treated retinas.

Conclusions:
After an initial period of normal physiology, dramatic changes in spontaneous and light-driven ganglion cell activity occur with a several-week latency following blast injury in this mouse model of mild-moderate TBI. The increased spontaneous activity and prolonged responses to light may be interpreted as “noise” in the retina’s message to the brain, which leads to abnormal visual perception in victims of TBI such as encountered in the modern battlefield. These findings may help develop additional means to diagnose and effectively treat these previously elusive visual distortions in TBI patients.

Keywords: Traumatic brain injury, Retinal ganglion cell, Visual distortion, Neural code for vision

Financial Disclosures: The authors had no disclosures.
Introduction:
There is controversy as to whether conversion disorders such as hysterical blindness should be treated with reassurance alone, placebo, or active therapies. In 1918, Babinski reported that neurologic conversion disorders such as functional vision loss could be treated with pithiatism, or persuasion. Accordingly, we suspected that treatment of our hysterically blind patients with persuasive techniques, a form of active therapy, would result in positive outcomes. Because there are no guidelines for management of persons with hysterical blindness, more data regarding outcomes such as these are needed.

Methods:
A review of eight consecutive patients who presented with acute hysterical blindness between 8/09 and 1/12. Patients who had co-morbid organic visual loss, neurologic disease, or a secondary gain such as disability reimbursements were not included. After initial neuro-ophthalmologic evaluation was complete and hysterical blindness was diagnosed, each patient was given a 20/200 optotype ‘Tumbling E’ card and encouraged that vision would improve by reading the card at increasing viewing distances daily, until 20 feet was reached.

Results:
Age range was 11-30 with mean of 17 years. Seven of eight patients were female. Seven had both acuity and field defects; one had a visual field defect only. Six had binocular defects, two had monocular vision loss. Three cases were associated with recent trauma. There were associations with bipolar and histrionic diagnoses, abusive relationships, recurrent childhood abdominal pain, and prior functional behaviors. In all eight cases, patients reported that they used the described technique, and confirmed that vision improved over time, over a range of two weeks to six months. There were no adverse events related to treatment.

Conclusions:
Treatment with persuasion techniques such as ‘vision expanding exercises’ can be a safe and effective management strategy for hysterical blindness.

References:

Keywords: Hysterical blindess, Functional vision loss, Conversion disorders

Financial Disclosures: The authors had no disclosures.
Demographic Associations With Optic Neuritis Outcomes: An Analysis of the Optic Neuritis Treatment Trial

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Introduction:
Poor visual recovery after optic neuritis is an important morbidity that affects activities of daily living and quality of life. Retrospective studies in the US, UK and South Africa have demonstrated disparate outcomes in individuals of African descent compared with Caucasians. However, published analyses of the prospectively collected Optic Neuritis Treatment Trial (ONTT) data did not find visual outcomes to be associated with race and ethnicity. We sought to further investigate demographic associations with visual outcomes following optic neuritis through application of longitudinal data analysis techniques to the ONTT data set.

Methods:
We analyzed prospectively collected data on black (n=58) and white (n=388) subjects presenting with unilateral acute optic neuritis who enrolled in the ONTT. Mixed effects regression techniques were used to model visual contrast sensitivity in the affected eye (CS-AFF) as a continuous outcome. All available follow up data (to a maximum of 15 years) were included. Models included effects of treatment (placebo, oral prednisone, IV methylprednisolone), time and treatment-time interaction as well as covariates of age, gender and race.

Results:
In the baseline model there were no significant effects of treatment on CS-AFF. There were significant interaction effects of treatment over time on CS-AFF (oral vs. placebo p=0.02, IV vs. placebo p=0.04). Neither age (p=0.25) nor gender (p=0.26) were significantly related to CS-AFF. Race was significantly related to CS-AFF (p=0.01) with black race being associated with worse scores for CS-AFF. The race effect persisted in a model that incorporated age, gender and race (p=0.008).

Conclusions:
We find race, but not gender or age to be significantly associated with contrast sensitivity in the affected eye following optic neuritis in an analysis of the ONTT. To our knowledge this is the first evidence from a prospectively collected data set to support a hypothesis of race dependent outcomes of optic neuritis. It is largest cohort of black subjects with optic neuritis to be studied.

References:
2. Pokroy, Modi, Saffer, Optic neuritis in an urban black African community, Eye, 15, 469, 2001

Keywords: Optic neuritis, Race, Epidemiology

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Anatomy and Image Correlation of Subjective Visual Vertical in Patients With Acute Brainstem Lesions

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Introduction:
Unilateral damage of the graviceptive pathways causes ocular tilt reaction (OTR) including tilt of the subjective visual vertical (SVV), ocular torsion, and skew deviation. The aim of this study was to analyze the topology of ischemic brainstem lesions generating pathological tilt of the SVV.

Methods:
We examined 28 patients with acute brainstem infarctions for tilt of otolith-ocular dysfunction. Lesions localized to the brainstem were identified by MRI and their subjective visual vertical (SVV) and ocular vestibular-evoked myogenic potentials (oVEMPs) during acute period of each patient were tested. Individualized brainstem lesions were analyzed by means of MRI-based voxel-wise lesion-behavior mapping, and the probabilistic lesion maps were constructed.

Results:
Most patients exhibited significant tilts of the internal representation of the gravity vector as deviated ipsiversive or contraversive tilt of subjective visual vertical. Patients with ipsiversive tilt of one or both eyes mostly had lesions at anteromedian pontomesencephalic and below the pons such as infarction at the lateral medulla. In contrast, patients with contraversive tilt of SVV had lesions mainly at dorsomedial pons and dorsomedial midbrain region.

Conclusions:
We hypothesize that both ipsilateral and contralateral graviceptive projections may explain the observations of ipsilesional or contralesional SVV deviation in patients with brainstem infarction.

Keywords: Subjective visual vertical (SVV), Otoliths, Brainstem, Ocular tilt reaction

Financial Disclosures: The authors had no disclosures.
Müller Cell Function in Neuromyelitis Optica as Measured by Electrophysiologic Testing

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Introduction:
Neuromyelitis optica (NMO) is an inflammatory demyelinating disorder of the central nervous system [1]. Antibodies against the astrocyte water channel protein aquaporin 4 (AQP4) have been implicated in the pathogenesis of NMO [2]. AQP4 has been found in the optic nerve as well as on Müller cells in the retina [3,4]. Patients who develop NMO-associated optic neuritis have a poorer prognosis for recovery with greater loss of retinal ganglion cell axons [5]. It is unclear if the visual prognosis in NMO is secondary to damage to the optic nerve alone or in combination with damage to Müller cells with subsequent retinal dysfunction. Intact Müller cell function may be reflected by a positive B-wave during ERG testing. This study was designed to evaluate the function of Müller cells as measured by electroretinography (ERG) testing in NMO patients.

Methods:
This was a retrospective study of patients with a known diagnosis of NMO and poor visual recovery after prior optic neuritis who underwent electrophysiologic testing.

Results:
Five patients with NMO-related optic neuritis and poor visual recovery underwent electrophysiologic testing. All patients were positive for anti-NMO AQP4 antibodies and were female. A total of 8 eyes had one or more prior episodes of optic neuritis with visual acuity of counting fingers or less. In all eight eyes, ERG B-wave was normal. In eyes with prior optic neuritis tested with visual evoked potential (VEP), there was severely increased latency and decreased amplitude. In eyes with no history of clinical optic neuritis, the ERG B-wave was normal and VEP showed normal latency and amplitude.

Conclusions:
Though AQP4 receptors are present on Müller cells, a preserved B-wave on electroretinography in eyes of NMO patients suggests that Müller cell function remains intact. VEP testing was abnormal as expected in eyes with prior optic neuritis and poor visual recovery in NMO patients.

References:

Keywords: Neuromyelitis optica, Aquaporin 4, Müller cell, Electrophysiology, Optic neuritis

Financial Disclosures: The authors had no disclosures.
Enhanced Depth Imaging Optical Coherence Tomography of Optic Nerve Head Drusen

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Introduction:
The usefulness of enhanced depth imaging optical coherence tomography (EDI OCT) for diagnosing and evaluating optic nerve head drusen (ONHD) was compared to conventional diagnostic methods.

Methods:
In this prospective, comparative, cross-sectional study, 34 patients with obvious or suspected ONHD in either eye were enrolled. Spectral-domain OCT of the optic nerve head in both conventional (non-EDI) and EDI modes, ultrasound B-scan, and standard automated perimetry (SAP) were performed for both eyes of all participants.

Results:
Eyes were clinically classified into 3 groups: 32 eyes with definite ONHD, 25 eyes with suspected ONHD and 11 normal-appearing fellow eyes. In the definite ONHD group, EDI OCT, non-EDI OCT, and ultrasound B-scan were positive for ONHD in all eyes, and VF was abnormal in 24 eyes. In the suspected ONHD group, 8 eyes had no evidence of ONHD in each test, and EDI OCT, non-EDI OCT, ultrasound B-scan, and visual field were positive in 17, 14, 7, and 3 eyes, respectively. In normal-appearing fellow eyes, 4 eyes had no evidence of ONHD in each test, and EDI OCT, non-EDI OCT, ultrasound B-scan, and visual field were positive in 17, 14, 7, and 3 eyes, respectively. In normal-appearing fellow eyes, 4 eyes had no evidence of ONHD in each test, and EDI OCT, non-EDI OCT, ultrasound B-scan, and visual field were positive in 17, 14, 7, and 3 eyes, respectively. EDI OCT had a significantly higher ONHD detection rate than ultrasound B-scan in all eyes (52/68 eyes vs. 40/68 eyes; p<0.001), in eyes with clinically suspected ONHD or normal-appearing fellow eyes (20/36 eyes vs. 8/36 eyes; p<0.001), and in eyes with clinically suspected ONHD (17/25 eyes vs. 7/25 eyes; p=0.002). EDI OCT detected ONHD mass as signal-poor regions surrounded by short, hyper-reflective bands or isolated/clustered hyper-reflective bands without a signal-poor core. In non-EDI OCT, posterior surfaces of the ONHD and deep-seated hyper-reflective bands were invisible or less clear than in EDI OCT.

Conclusions:
EDI OCT may be more useful in diagnosing ONHD and assessing their structures than conventional tests.

Keywords: Optic nerve head drusen, Optical coherence tomography, Enhanced definition imaging, Ultrasound B-scan

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Poster 31

Retinal Ganglion Cell Layer Thickness in Children with Vision Loss from Optic Pathway Gliomas

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Introduction:
Accurate and reliable vision testing in young children with optic pathway gliomas (OPGs) can be limited by their cooperation. Since not all OPGs cause vision loss, a surrogate marker of visual pathway integrity that does not require cooperation would be helpful. Therefore, we explored whether structural measures of the macular ganglion cell layer-inner plexiform layer (GCL-IPL) using spectral-domain optical coherence tomography (SD-OCT) were related to visual acuity (VA) and visual field (VF) in children with OPG.

Methods:
Children with OPGs (sporadic or secondary to Neurofibromatosis type 1, NF1) enrolled in a prospective study of SD-OCT were analyzed. Enrolled subjects were included only if macular SD-OCT images were acquired and they were cooperative for VA and VF testing. Manual segmentation of the macular GCL-IPL was performed across an ETDRS grid at 1.5, 3.0 and 4.5mm diameter. GCL-IPL thickness measures (microns) were compared to VA and VF outcomes.

Results:
Forty-seven study eyes from 26 children with OPGs (18 NF1-related, 8 sporadic) were included. Median age was 5.3 years (range, 2.5 – 12.8). The GCL-IPL thickness (3.0mm diameter) was decreased in the abnormal VA (61 ± 2 microns), abnormal VF (73 ± 11 microns) and abnormal VA/VF (53 ± 5 microns) groups when compared to the normal VA/VF group (92 ± 9 microns; F = 44.9, p <0.001). GCL-IPL thickness in the abnormal VA group was not different from the abnormal VF (Post-hoc Scheffe’s test, p = .28) and abnormal VA/VF groups (p = 0.67), however the abnormal VA/VF group had a lower GCL-IPL than the abnormal VF group (p <0.01).

Conclusions:
Children with vision loss (VA, VF or both) from their OPG have decreased GCL-IPL thickness compared to those children with normal vision. GCL-IPL could be used as a surrogate marker of vision in children with OPGs.

Keywords: Retinal ganglion cells, Optical coherence tomography, Optic pathway glioma, Children

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Poster 32

Meningoceles in Idiopathic Intracranial Hypertension (IIH)

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Introduction:
Acquired skull base meningoceles can be seen in patients with chronic intracranial hypertension and are sometimes observed in IIH patients. Our aim was to evaluate the frequency of meningoceles among IIH patients and to determine their association with disease severity and outcome.

Methods:
Retrospective case-control study of consecutive IIH patients and controls seen between 2000-2011 with axial and coronal MRI T2 series performed at our institution. IIH patients’ demographics, symptoms/signs of IIH, CSF-opening pressure, visual outcome and treatments were collected. Controls included patients without headache or visual complaints, who had an MRI read as normal (meningoceles are not routinely evaluated) [reasons for MRI included sensory disturbances, peripheral neuropathies, seizure, and metastatic screening]. A neuroradiologist reviewed all imaging studies for meningoceles and enlargement of Meckel’s cave. Wilcoxson Rank Sum Test and Fisher Exact Test were used for statistical comparisons.

Results:
We included 79 IIH patients [median age 28 years; range:24-36; 77 women (97%); BMI 40 kg/m², range:33-43] and 76 controls [median age 33 years, range:30-40; 62 women (86%)]. 9/79 (11%) IIH patients had meningoceles (Meckel’s cave or petrous apex) and 7/79 (9%) had enlargement of Meckel’s cave, compared to none of the controls (p<0.003). No anterior fossa meningoceles were found. Among IIH patients, the presence of meningoceles or Meckel’s cave enlargement was not associated with their demographics, symptoms, degree of papilledema, visual acuity (median:20/25 vs. 20/20;p=0.42), or visual field defect severity (median HVF MD:-3.6 vs. -3.4;p=0.94), CSF opening pressure, or need for surgical treatment.

Conclusions:
Meningoceles and Meckel’s cave enlargement are more common among IIH patients than controls, likely secondary to chronically elevated ICP. However, they are relatively rare (around 10%) and their presence or absence does not seem to influence the prognosis of IIH, and therefore should have no therapeutic implications.

References:

Keywords: Idiopathic intracranial hypertension, Meningoceles, Radiology

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Uveitis With Optic Disc Edema Secondary to Concomitant Idiopathic Intracranial Hypertension

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Introduction:
Optic disc edema has been reported in association with anterior uveitis; resolution of uveitic optic disc edema has been demonstrated with treatment of uveitis. We describe 5 patients with uveitis and optic disc edema found to have concomitant idiopathic intracranial hypertension (IIH) requiring treatment.

Methods:
A retrospective review of patients presenting from January 2007 through September 2012 with uveitis and concomitant optic disc edema. Data was collected regarding patient demographics, symptoms, clinical findings, work-up, management, and outcomes.

Results:
Of 24 patients identified to have uveitis and concomitant optic disc edema, six (25%) had an elevated opening pressure (OP) on lumbar puncture (LP), of which five (21%) were diagnosed with IIH, while one (4%) was diagnosed with neurosarcoidosis. All 5 IIH patients were female with mean age of 37.6 years. Mean BMI was 38.9, with 3/5 patients reporting significant weight gain prior to the development of papilledema. All 5 patients reported symptoms consistent with IIH including: headaches (4/5 patients), pulsatile tinnitus (3/5), diplopia (1/5), and transient visual obscurations (1/5). All 5 patients had history of bilateral uveitis, 2 with anterior and 3 with panuveitis. In 3/5 patients, the uveitis preceded papilledema, though 1 patient had no active uveitis at the time of IIH diagnosis. In 2/5 patients, IIH was diagnosed prior to the development of uveitis, and in 1/5 patients, the diagnoses were made concurrently. Average OP was 32.3 with normal cerebrospinal fluid analysis. All patients were treated medically for IIH with weight loss and acetazolamide. In 2/5 patients, symptoms and papilledema resolved, while 3/5 are followed on medical treatment.

Conclusions:
In addition to uveitic disc edema, diagnostic considerations in a patient with uveitis and optic disc edema should include IIH, taking into account patient demographics, history of recent weight gain, and symptoms. If suspected, appropriate work-up and treatment for IIH should be initiated.

Keywords: Optic Disc Edema, Idiopathic Intracranial Hypertension, Uveitis, Papilledema

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Pattern Electroretinogram for Detecting Retinal Neural Loss in Neuromyelitis Optica and Multiple Sclerosis Patients With or Without Optic Neuritis. Correlation With OCT and Automated Perimetry

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Introduction:
Multiple sclerosis (MS) and neuromyelitis optica (NMO) are related diseases that typically affect the anterior visual pathway leading to visual loss. The quantification of axonal loss in both MS and NMO has received great attention with OCT but may also be assessed by pattern electroretinography (PERG). The purpose of this study is to evaluate the ability of transient PERG parameters to differentiate between eyes of patients with NMO, longitudinally extensive transverse myelitis (LETM), MS with (MS+ON) and without optic neuritis (MS-ON) and controls, and to assess the relationship between PERG amplitude and OCT or standard automated perimetry.

Methods:
Visual fields (VF) and full-field stimulation PERGs using both 48 and 14 min. stimuli were obtained from patients with MS (n=28), NMO (n=20), (LETM) (n=18) and healthy controls (n=26). In addition, retinal nerve fiber layer (RNFL) thickness was measured from peripapillary OCT scans of the optic disc and retinal ganglion cell plus inner plexiform layer (RGCL+) and total retinal thickness of the macula were obtained from macular cube scans. Correlations between PERG amplitude and OCT or standard automated perimetry were investigated.

Results:
Compared to controls, PERG amplitude measurements were significantly reduced in NMO and MS+ON eyes, but not in MS-ON and LETM eyes. Abnormalities were most evident in N95 amplitude measurements. The PERG amplitudes in NMO and MS+ON eyes were significantly smaller when compared to MS-ON eyes. A significant correlation was found between N95 amplitude measurements and OCT-measured macular NFL ($R^2=0.24$), RGCL+ ($R^2=0.25$), total retinal ($R^2=0.25$) thickness as with peripapillary RNFL ($R^2=0.16$) thickness in the temporal disc sector. PERG amplitude was significantly associated with VF sensitivity loss.

Conclusions:
PERG measurements were abnormal both in MS+ON and NMO eyes, but we were not able to differentiate between the two conditions. PERG amplitude measurements correlate well with OCT-measured parameters, suggesting that the technologies can be complementary for quantifying axonal loss in NMO and MS.

Keywords: Pattern electroretinography, Optical coherence tomography, Multiple sclerosis, Neuromyelities optica

Financial Disclosures: The authors had no disclosures.
Poster 35

Ventriculoperitoneal Shunts as a Treatment in Idiopathic Intracranial Hypertension

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Introduction:
Ventriculoperitoneal (VP) shunt is a treatment for visual loss and headache in idiopathic intracranial hypertension (IIH). The current investigation evaluates outcomes of IIH patients who underwent VP shunts in a single institution.

Methods:
A retrospective chart review was performed of all new IIH patients first evaluated in our eye institution over an 11 year period (2000-2010). Patients who underwent VP shunt at our medical center were included.

Results:
Of 425 new patients with IIH, 19 (5%) met inclusion criteria. The mean age was 29±13 years, and one (5%) patient was male. Seven patients had surgery before VP shunt: 6 had an optic nerve sheath fenestration performed in one or both eyes, and 1 patient had a failed lumboperitoneal shunt elsewhere. All 19 patients had VP shunts placed between 12/2006 to 8/2011, and the average follow up was 646 days (range 5-1342). Median baseline visual acuity was 20/50 OD and 20/45 OS (range 20/15 to NLP). Median acuity 18 months after VP shunt was available in 8 patients and was 20/25 OD and 20/40 OS (range 20/20 to hand motion); mean improvement was 0.47±0.36 logMAR OD and 0.24±0.16 logMAR OS. Mean HVF improvement was from -18.5 dB to -13.0 dB with 65% of the patients experiencing improvement. Kaplan-Meier analysis showed 62% of VP shunts were functioning at 18 months without replacement, removal, or revision but may have had an adjustment; 2 additional patients subsequently required shunt revisions (at 27 and 38 months).

Conclusions:
VP shunts usually improve vision in IIH, but come at a price of moderately high complication rates over time which necessitates close monitoring of patients’ shunt function.

References:

Keywords: Idiopathic intracranial hypertension, Ventriculoperitoneal shunt

Financial Disclosures: The authors had no disclosures.
Optic Nerve Sheath Decompression: A Surgical Technique With Minimal Operative Complications

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Introduction:
The purpose of this study was to determine the safety and efficacy of optic nerve sheath decompression (ONSD) with a medial transconjunctival approach for a variety of indications in a larger population of patients that has previously been reported.

Methods:
A retrospective chart review was performed on consecutive patients who underwent ONSD between January 1992 and December 2010. Prior to ONSD, all patients had documented evidence of progressive loss of visual acuity or visual field, or both. Postoperative follow-up visits were scheduled at 1 week, 1 month, and then every 3 to 6 months. Main outcome measures were visual acuity, visual fields, and surgical complications.

Results:
Five hundred seventy-eight eyes of 331 patients underwent ONSD for progressive vision loss due to various indications, which included but were not limited to IIH, progressive non-arteritic ischemic optic neuropathy (pNAION), and optic nerve drusen (OND). During a mean follow-up of 18.7 months (range, 1 week - 10 years), postoperative visual acuity remained stable or improved in 536 of 568 eyes (94.4%) and progressively worsened in 32 of 568 eyes (5.6%). Visual fields remained stable or improved in 257 of 268 eyes (95.9%) and progressive visual field loss occurred in 11 of 268 eyes (4.1%). There were no reported intra-operative complications. The most common postoperative complication involved motility disturbance (6.0%).

Conclusions:
To our knowledge, this review represents the largest series of patients who have undergone ONSD for any indication. Our data is consistent with current literature supporting ONSD as a safe and effective procedure for IIH. Despite the small sample sizes, this study confirms previous reports that ONSD provides no benefit for pNAION, but suggests ONSD might remain a therapeutic consideration for halting progressive visual field loss associated with OND. Regardless of the indication, complications following ONSD with the technique described in this review are infrequent.

Keywords: Optic Nerve Sheath Decompression Safe

Financial Disclosures: The authors had no disclosures.
Occult Pancraniosynostosis Masquerading as Idiopathic Intracranial Hypertension in a Normocephalic Patient during Late Childhood

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Introduction:
Craniosynostosis refers to premature fusion of a cranial suture, potentially causing characteristic deformations of the skull in a pattern originally described by Virchow. Often, craniosynostosis presents early in infancy or childhood due to anomalous skull shape, associated facial anomalies, developmental delay, or signs and symptoms of neurologic impairment or elevated intracranial pressure (ICP). Here we report a case of secondary occult pancraniosynostosis masquerading as idiopathic intracranial hypertension (IIH) in 10-year-old girl with a history of Graves Disease and otherwise normal development, resulting in rapid and severe vision loss. The purpose of this case report and search of relevant literature is to determine if occult craniosynostosis is an under-recognized cause of secondary intracranial hypertension or altogether misdiagnosed as IIH, leading to delayed diagnosis and visual impairment.

Methods:
A PubMed and MEDLINE literature search was performed using the following words alone or in combination: "craniosynostosis", "craniosynostoses", "synostosis", "pancraniosynostosis", "mild", "occult", "intracranial pressure", "intracranial hypertension", "pseudotumor cerebri", "hyperthyroidism", "hypothyroidism", "Graves Disease".

Results:
A search of PubMed and MEDLINE literature yielded 6 publications of case series describing late diagnosis of craniosynostosis. The maximum age at presentation of these cases ranged from age 4 to 13. Elevated intracranial pressure (ICP) is a well-known sequela of craniosynostosis. Common features that this case shares with cases reported in the literature of late diagnosis of craniosynostosis include: 1) normocephalic configuration, 2) absence of prior signs or symptoms of elevated ICP, 3) acute onset of signs and symptoms of elevated ICP, 4) rapid progression, and 5) visual impairment.

Conclusions:
Craniosynostosis in normocephalic patients can present late in childhood as an acute episode of elevated intracranial hypertension, which can be misdiagnosed as IIH. Careful review of radiologic images for signs of premature suture fusion and clinical suspicion must be employed to identify occult craniosynostosis in order to prevent rapid progression to severe vision loss.

References:


Keywords: Childhood Idiopathic intracranial hypertension, Craniosynostosis, Pancraniosynostosis

Financial Disclosures: The authors had no disclosures.
Cerebrovascular Manifestations in Giant Cell Arteritis

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Introduction:
Giant cell arteritis (GCA) can present in multiple ways, among the most serious of which are cranial ischemic complications (CIC). The majority of CICs involve the ocular circulation; however, less commonly present with cerebrovascular ischemia which may lead to delayed or missed diagnosis. Our study aims to characterize the incidence of cerebrovascular ischemia as a presenting manifestation of GCA, as well as identify demographic, clinical or serological features within this cohort of patients, with the potential to predict which GCA patients may be at higher risk of developing intracranial involvement.

Methods:
We performed a retrospective chart review of all patients under the care of a single physician, who underwent temporal artery biopsy from August 1, 2007 to August 31, 2011 in a Canadian tertiary care institution and identified patients within this cohort who presented with or developed cerebrovascular ischemia. Demographic, clinical, and serological data were gathered and analyzed.

Results:
During the study period, 95 consecutive patients with biopsy-proven GCA were identified, of which 9 (9.5%) experienced cerebrovascular ischemia. Six patients presented with vasculitic involvement of anterior and posterior circulations, while 3 patients experienced an ischemic event confined to the vertebrobasilar territory. There was no significant difference between patients with and without stroke in terms of age (76.2, 74.2, p=0.49), ESR (48, 52, p=0.87), CRP (42.4, 44.8, p=0.91), and platelet count (322, 344, p=0.36). Features such as male gender, headache, scalp tenderness, visual disturbance, and jaw claudication were not predictive of CVA in GCA patients.

Conclusions:
No single demographic, clinical, or serological feature was shown to predict the occurrence of cerebrovascular manifestations in GCA patients. A high index of suspicion for GCA needs to be maintained for patients presenting stroke symptoms, as well as awareness of potential CVA in GCA patients. This paper describes the varied neurological presentations of our patients.

Keywords: Giant Cell Arteritis, Stroke, Temporal Arteritis, cranial ischemic complications

Financial Disclosures: The authors had no disclosures.
Magnetic Resonance Diffusion-Weighted Imaging of the Optic Nerve Head in Papilledema

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Introduction:
Increased intracranial pressure (ICP) may lead to characteristic neuroimaging signs, which have been previously described.¹ Although isolated restricted diffusion of the optic nerve head (ONH) in the setting of papilledema has been described,² it has not been evaluated as a predictor of papilledema, nor as a prognostic indicator of visual outcome.

Methods:
More than 100 pediatric cases with neuroimaging signs of elevated ICP (ie, protrusion, enhancement, or restricted diffusion of the ONH; dilation of the optic nerve sheath), or MRIs performed for an indication of papilledema were collected. A retrospective chart review was performed to confirm the ophthalmoscopic evidence of papilledema, diagnosis, and visual acuities and visual fields at presentation (at the time of the MRI) and at the most recent follow-up.

Results:
We will evaluate the ability of restricted diffusion of the ONH to diagnose papilledema (ie, sensitivity and specificity), and see if certain etiologies may predispose patients to this neuroimaging sign. We will compare initial vision and final visual outcomes in patients with papilledema and restricted diffusion to patients with papilledema and no restricted diffusion, to see whether this MRI sign has short-term predictive or long-term prognostic value, respectively. Of the first 10 patients analyzed with restricted diffusion, all had clinical papilledema from posterior fossa tumors, pseudotumor cerebri, cerebral venous thrombosis (CVT), and meningitis. Nine out of 10 patients had initial acuities of 20/40 or better and final acuities of 20/32 or better (patient with CVT had poor initial and final visual function).

Conclusions:
Some preliminary conclusions regarding restricted diffusion of the ONH include: 1) it is not found in pseudopapilledema, 2) it is highly suggestive of papilledema, 3) visual outcomes are favorable, 4) it does not seem to be etiology-dependent, 5) it is commonly seen with other neuroimaging signs of increased ICP.

References:

Keywords: Diffusion-weighted imaging, Papilledema

Financial Disclosures: Dr. Liu has served as a consultant for Ipsen. No other authors had disclosures.
The Orbital Volume Measurement in Patients With Ventriculoperitoneal Shunt

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Introduction:
Ventriculoperitoneal shunt (VPS) operation is a surgical approach for treatment of increased intracranial pressure. There were reports of patients with congenital hydrocephalus who developed bilateral enophthalmos after ventriculoperitoneal shunt. We analyzed orbital volume before and after shunt operation in patients underwent VP shunt.

Methods:
Orbital volumetric analysis was performed on 30 patients (mean age 58 years, 16 male and 14 female patients) who underwent ventriculoperitoneal shunt with no evidences of orbital fractures or maxillary lesions. ITK-snap(version 2.0 software) was used to analyze orbital CT axial images by measuring the orbital area. The calculated orbital volumes at pre-shunt were compared with post-shunt using the Wilcoxon matched-pairs signed rank test. (GraphPad Software, San Diego, CA).

Results:
There was no case of enophthalmos or related symptoms in all patients. The average follow up period was 8.9 months after initial shunt. Pre-shunt mean orbital volume were 23.9cm³ (right), and 24.6cm³ (left), which was not statistically significantly different from post-shunt mean 23.6cm³ (right), and 24.1cm³ (left) (P=0.5959). However, the orbit volume was significantly increased in 4 patients (8 eyes) who were followed for more than 23 months after shunting. (P= 0.0156).

Conclusions:
Previously suggested mechanisms for enophthalmos and increased orbital volume was intracranial hypotension-related skull remodeling. In this study, the increased orbital volume can only be observed in patients underwent VPS for more than 23 months. We should consider the possibility of secondary enophthalmos with long term intracranial hypotension.

References:

Keywords: Orbital volume, Ventriculoperitoneal shunt, Enophthalmos

Financial Disclosures: The authors had no disclosures.
Abnormal Thickness of Ganglion Cell-Inner Plexiform Layer in Patients with Alzheimer's Disease and Mild Cognitive Impairment

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Introduction:
A depletion of retinal ganglion cells and axons has been histologically observed in Alzheimer’s disease patients.¹ Previous studies showed that retinal nerve fiber layer (RNFL) is reduced in Alzheimer’s Disease (AD) and mild cognitive impairment (MCI) patients.²⁻⁴ The purpose of this study is to find the differences of Macular and Ganglion Cell-Inner Plexiform Layer thickness in AD, MCI, and control subjects.

Methods:
The patients compatible with the diagnostic criteria of AD(n=10), MCI(n=5) and age-matched control(n=10) subjects have been evaluated neurologic, ophthalmologic examination and measured the average ganglion cell-inner plexiform layer, macular and retinal nerve fiber layer (RNFL) thickness by optical coherence tomography.

Results:
Mean average ganglion cell-inner plexiform layer thickness was 63.60±14.00µm (AD), 71.44±14.45µm (MCI), 78.20±11.09µm (Control). The correlation in thickness of average ganglion cell-inner plexiform layer (p=0.003) and RNFL thickness (p<0.05) among 3 groups were statistically significant. But, there was no significant correlation in macular thickness.

Conclusions:
The decreased thickness of ganglion cell-inner plexiform layer in patients with AD suggest the thickness of ganglion cell-inner plexiform layer could be helpful for diagnosis of AD and MCI.

References:

Keywords: Alzheimer’s disease, Ganglion Cell Layer, Optical coherence tomography

Financial Disclosures: The authors had no disclosures.
Visual Outcomes of the Surgical Treatment of Idiopathic Intracranial Hypertension

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Introduction:
Surgical treatment of increased intracranial pressure is performed for severe visual loss, intractable headache, or both, usually after failed medical treatment. Options include optic nerve sheath fenestration and CSF diversion. Both procedures are felt to be effective, but no comparative data exist. The objective of this study is to evaluate the visual outcomes in surgically treated IIH patients.

Methods:
Retrospective observational case series in which the medical records of patients with severe idiopathic intracranial hypertension who underwent surgical treatment at our institution were reviewed for demographics and clinical findings. Patients who underwent ONSF were compared to shunted patients.

Results:
Thirty-eight patients (35 women and 3 men) with a mean age of 30.3 years (range, 9-52) underwent surgical treatment of IIH (16 ONSF, 22 shunt). Pre-operative Frisen scale papilledema in the ONSF group was grade 3 (14.3%), grade 4 (71.4%) and grade 5 (14.3%). In the shunt group 27.8% of patients had pre-operative optic atrophy, grade 1 (16.7%), grade 2 (33.3%), grade 3 (16.7%) and grade 4 (5.5%) swelling. Pre-operative visual acuity (VA) was 60.6 ETDRS equivalent letters in the ONSF group and 73.9 letters in the shunt group (p=0.13). There was a trend towards worse pre-operative mean deviation in the ONSF group (-22.94 dB vs -14.86 db, p=0.09). At the last follow-up mean VA was 75.3 letters in the ONSF group and 77.0 letters in the shunt group (p=0.56). Perimetric mean deviation was worse after ONSF (-17.36 dB vs -8.78 dB, p<0.05).

Conclusions:
Both ONSF and shunting are effective procedures in stabilizing or improving visual acuity and visual fields in IIH patients. In general there was a trend towards worse pre-operative visual function that could explain the worse visual fields outcomes after ONSF. Prospective comparison of these procedures is needed.

Keywords: Idiopathic intracranial hypertension, Papilledema, Visual outcomes, Optic nerve sheath fenestration, Cerebrospinal fluid diversion

Financial Disclosures: The authors had no disclosures.
Introduction:
Patients with neuro-ophthalmologic disease often have laterally incomitant strabismus. The study purpose was to determine the effect of monocular strabismus surgery on lateral incomitance in patients with exotropia.

Methods:
Prospective evaluation of patients > 7 years old with exotropia who had unilateral horizontal rectus muscle surgery between 12/09 and 1/12. Prism and alternate cover testing was performed with distance fixation in primary position, right gaze, and left gaze after > 45 minutes of monocular occlusion, within one week prior to surgery, within one week after surgery, and > 3 months after surgery. The surgical procedure was at the discretion of the surgeon. Patients with extraocular muscle paralysis, fibrosis or trauma were excluded. The change in deviation induced by strabismus surgery in lateral gaze was expressed as a percentage of the change in deviation induced in primary position.

Results:
Fourteen patients with an age range of 17-74 years and a preoperative exotropia of 12-85 prism diopters met inclusion criteria. Nine patients had unilateral recessions/resections, three had unilateral lateral rectus recessions, and two had unilateral medial rectus resections. Thirteen patients (93%) had greater effect from strabismus surgery with gaze towards the operated eye (p=0.0013). On average, the surgical effect in gaze towards the operated eye was 148% that achieved in primary position whereas the surgical effect in gaze away from the operated eye was 82%. Ten patients had >3 months follow-up. Nine of these patients (90%) had greater surgical effect with gaze towards the operated eye (p=0.0114). On average, the surgical effect in gaze towards the operated eye was 121% that achieved in primary position whereas the surgical effect in gaze away from the operated eye was 85%.

Conclusions:
Unilateral strabismus surgery induces clinically significant lateral incomitance in patients with exotropia. The lateral incomitance is reduced, but still remains 3 months after surgery.

Keywords: Strabismus surgery, Exotropia, Lateral incomitance

Financial Disclosures: The authors had no disclosures.
Enhanced Image Guided Endoscopic Orbital Surgery Technique

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Introduction:
Retrobulbar access, specifically treatment to the optic nerve, is limited by the orbital anatomy. We have developed a minimally invasive, endoscopic procedure for optic nerve treatment. This approach is based on an electromagnetic image guidance system (IGS). In this experiment, we compared performance with and without an enhanced or augmented reality, modification of the system.

Methods:
Twelve orbital interventions were performed in six live pigs. Following an Institutional Animal Care and Use Committee approved protocol, the pigs were anesthetized, A matched set of microspherical bulbs were placed in each orbit. One bulb held pure water, the other clear magnevist. Red dye was also placed to either the water or the magnevist bulb to make it have a color. Fiducial markers were applied and magnetic resonance imaging (MRI) performed. The bulbs had opposite image intensity and in surgical observation the bulbs were distinct. The surgeon reviewed the pre-operative image set and used the IGS to locate the target bulb. The surgeon identified the color of the target. In the first 3 pigs the system was used without the enhanced guidance. In the last 3 pigs the video display module was modified to have an overlay of a 2D/3D registration enhancement which designated the location of the target.

Results:
The correct target was identified in all procedures. With the standard IGS, the average time to find and identify the target was 20 minutes. With the enhanced IGS, time to target was reduced to 3 minutes.

Conclusions:
The enhanced IGS afforded a substantial decrease in procedure time. Augmented reality has been demonstrated in rigidly tracked endoscopes. This is first proof of concept that endoscopic enhancement, which helps to provide a geographic anatomic framework, facilitates minimally invasive flexible orbital endoscopic procedures.

References:

Keywords: Optic nerve, Endoscopy, Surgery, Image Guidance, Augmented reality

Financial Disclosures: The authors had no disclosures.
Optical Coherence Tomography in the Evaluation of Long-Term Hydroxychloroquine Therapy: What does it tell us?

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Introduction:
In 2011, revised guidelines for screening of hydroxychloroquine (HCQ) retinopathy recommended spectral domain-optical coherence tomography (SD-OCT), fundus autofluorescence or multifocal electroretinogram to be performed in addition to ophthalmologic examination and automated 10-2 perimetry based on studies that showed SD-OCT changes can occur before visual field loss. Sensitivity and predictive value of SD-OCT compared to visual fields are unknown. We undertook a point prevalence study of SD-OCT in a well characterized population of 64 patients with various rheumatological diseases on long-term HCQ therapy in order to determine the utility of this test in the evaluation of potential toxicity.

Methods:
Prior standard recommendations for screening included visual acuity, automated 10-2 perimetry and fundus appearance. Although there are no established criteria for predicting toxicity, these tests were used as the “gold standard” for evaluation of SD-OCT using Fisher’s exact test. Each eye was evaluated separately and considered positive if 2 or more of these tests were abnormal.

Results:
Sensitivity of SD-OCT using 2 or more test abnormalities was 0.25. Specificity was 0.98. Positive predictive value was 0.50 and negative predictive value was 0.94.

Conclusions:
We conclude that in the presence of an abnormal clinical examination, an abnormal SD-OCT can be used to validate HCQ toxicity when the diagnosis is suspected but not established. However, a normal SD-OCT cannot be used to exclude HCQ retinopathy when visual fields or fundus examination are abnormal. Likewise, SD-OCT was not sensitive enough to predict incipient HCQ retinal toxicity in the absence of other clinical abnormalities. Prospective studies are underway to determine the positive predictive value of SD-OCT in HCQ retinal toxicity. In this way, rational guidelines can be established for the use of SD-OCT in patients on long-term HCQ therapy bearing in mind the control of their rheumatological disorder, availability of SD-OCT and rising health care costs.

Keywords: Hydroxychloroquine, OCT, Retinopathy

Financial Disclosures: The authors had no disclosures.
Quantitative Changes in Perimacular Retinal Thickness by Optical Coherence Tomography in Patients on Long-Term Hydroxychloroquine Therapy

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Introduction:
Spectral domain-optical coherence tomography (SD-OCT) can show qualitative changes prior to visual field loss in patients on long-term hydroxychloroquine (HCQ) therapy. Loss of the inner-/outer-segment line, thinning of the outer nuclear layer and downward displacement of inner retinal layers involving the perifoveal retina are signs of toxicity, but may be difficult to identify conclusively. A simpler quantitative marker of incipient HCQ retinopathy would be of value in assessment of patients on HCQ as a disease modifying agent in rheumatological disease.

Methods:
We undertook a point-prevalence study to evaluate quantitative SD-OCT changes in a well characterized population of 67 patients with various rheumatological conditions who had been on HCQ therapy (mean duration 12.0 years; range 1.5-22 years).

Results:
There was a significant inverse relationship between retinal thickness and cumulative dosage involving all quadrants of the inner perifoveal ring, but only temporal and inferior quadrants of the outer ring. The rate of loss of retinal thickness was significantly greater in the inner ring (-0.34 microns/g/kg) than the outer ring (-0.31 microns/g/kg). Temporal and nasal quadrants of the inner ring were most affected. Based on the 95% prediction band, minimum predicted retinal thickness of temporal and nasal quadrants prior to HCQ therapy would be 314.0 (SE 7.3) microns and 320.6 (SE 7.7) microns respectively.

Conclusions:
We recommend that HCQ not be considered as disease modifying therapy in patients with rheumatological disorders whose initial SD-OCT show perifoveal inner ring retinal thickness of less than 314 microns in the temporal quadrant and 321 in the nasal quadrant. These patients may be at greater risk for HCQ retinal toxicity. If patients are already on HCQ then consideration should be given to lowering the dosage of HCQ and performing more frequent examinations and SD-OCT in order to avoid HCQ retinopathy.

Keywords: Hydroxychloroquine, OCT, Retinal thickness, Retinopathy

Financial Disclosures: The authors had no disclosures.
Repeatability of Blood Flow Velocity Measurements of Human Conjunctiva Using the Retinal Function Imager (RFI)

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Introduction:
The eye provides a window to study the etiology and pathogenesis of cerebrovascular diseases. Ocular microvascular structural alterations may reflect micro-angiopathic processes in the brain. The objective of this study was to test the repeatability of measuring blood flow velocities in the conjunctiva.

Methods:
A Retinal Function Imager (RFI, Optical Imaging Ltd, Rehovot, Israel) was used to capture reflectance changes as a function of time under stroboscopic illumination. The system was used to image the conjunctiva microvasculature. Hemoglobin in red blood cells was used as an intrinsic motion-contrast agent in the calculation of the velocity of blood flow. Seven health subjects (4 males and 3 females, age 36.4 ± 8.4 years) were recruited. The temporal conjunctivas of the right eye were imaged in two sessions on the same visit. The blood flow velocities were measured at about twenty vessel segments by the same ophthalmologist. Intraclass correlation coefficients (ICC) and the coefficient of repeatability (CR) were calculated.

Results:
The mean blood flow velocities of the arteries in the temporal conjunctiva were 0.77 ± 0.10 (mean ± SD, mm/s) and 0.80 ± 0.10 mm/s for sessions 1 and 2 (p>0.05), respectively. The blood flow velocities of the veins were 0.74 ± 0.12 and 0.71 ± 0.17 mm/s for sessions 1 and 2, respectively (p > 0.05). The ICC was 0.84 and CR 0.11 mm/s for arterial and ICC 0.91 and CR 0.15 mm/s for venous flow velocities. There was no significant difference of mean blood flow velocities (P>0.05) between arteries and veins.

Conclusions:
To the best of our knowledge, this is the first study to demonstrate the repeatability of measuring conjunctival blood flow velocity using RFI. The repeated reliability appeared to be moderate to excellent. {Supported in part by the research grants NIH R01EY020607, NIH R01EY020607S, NINDS K24 NS 062737 (TR)}

Keywords: Conjunctiva, Microvasculature, Velocity, Optical image, Cerebrovascular disease

Financial Disclosures: The authors had no disclosures.
Poster 48

Magnetic Resonance Imaging of Superior Oblique Muscle Atrophy in Trochlear Nerve Schwannoma

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Introduction:
Fewer than 80 cases of trochlear nerve sheath tumor (schwannoma) have been reported since 1976. Identification of small trochlear schwannomas was elusive prior to the advent of MRI, and gadolinium-enhanced MRI has facilitated more frequent and earlier identification of these tumors.

Methods:
Case Report

Results:
We present a case report of a 44-year-old male with a history of hyperthyroidism, who presented with vertical diplopia consistent with a left trochlear nerve palsy. His initial MRI study demonstrated a small enhancing mass in the left perimesencephalic cistern along the expected course of the trochlear nerve. The superior oblique muscles appeared normal and symmetric. Eight months later, there was an interval worsening of the left superior oblique palsy. A second MRI exam showed development of selective atrophy of the left superior oblique muscle ipsilateral to the cisternal trochlear nerve tumor. The presumed trochlear schwannoma showed only slight increase in size, increasing from 5 x 3 mm to 6 x 4 mm over the eight-month period. Also of interest is the finding of FLAIR hyperintensity of the tumor on pre-gadolinium images. Diagnosis is consistent with left trochlear schwannoma with denervation atrophy of the left superior oblique muscle. Treatment was initiated with corrective prisms and monitored with follow up examinations. Due to the increased size of the tumor and denervation atrophy of the superior oblique muscle, it was elected to proceed with stereotactic radiotherapy. Followup examinations have remained stable.

Conclusions:
This case documents evolution of superior oblique muscle denervation atrophy in a patient with a trochlear schwannoma. Obtaining serial MRI scans of both the brain and orbit may not only be of benefit in establishing the diagnosis of a trochlear schwannoma, but will also aid in the consideration of therapeutic options.

References:


Keywords: Schwannoma, Muscle atrophy, Trochlear, Nerve, Imaging

Financial Disclosures: The authors had no disclosures.
Arteritic and Non-Arteritic Anterior Ischemic Optic Neuropathy: Relative Incidence in Saudi Arabia

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Introduction:
Based on the small absolute number of patients with biopsy proven giant cell arteritis (GCA), it has been suggested that the incidence of GCA is lower in the Saudi Arabian than western (Caucasian) population. (1) To further assess this claim, we evaluate the proportion of patients presenting with anterior ischemic optic neuropathy (AION) found to have GCA in Saudi Arabia.

Methods:
In this University Based Study, the computer database at a single institute was used to identify 171 consecutive patients presenting with AION between 1997 and 2012. Diagnostic criteria for non-arteritic AION (NAION) were similar to that used in the IONDT. (2) The diagnosis of arteritic AION (AAION) was based on clinical presentation and positive temporal artery biopsy. The number of patients with and without GCA was tabulated and the proportion and 95% confidence interval (CI) determined.

Results:
Four patients (2 male, 2 female; age: mean 76, range 70 to 80 years) were diagnosed with AAION and 167 patients (117 male, 50 female; age: mean 57, range 30 to 93 years) with NAION. The proportion of patients presenting with AION found to have GCA was 2.3% (95% CI, 0.74 to 5.6%).

Conclusions:
In this Arabic population, GCA is the cause of AION in 2.3% (95% CI, 0.74 to 5.6%) of patients. This is less than that reported for western (primarily Caucasian) populations, roughly 6%. (3) This is consistent with previous assertions that the prevalence of GCA is relatively low in Saudi Arabia. An alternate explanation is that our results reflect a higher incidence of NAION. The difference between Arabic and Western populations is relatively small, with the upper confidence interval (0.74 to 5.6%) approaching close to the Western 6% estimate. An assessment of additional, possibly larger populations is necessary to determine the true significance of this observation.

References:

Keywords: NAION, Giant cell arteritis, Epidemiology, Arteritic AION, International medicine

Financial Disclosures: The authors had no disclosures.
Transorbital Approach for Diagnosis of Lesions of the Cavernous Sinus

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Introduction:
Cavernous sinus lesions frequently present with neuro-ophthalmic signs and symptoms including diplopia and sensory changes. Treatment (surgery, radiation, chemotherapy) depends on specific diagnosis. While imaging studies may localize pathology, having tissue for pathologic and immunologic evaluation can be critical. This is particularly true for malignant lesions affecting the cavernous sinus. Previous attempts at diagnosis include open craniotomy and potentially fine needle aspiration biopsy. In order to reduce the morbidity associated, we have extended the use of a lateral orbitotomy to allow biopsy of pathology in and around the cavernous sinus.

Methods:
Case report of two patients undergoing translateral orbitotomy approach to the cavernous sinus with biopsy of lesions affecting the cavernous sinus.

Results:
In both cases a previous attempt at fine needle aspiration biopsy through the foramen ovale was unsuccessful in providing adequate tissue to direct therapy. In one patient sclerosing idiopathic orbital inflammatory disease with extension into the cavernous sinus (Tolosa Hunt) was diagnosed and the patient was subsequently treated with biologic agents. In the second case, metastatic melanoma was diagnosed and adequate tissue was obtained to look for BRAF markers.

Conclusions:
Translateral orbital approach to the cavernous sinus is safe and effective in making a specific diagnosis. A transorbital approach to the cavernous sinus has substantial less morbidity than a transcranial approach. These patients were both treated as outpatients, and suffered no ill effects of their surgery. This can reduce the morbidity present in a more classic open craniotomy approach.

Keywords: Cavernous sinus, Orbital surgery, Metastatic melanoma, Sclerosing orbital inflammatory disease, Tolosa Hunt Syndrome

Financial Disclosures: The author had no disclosures.
Use of Posterior Fixation Sutures (Myopexy) to Expand Binocularity: Indications and Limitations

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Introduction:
Intractable diplopia is not a rare complication of either paretic or restrictive limitation in extra ocular motility. The primary goal of muscle surgery is to achieve binocularity in primary position and down reading gaze. To maximize the area of binocular, patients may be treated with surgery to limit the motility of the better moving eye. The Posterior fixation suture is an effective means of limiting motility.

Methods:
A retrospective review of 43 patients undergoing 44 posterior fixation procedures over a 13 year period. Patients were assessed quantitatively with Hess screen and binocular single vision fields.

Results:
There were a total of 26 women and 17 men (age 14-88 mean 58). The most common cause of paretic limitation in motility was the presence of skull base tumors (9 meningiomas, 2 pituitary tumors) with additional malignant tumors and a midbrain glioma. Four patients had aneurysms and 2 had carotid cavernous fistulae. Additional etiology included trauma in 11 and 1 patient who had restrictive strabismus secondary to infection following surgery for fibrous dysplasia. The most common muscle involved was the medial rectus (21 cases) followed by the inferior rectus in 12, the lateral rectus in 10 and the superior rectus in 1. Two patients required repeat procedures. One patient had inadequate follow up, 1 patient continued to wear an occluder over 1 eye to avoid diplopia, and 1 patient was considered a failure even though the area of binocularity remained no worse. Only 4 patients had no diplopia at all, but most the patients had expansion of binocularity so that double vision occurred only in extreme eccentric gaze.

Conclusions:
Posterior fixation myopexy is a useful technique for expanding binocularity in patients with persistent diplopia secondary to cranial nerve palsies and restrictive strabismus.

Keywords: Diplopia, Binocular single vision fields, Hess screen, Muscle surgery

Financial Disclosures: The author had no disclosures.
Ocular Motor Abnormalities and Congenital Facial Weakness

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Introduction:
The classic condition of ocular motor deficits and facial weakness is Mobius syndrome, attributed to abducens and facial nerve involvement. Reports of facial weakness with oculomotor dysfunction or accommodative esotropia due to genetic mutations (TUBB3 and HOXB1, respectively), have recently broadened the clinical spectrum.

Methods:
We report ocular motor findings in 39 patients with congenital facial weakness.

Results:
We examined 39 patients (age range 2-64, 25 females), 36 with bilateral and 3 with unilateral facial weakness. Two had isolated bilateral abduction defects. The most common pattern (n=16) was bilateral horizontal gaze paresis (BHGP) with spared vertical range. Ten patients had BHGP plus vertical involvement, 4 with impaired elevation and depression and 6 with impaired elevation only. Nine patients had full motility range; 2 with a large esotropia. One patient had isolated unilateral limited adduction and one could not be characterized. Two patients had bilateral ptosis – one with diffuse ophthalmoplegia and one with spared vertical eye movements. No patient had pupillary involvement. Twenty-nine patients performed pursuit, saccade, and optokinetic nystagmus testing. Of particular importance was the absence of vertical optokinetic nystagmus in 19 patients, even when vertical range of motion was unaffected. Twenty-three patients had not undergone strabismus surgery. Of these, 15 had normal alignment. By history and exam, 59% had tongue abnormalities, 51% imbalance, and 26% cognitive impairment. Mirror movements, seizures, and psychiatric abnormalities occurred, but were less common.

Conclusions:
A range of ocular motor abnormalities and other deficits may occur with congenital facial weakness. The most common pattern of BHGP suggests involvement of motoneurons and interneurons in the abducens nuclei. Characterization of the patterns of ocular motor deficits in congenital facial weakness may assist continued advancement in identification of the genetic defects underlying these disorders.

Keywords: Congenital facial weakness, Mobius syndrome, Horizontal gaze palsy, Optokinetic nystagmus

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Poster 53

Measuring Retinal Vascular Tortuosity – A Harmonic Analysis-Ergodic Theory Based Approach

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Introduction:
This presentation (1) introduces the ergodicity defect (ED) as a measure for type I (e.g., retinal disease of prematurity) and type III (e.g., atherosclerosis) retinal vascular tortuosity [1, 2] and (2) considers a connection between the ED values and the diagnosis of background diabetic retinopathy. In [3], ED identified tortuosity in vessels segmented from brain MRA images and with both 2D and 3D synthetic data, ED worked even in cases where other metrics failed.

Methods:
ED measures the complexity of a signal in terms of how the signal covers the space [4, 5]. A vessel that is tortuous is more complex and has a lower ergodicity defect than nontortuous vessels. ED draws from several mathematical theories and the computation takes as input a set of points describing the vessels and gives an output corresponding to the extent of tortuosity in the vessel. An open source, image analysis software was used to manually segment retinal vessels from images obtained from the DRIVE and Messidor databases and, for ease of computation, the ergodicity defect is computed with respect to the Haar wavelets. Diagnosis for the DRIVE images was provided by an expert and a retinopathy grade accompanies the Messidor images.

Results:
7 DRIVE subjects have diabetic retinopathy or vascular abnormalities and ED identified abnormal vascular tortuosity in 6 of the images. 25 Messidor images were analyzed and those ED values indicate that the higher the retinopathy grade, the lower the ED value. However, this correspondence does not always hold in the case of type III tortuosity.

Conclusions:
The ergodicity defect (ED) is shown to be an effective measure for retinal type I and III vascular tortuosity in retinal data images. The preliminary findings also suggest a correspondence between the retinopathy grade and the ED value, but this connection is inconsistent for type III tortuosity.

References:

Keywords: Retinal vessels, Tortuosity measure, Diabetic retinopathy, Retinal pathology diagnosis, Retinal image data

Financial Disclosures: The author had no disclosures.
Post Infectious Optic Neuritis: Manifestations in Children Differ From Adults

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Introduction:
Post infectious optic neuritis (ON) appears at any age. Though children very commonly suffer from infectious diseases, ON is reported to be less common than that in adults, with annual incidence of 0.33-1.66/10000 vs. 5.1/100000. The aim of this work is to compare the clinical manifestations and outcomes of the disease in adults and children.

Methods:
A retrospective case-series from two neuro-ophthalmology services. Included were all patients with a diagnosis of ON associated with infectious disease between 2005 – 2012. Excluded were all patients with any other possible cause for optic neuropathy such as compressive, immune or demyelinating.

Results:
Included were 11 children and 8 adults. Among children and adults 55% (6/11) and 50% (4/8) were female, respectively. The mean follow-up was 27 months (2-72 months) and 18 months (2-80 months) in children and adults, respectively. Bilateral ON was found in 55% (6/11) of the children and 38% (3/8) of the adults. Causative pathogen was identified in 55% (6/11) of the children’s group, 67% (4/6) of them were Mycoplasma Pneumonia. In adults causative pathogen was identified in 75% (6/8) however without any common denominator. Time interval between febrile illness and ON was 6 days (range 1-14 days) in children and 19.5 days (range 14-30 days) in adults. Acute Disseminated Encephalomyelitis (ADEM) was diagnosed in 45% (5/11) of children and none of the adults. Final visual functions were good in both groups with visual acuity 20/30 or better in all patients.

Conclusions:
Though the final outcome of post infectious optic neuritis is good at any age, the clinical manifestation differs in children, probably a different entity of that seen in adults. Children tend to develop the disease earlier and more frequently in conjunction to ADEM. The disease was bilateral and the causative agent was isolated more frequently.

Keywords: Post infectious, Optic Neuritis, Children, Adults

Financial Disclosures: The authors had no disclosures.
Intracranial Pressure Measurements in Astronauts With Optic Disc Edema, Choroidal Folds, and Cotton Wool Patches Following Long Duration Space Flight

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Introduction:
Introduction: Optic disc edema, choroidal folds, hyperopic shifts, and cotton wool patches have recently been reported in astronauts following long duration space flight. The etiology of these findings remains unknown but one hypothesis has been increased intracranial pressure resulting from long term microgravity exposure.

Methods:
Methods: A retrospective case series of astronauts undergoing lumbar puncture following long duration space flight.

Results:
Results: Intracranial pressure measurements taken at various intervals following return to earth after long duration space flight have been variable ranging from normal (opening pressure less than 20 cm of water) to borderline high (20-25 cm), to elevated (up to 28.5 cm).

Conclusions:
Conclusion: Increased intracranial pressure may be a contributing factor in the development of the ocular and orbital findings in astronauts after long duration space flight but may not be the primary or only mechanism for the observed findings. Cephalad fluid shift to the eye or orbit may be a better primary or single unifying etiologic diagnosis in this setting but further prospective evaluation is ongoing.

References:


Keywords: Astronaut, Optic disc edema, Papilledema, Choroidal folds, Space flight

Financial Disclosures: The authors had no disclosures.
Serial OCT Examination of the Macula in Normal Patients Talking Hydroxychloroquine

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Introduction:
Spectral Domain OCT has been suggested as a modality for monitoring hydroxychloroquine (plaquenil) retinopathy. It has been shown that patients with confirmed hydroxychloroquine toxicity exhibit global macular thinning and “sinkhole effect” with loss of inner retinal layers.

Methods:
We have investigated clinical parameters on patients without clinical toxic maculopathy undergoing monitoring including serial OCT measurements. 15 patients were randomly selected with history of hydroxychloroquine use ranging from 30 to 340 months. Differences in OCT taken an average 18 months apart were examined.

Results:
Two of the 15 patients showed increased thinning in all quadrants and were recommended to cease hydroxychloroquine. The remainder showed stability or more commonly, a slight (2-7 micron) increase in macular thickness.

Conclusions:
Hydroxychloroquine maculopathy is rare. Serial OCT may be a more objective method for following patients. The most common finding in patients without toxicity is a slight but consistent thickening in both central, inner and outer macular quadrants.

References:
1. Spectral-Domain Optical Coherence Tomography and Adaptive Optics may Detect Hydroxychloroquine Retinal Toxicity before Symptomatic Vision Loss

Keywords: Hydroxychloroquine, Ocular Coherence Tomography, Drug Toxicity

Financial Disclosures: The authors had no disclosures.
Pallid disc edema and choroidal perfusion delay in post-hemodialysis non-arteritic ischemic optic neuropathy

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Introduction:
Non-arteritic anterior ischemic optic neuropathy (NAION) is a rare but recognized ophthalmic complication in patients with end-stage renal disease (ESRD) on chronic hemodialysis characterized by unilateral or bilateral acute visual loss. Anemia, concomitant vaculopathic risk factors (i.e. diabetes, atherosclerosis), autonomic dysfunction and the hypotensive effect of hemodialysis are among the predisposing factors to the development of ischemic optic neuropathy (ION) in this patient population. Although pallid disc edema, cotton wool spots, and a choroidal perfusion delay have been classically associated with giant cell arteritis (GCA). We reported two cases of NAION in patients with ESRD on chronic hemodialysis who demonstrated pallid disc edema in conjunction with choroidal and retinal ischemia. To our knowledge, this is the first such report in the English language ophthalmic literature.

Methods:
Retrospective review of charts, Visual field and Fluorescein angiography (FA) of two patients with ESRD on chronic hemodialysis.

Results:
Post-hemodialysis NAION is a demonstrable non-GCA cause of pallid disc edema, retinal ischemia, and choroidal filling defects.

Conclusions:
Post-dialysis NAION can present with pallid edema, retinal ischemia, and achoroidal perfusion defect similar to that of arteritic ischemic optic neuropathy (ION). Ophthalmologists should be aware that post-dialysis NAION can present with pallid edema and a choroidal perfusion defect similar to that seen in arteritic ION.

References:

Financial Disclosures: The authors had no disclosures.
Invasive Thymoma Presenting with Ocular Myasthenia Gravis

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Introduction:
Invasive thymoma is an uncommon neoplasm with an aggressive course that has been noted with increasing frequency in recent years. Forty percent of invasive thymomas are associated with paraneoplastic syndromes including myasthenia gravis. We report two patients with ocular MG with biopsy confirmed invasive thymoma. Prompt computed tomography (CT) of the chest should be performed in all patients with MG including ocular MG. To our knowledge this is the largest series in the English language ophthalmic literature of invasive thymoma with ocular MG.

Methods:
Retrospective chart review including radiographic imaging studies of two patients with ocular MG and thymoma.

Results:
Patient 1 had a thymic lesion characterized by mixed pattern grade B1, B2 and focal B3, stage III. Patient 2 had invasive thymic epithelial neoplasm with conventional and atypical features, stage II. Patient 2 had post-thymectomy and radiation recurrence of MG symptoms.

Conclusions:
Patients with ocular MG should undergo prompt CT of the chest to rule out thymoma. Delayed diagnosis and surgical treatment of thymoma might affect prognosis as more invasive stages have significantly higher mortality rates. In patients with prior history of thymoma presenting with worsening symptoms, repeat radiographic imaging to exclude recurrent thymoma should be considered.

References:


Financial Disclosures: The authors had no disclosures.
First Cases of Dominant Optic Atrophy (DOA) in Saudi Arabia: A Report on a Novel Genetic Mutation

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Introduction:
The majority of DOA cases (50-60% of the patients) are caused by mutations of the OPA-1 gene, which codes for a protein (dynamin-related GTPase) of the internal mitochondrial membrane. Currently, more than 200 pathogenic mutations in the OPA-1 gene have been described. Regardless, in large-scale studies, DOA in certain families has been related to other chromosomal loci, i.e., OPA-3, OPA-4, OPA-5 and OPA-7.

Methods:
In this study, we describe a clinical series of 40 patients from Saudi Arabia with a positive DOA phenotype (i.e., decreased VA during the first two decades of life, temporal or global optic nerve pallor, and an absence of other neurological or ophthalmological diseases that could explain the optic neuropathy) who underwent testing for OPA-1 (and, in some cases, for OPA-3).

Results:
We present the results of the evaluation, including the first five cases (to the best of our knowledge) of a confirmed OPA-1 mutation in Saudi Arabia, and report a previously undescribed novel mutation that was found in one patient.

Conclusions:
The question remains whether certain patients in Saudi Arabia with a clearly defined DOA phenotype may have chromosomal loci other than OPA-1 and OPA-3. It is likely that genetic alterations associated with different loci will be discovered in the future, thereby allowing us to answer these questions.

References:

Keywords: Dominant optic atrophy, OPA-1, Congenital optic neuropathy

Financial Disclosures: The author had no disclosures.
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Idiopathic Sclerosing Orbital Inflammation: Presentation of an Unusual Case with Isolated Bilateral Optic Nerve Involvement

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Introduction:
Idiopathic sclerosing orbital inflammation (ISOI) manifests clinically as orbital inflammation with the main symptoms of pain, proptosis and vision loss. However, unlike idiopathic orbital inflammation (IOI), which has an acute clinical course, the progression of ISOI cases typically have a chronic and indolent course.

Methods:
Herein, we report an unusual clinical case of ISOI with predominant and almost exclusive bilateral optic nerve involvement.

Results:
The presumptive diagnosis was sarcoidosis according to the previous existence of facial paralysis and increased angiotensin-converting enzyme (ACE) levels; however, a biopsy of the optic nerve (no light perception) clearly indicated a definitive diagnosis of ISOI.

Conclusions:
ISOI is a rare clinical condition for which proper treatment remains poorly established. To our knowledge, and after reviewing the scientific literature, we believe that this is the first clinical case of ISOI with predominant optic nerve infiltration.

References:


Keywords: Idiopathic sclerosing orbital inflammation, Optic neuropathy, Idiopathic orbital inflammation

Financial Disclosures: The authors had no disclosures.
Lambert-Eaton Myasthenic Syndrome And The Eye

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Introduction:
Wide ranges on the frequency of ophthalmic symptoms and signs in Lambert-Eaton Myasthenic Syndrome (LEMS) have led to mixed and occasionally contradictory messages on how frequently the visual system is involved.

Methods:
Records of 176 patients with diagnosis of LEMS made at a single tertiary medical center from Jan. 1, 1976-Dec. 31, 2010 were reviewed with special attention to ophthalmic symptoms and signs.

Results:
Ophthalmic symptoms: 23.3% of patients had ptosis, 20.5% had diplopia, 13.6% had decreased vision, and 6.8% had dry eyes. Ophthalmic signs: 25.6% had ptosis, 8.5% had motility dysfunction, 8% had strabismus, 6.8% had pupillary dysfunction, and 2.3% had clinical signs of dry eye.

Conclusions:
Although they are not the most common symptoms in LEMS, a significant number of patients reported ophthalmic symptoms and many had objective findings of ophthalmic involvement on clinical exam. The findings of this study, the largest series to focus on ophthalmic findings, suggest that LEMS should continue be a diagnostic consideration in patients with ophthalmic symptoms and signs, especially in those with systemic symptoms including weakness and autonomic dysfunction.

Keywords: Lambert Eaton, Myasthenia

Financial Disclosures: The authors had no disclosures.
Patients with a Homonymous Hemianopia Able to Drive With the Use of Novel Monocular Sector Prisms

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Introduction:
Patients with homonymous hemianopia (HH) often fail to meet visual field (VF) requirements for a driver license. We describe two patients with complete HH who met the minimum VF requirements for driving using a novel, high-power, monocular sector prism system.

Methods:
Two patients with complete HH were identified retrospectively from billing records. Each patient underwent full neuro-ophthalmologic examination. Baseline VFs were assessed using automated (Humphrey) perimetry using the SITA-Fast 24-2 program and kinetic (Goldmann) perimetry using dynamic and static size-III stimuli. VFs were reassessed at multiple time points to confirm stability. Patients were fitted with glasses and press-on 57-PD peripheral monocular sector prisms placed above and below the visual axis with prisms oriented obliquely. Kinetic perimetry was re-assessed both monocularly and binocularly, with and without prisms.

Results:
The state requires 105 degrees of horizontal VF. Case 1 had a left HH with 95 and 65 degrees of horizontal VF in the right and left eyes, respectively, and 95 degrees binocularly. With the use of the prism system described above, the binocular VF increased to 115 degrees of continuous, horizontal VF. This patient passed a comprehensive, behind-the-wheel driver evaluation. Case 2 had a left HH with 79 and 40 degrees of VF in the right and left eyes, respectively, and 82 degrees binocularly. With the use of the prism system, the binocular VF increased to 112 degrees of continuous, horizontal VF. Both patients reported significant improvement in quality of life and satisfaction with their prism-containing glasses. Each holds a valid driver license and has successfully operated a motor vehicle without any restrictions or accidents.

Conclusions:
Clinicians should consider the addition of oblique, 57-PD prisms to the ipsilateral spectacle lens above and below the visual axis for patients with complete or nearly complete HH.

Keywords: Homonymous hemianopia, Prism, Visual field, Drive

Financial Disclosures: The authors had no disclosures.
Optic Nerve Tortuosity and Vision Loss in Neurofibromatosis Type 1
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Introduction:
In children with neurofibromatosis type 1 (NF-1), the prevalence of optic pathway glioma (OPG) is as high as 20%, with nearly one-half of these patients developing vision loss. Optic nervetortuosity (ONT) in the absence of thickening or gadolinium enhancement is often found on MRI of NF-1 patients. However, it is unknown whether ONT predicts future development of OPG or vision loss.

Methods:
Children with NF-1 diagnosed at a national referral center between 1992 and 2005 were identified. Those with at least one MRI rain/orbital MRI and with subsequent visual acuity data from follow-up eye exams were then studied. Clinical and vision data was obtained through standardized chart review. Three pediatric neuroradiologists independently reviewed the initial MRI and identified patients with ONT based on the operational criteria established by Armstrong et al (Am J NeuroRadiol, 2005, 28:666-671). Decreased visual acuity was defined as ≥0.2 logMAR below the age-matched normative value at last vision testing, or at the last visit before treatment of an OPG was initiated. The value of ONT in predicting poor visual acuity was assessed by chi-squared testing.

Results:
263 NF-1 patients had a brain MRI during the study period, and 202 of those (404 eyes) had visual acuities documented at least one year after the initial imaging study. Standardized interpretation of each MRI is underway and has identified more than 30 patients with ONT. The radiographic assessment will allow for a well-powered assessment of ONT as a possible predictor of poor vision.

Conclusions:
This study will establish whether ONT predicts subsequent vision loss, and will thus help to clarify whether frequent serial eye exams and imaging studies are necessary in this population.

References:

Keywords: NF-1, Optic pathway glioma, Vision loss

Financial Disclosures: The authors had no disclosures.
Is There Value to the Use of Brain Atlas in Idiopathic Intracranial Hypertension?

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Introduction:
This study aimed to develop disease-specific probability brain atlas with disease-relevant parcellation maps from a small cohort of patients with idiopathic intracranial hypertension (IIH). Probability atlases can identify patterns of altered structure and/or function, and can guide image analysis algorithms for further structural and functional examinations, automated image labeling, and tissue classification for volume and shape analysis.

Methods:
Brain MRI was performed in 5 patients with IIH diagnosed according to the modified Dandy criteria. High resolution, 3D T2-weighted FLAIR fat-suppressed and FSE MRI datasets were obtained in the coronal plane at 3T. Firstly, the individual datasets were mapped into stereotaxic space. Secondly, the brain data were manually segmented and labeled into sub-volumes of neuro-ophthalmic interest, such as optic nerve head, optic nerve, optic nerve sheath, optic chiasm, pituitary gland and cerebellar tonsils. Thirdly, the probability map was then constructed for each segmented structure by determining the proportion of subjects assigned a given anatomic label at each voxel position in stereotaxic coordinates. For visualization and atlas creation we used imaging processing software.

Results:
To our knowledge, this is the first brain probability atlas with manual parcellation maps in patients with IIH. The processing of images from patients with such complex disease as IIH in the context of a disease-specific probabilistic atlas enables a more meaningful interpretation of data. The construction of averages, templates and models allow describing how the brain and its component parts are organized and allow further analysis and comparison with other datasets.

Conclusions:
In this preliminary study of IIH-specific brain atlas construction of optic nerve sheath and other seemingly involved structures is shown. The algorithm used in the Atlas Creator module that aligns images paired with segmentations to generate statistical atlases proved to be a valuable method that requires verification on larger datasets and against other available algorithms.

References:


Keywords: Idiopathic intracranial hypertension, Brain atlas, Parcellation Label maps, MRI, Optic nerve segmentation

Financial Disclosures: The authors had no disclosures.
Optical Measurement of Craniofacial Autonomic Function

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Introduction:
Autonomic function, including systemic sudomotor, adrenergic, and cardiovagal responses, is widely tested. However, there is no routinely available method of measuring craniofacial autonomic phenomena. Pupillary responses are commonly tested, but they are rarely observed in the context of the broader craniofacial autonomic response. We are developing a clinically feasible protocol for measurement of craniofacial autonomic function.

Methods:
Subjects were placed supine and imaged under green, red and blue light. A high sensitivity CCD camera was used to image the upper two-thirds of the face. A pulse oximetry probe was placed on the earlobe. A twin-bore tube was inserted into one nostril, and the contralateral nostril was occluded. Imaging and pulse oximetry were recorded before, during and after stimulation with ammonia vapor, which was directed into the nostril via vacuum pump. Additional testing in selected cases included facial skin potentials, Schirmer’s test and slit-lamp evaluation.

Results:
Data was collected in six subjects. At baseline, low frequency fluctuations in skin reflectance were noted, along with reflectance changes due to heartbeat. Between 1-3 seconds after ammonia stimulus, bilateral consensual pupillary dilation, followed by relative constriction, was observed. During and shortly following the constriction phase, reflectance over the forehead and cheeks decreased bilaterally (most prominent ipsilaterally), concurrent with a reduction in spontaneous vascular fluctuations. Scleral/conjunctival reflectance also decreased bilaterally and tearing increased ipsilaterally. Imaging at multiple wavelengths, as well as slit lamp exam with a similar paradigm, suggested that the mechanism of reflectance change is an increase in blood volume due to vasodilation.

Conclusions:
Our optical, multi-modal measure of facial and ocular trigeminovascular autonomic function fills a gap in current clinical measurement of autonomic function. This technique is clinically feasible and may serve as a supplement to the characterization of neuro-ophthalmologic clinical entities with prominent facial autonomic responses, including migraine, cluster headache, and multiple sclerosis.

Keywords: Autonomic testing, Trigeminal, Optical imaging

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Binocular Treatment of Lazy Eye (Amblyopia)

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Introduction:
Amblyopia, a developmental disorder of the visual system that results in information from one eye being ignored by the brain, is currently untreatable in adults. Since amblyopia involves visual loss in one eye, one of the primary deficits is deficient cooperation between the two eyes, which is caused by suppression of the input from the amblyopic eye by the input from the normal eye. Misalignment of the eyes (strabismus) is a common cause of amblyopia. Corrective surgery in strabismus improves motor binocular fusion and improves vision in children; however, this effect is modest in adults.

Methods:
A technique whereby images were presented separately to each eye was used. The images presented to the amblyopic eye were of a much higher contrast than those presented to the normal eye. The participants performed a visual task that required binocular integration of information. The contrast difference between the two eyes varied until binocular integration occurred (balance point), a useful clinical measure of suppression. Repeated measurement of this balance point resulted in improved sensory binocular fusion and the vision in the amblyopic eye.

Results:
22 patients from 5 to 73 year-old (mean 36.2 year-old) have been tested and showed on average 3.2 lines improvement in visual acuity, which was stable on average for six months. The visual improvement correlated with number of sessions of treatment and age but not with the severity of amblyopia.

Conclusions:
The results suggest that the treatment procedure used in this study can improve visual function in amblyopic eyes of adults. In a subsequent study, we plan to use this binocular treatment before and after strabismic surgery and hypothesize that combining improvement in sensory and motor binocular fusion has an augmenting effect. This is a novel approach to treatment of strabismic amblyopia and clinically significant as it might conclude in a new treatment approach.

Keywords: Amblyopia, Strabismus, Binocular vision, Treatment, Surgery

Financial Disclosures: The authors had no disclosures.
Simultaneous Paraneoplastic Optic Neuropathy and Ocular Flutter-Myoclonus Syndrome Associated With Lung Adenosquamous Carcinoma and Circulating Collapsin Response-Mediating Protein (CRMP-5)

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Introduction:
We describe a patient with a complex paraneoplastic visual and oculomotor syndrome associated with circulating CRMP-5 antibody and lung cancer. After treatment with oral clonazepam and intravenous immunoglobulin, the ocular fixation instability dramatically improved while severe bilateral visual loss only partially recovered. Subsequent chemotherapy resulted in decreased tumor volume.

Methods:
A 63 year-old man developed subacute bilateral vision loss, upper limb tremor, gait unsteadiness, memory loss and confusion. Physical examination was performed, followed by Onconeural antibodies panel, Head magnetic resonance imaging (MRI), Cerebrospinal fluid (CSF) analysis, Electroencephalogram (EEG), Chest computed tomography (CT) and Transbronchial biopsy (TB).

Results:
On neuro-ophthalmological examination, intermittent ocular flutter, bilateral severe loss of visual acuity (<1/10), left relative afferent pupillary defect and bilateral optic disk edema with macular exudates were noted. Visual field testing revealed severe overall depression. Additionally, upper limb myoclonic tremor, gait ataxia and fluctuating state of consciousness were present. Head MRI showed bilateral basal ganglia T2 hyperintensities with extension to the hippocampal regions. A paraneoplastic antibody panel disclosed a positive CRMP-5 IgG. CSF analysis revealed 150 lymphocytes /mm3, a protein level of 155 mg/dL and the presence of oligoclonal bands. EEG showed focal slow-wave fronto-temporal activity. Chest CT revealed a right upper lobe lung lesion and TB disclosed adenosquamous carcinoma of the lung. A 5-day course of 1 g/day intravenous methylprednisolone provided no clinical improvement. Further treatment with oral clonazepam 2mg/day and monthly intravenous immunoglobulin was started. Ocular flutter, myoclonus and ataxia completely remitted while only right eye visual acuity has partially improved (7/10). Systemic chemotherapy was then instituted, with partial remission of the tumour.

Conclusions:
This is the first report of paraneoplastic ocular flutter associated with circulating CRMP-5 antibody. The co-occurrence of bilateral optic neuropathy and the association with an unusual variant of lung cancer are further underlined.

References:

Keywords: Ocular Motility Disorders, Optic Nerve Diseases, Paraneoplastic Syndromes, Lung Neoplasms

Financial Disclosures: The authors had no disclosures.
In this report, we describe a patient with downbeat nystagmus evoked only by eye closure, associated with a localized paramedian lesion of the lower basis pontis possibly due to involvement of the decussating ventral tegmental tract by ephaptic transmission.

Methods: A 43-year-old woman developed subacute asymmetric distal weakness and paresthesias of the lower limbs and urinary retention, followed 2 weeks later by the sensation of rhythmic eye movement and sustained vertigo provoked strictly by eye closure. Neurological examination, video-nystagmography (VNG), electro-oculography (EOG) and brain and spinal MRI were performed.

Results: Exam demonstrated full ocular motility without spontaneous, gaze-evoked, positional or head-shaking nystagmus. With eyes closed, rhythmic vertical ocular excursions were evident under closed lids associated with imbalance. Lower limb proprioception and vibration were impaired, with a sensory level at T10. She had mild bilateral ankle dorsiflexion weakness and bilateral extensor responses. VNG with eyes open in light or dark was normal. EOG during eye closure in dark revealed downbeat nystagmus (DBN) in primary position with maximal slow phase velocity 18°/s; this disappeared with eyes open. Brain and cord MRI revealed a left lower paramedian pontine T2 lesion with nodular enhancement, and 9 other brain and cord T2 lesions fulfilling MRI criteria for multiple sclerosis. The patient was hospitalized and treated with high dose intravenous corticosteroids. After one month, motor and sensory involvement had completely subsided while DBN evoked by eye closure remained unabated.

Conclusions: This is the first report of DBN evoked only by eye closure associated with focal pontine demyelination. One suggested mechanism for DBN in this case involves transverse ephaptic spread of excitation from areas that subserve coordinated lid closure to the decussating ventral tegmental tract.

References:


Keywords: Nystagmus, Electrooculography, Eye movements, Eyelids, Vertigo

Financial Disclosures: The authors had no disclosures.
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Vector Gradient Analysis of Cupping in Optic Atrophy

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Introduction:
With vector gradient analysis as numeric representative of disk cupping, we compared optic disk cuppings between optic atrophy and control group.

Methods:
A prospective study was conducted in consecutive eighteen eyes of eighteen optic atrophy subjects and twenty-eight eyes of twenty-eight normal subjects between Jul 2011 and Jul 2012. All subjects were assessed with SAP and OCT. The individual OCT data, which was acquired from The Optic Disc Cube protocol of Cirrus-HD OCT, were reconstructed as the 3-dimensional surface map of the optic disc cup, and analyzed the slope and size of cupping of optic disc with custom developed software. Each parameter, the numeric representatives of optic disc cupping and MD and PSD of SAP, was statistically analyzed with Mann-Whitney U test.

Results:
Averages of mean deviation and pattern standard deviation and RNFL thickness in optic atrophy and normal group were statistically different (P<0.05). Averages of "sum of cup vector gradient", "cup size" and "cup volume" were not statistically different between two groups. (P=0.278, 0.455, 0.415, respectively).

Conclusions:
The vector gradient, size and volume of optic disc cupping were not different between normal group and optic atrophy group.

Keywords: Vector gradient, Optic atrophy, Cupping

Financial Disclosures: The authors had no disclosures.
Assessment of Smartphone Color Vision Testing

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Introduction:
The widespread use of handheld computers (‘smartphones’) with high-quality displays has prompted the development of software designed to evaluate color vision at the bedside. Software versions of Ishihara pseudoisochromatic plates may become a popular alternative to the original printed version. As there is no standard for electronic representation of Ishihara plates, the optimal software and display characteristics for electronic color vision evaluation are unknown.

Methods:
The display characteristics of several smartphones were evaluated using commercially-available video diagnostic software. Searching a smartphone application store with defined search parameters, we assessed 13 existing free English-language color vision testing applications. Specifically, we evaluated characteristics likely to influence the clinical reliability of results.

Results:
We found that among six popular smartphones, there was considerable variability in display technology, including differences in maximum brightness, contrast ratio, and color accuracy. The applications that met our inclusion criteria varied in number of plates displayed, plate diameter, color quality, plate resolution, and method of testing.

Conclusions:
The ability to perform smartphone-based color vision testing will potentially allow more convenient testing of this portion of the examination in settings where standard color vision testing is not possible. Our results suggest that several variables exist across existing smartphone-based color vision testing applications and handheld devices. Further studies will elucidate which of these parameters impact the validity of results obtained through this novel method.

Keywords: Color Vision, Smartphone, Computer, Electronic Display, Handheld

Financial Disclosures: The authors had no disclosures.
Castleman Disease Presenting As Pseudotumor Cerebri Syndrome

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Introduction:
Castleman disease is a rare lymphoproliferative disorders with an unknown pathogenesis.

Methods:
A 28-year-old female complained of intermittent bilateral blurred vision for 2 months. Deny headache and tinnitus. No remarkable past medical history except loss of body weight for 5kg in the past half year.

Results:
On examination, slim and pale woman. BCVA were 20/20 OS and 60/200 OU with a right RAPD. Fundus examination showed bilateral severe elevated disc. Constricted visual field bilaterally on HVF. Brain MRI was grossly normal except enlarged optic nerve sheath space and herniated sella. Lumber puncture showed an increased intracranial pressure (>330 mmH2O) with slightly increase protein (71mg/dl). Routine blood test showed an otherwise normal result except slightly reduced RBC (3.26×1012/L) and Hb (108g/L). ESR was 35mm. CRP 8.6mg/L. ANA,ENA penal and C5 penal were normal. Enlargement of mediastinum was found by chest CT and then multiple enlarged lymph nodes were noticed. Biopsy was done on neck and mediastinum lymph node, bone marrow. Together with the increased serum level of total IgG, monoclonal spike k-and λ-, and β2-microglobulin levels, Castleman disease was diagnosed. Patient was put on chemotherapy (CHOP) without improvement, instead developing polyneuropathy. Then she was included in a Rituximab clinical trial and is now followed.

Conclusions:
Intracranial hypertension complicated in Castleman disease was not found in literature and the pathogenesis is not clear. CSF drainage disturbance due to increased central vein pressure caused by compression from the enlarged mediastinum is the suspected cause in this case.

References:

Keywords: Castleman Disease, Pseudotumor Cerebri Syndrome, Multiple enlarged lymph nodes

Financial Disclosures: The authors had no disclosures.
Diffusion Weighted Imaging (DWI) and Gadolinium Enhancement in the Differential Diagnosis of NAION and Optic Neuritis (ON)

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Introduction:
Although previous reports have evaluated the utility of DWI and gadolinium enhancement in the differential diagnosis of NAION and ON, none of these reports included a sufficient number of subjects to develop firm diagnostic imaging criteria. In this study, we proposed to review a larger series of cases to develop imaging criteria for the differential diagnosis of acute NAION and ON.

Methods:
We retrospectively reviewed patients evaluated at the Moran Eye Center with a diagnosis of NAION and ON from 2003-present. We included patients over the age of 18 with orbital MRI performed within 1 month of symptom onset. Neuroradiologists masked to the clinical diagnosis reviewed the DWI and gadolinium enhancement characteristics of the optic nerve.

Results:
594 charts have been reviewed to date and 52 patients have met criteria; 26 with NAION and 26 with ON. Diffusion restriction was seen in 9 cases (35%) of NAION and 13 cases (50%) of ON. Optic nerve enhancement was seen in 21 cases (81%) of NAION and 25 cases (90%) of ON. Of 9 NAION cases with diffusion restriction, 8 cases (89%) demonstrated abnormalities confined to the optic disc. Of 21 NAION cases with enhancement, 19 (90%) demonstrated enhancement confined to the optic disc. In none of the cases of optic neuritis was enhancement or diffusion restriction confined to the optic disc.

Conclusions:
Enhancement of the optic nerve is nearly always present in cases of both NAION and ON. However, in those cases of NAION in which enhancement is present, it is almost always confined to the optic disc. DWI characteristics alone cannot be used to distinguish NAION from ON. However, in cases in which diffusion restriction is present, confinement of the restriction to the disc is most consistent with a diagnosis of NAION. Additional data will be reviewed and analyzed prior to the Annual Meeting.

References:

Keywords: Optic Neuritis, Ischemic optic neuropathy, Diffusion weighted imaging, Gadolinium Enhancement

Financial Disclosures: The authors had no disclosures.
The Ability to Fake a Neurologic Field Defect

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Introduction:
To determine if volunteers can simulate and reproduce three automated visual field (Humphrey) (HVF) defects – hemianopia (HA), quadrantanopia (QA) and central scotoma (CS).

Methods:
Healthy volunteers, new to the HVF test, were randomized to a field defect (HA, QA or CS). After standard new patient instructions, they were shown a diagram of the defect to be simulated (a right temporal hemianopia, a right supronasal quadrantanopia and a right central scotoma). Two sets of HVF SITA Fast 24-2 fields were performed on the right eye with 5-10 minutes between tests. Three masked experts reviewed all the HVFs. If two of the three expert interpretations matched the attempted HVF defect, the simulation was deemed successful.

Results:
There were thirty volunteers randomized to ten HA, ten QA and ten CS. A HA was successfully simulated by all ten volunteers (100%) in the first and second HVF. A QA was successfully simulated by all ten volunteers (100%) in the first and nine (90%) in the 2nd HVF. A CS was successfully simulated by eight volunteers (80%) in the first and ten (100%) in the 2nd HVF. Reliability criteria were excellent. Forty-seven HVFs (78%) had zero fixation losses, forty-eight (80%) had zero false positives and forty-four (73%) had zero false negatives.

Conclusions:
It is easy to outwit the automated visual field (Humphrey) analyzer and simulate a reproducible and reliable neurologic field defect.

References:

Keywords: Humphrey visual field, Malingering

Financial Disclosures: The authors had no disclosures.
Spontaneous Cerebrospinal Fluid (CSF) Leak and Idiopathic Intracranial Hypertension (IIH)

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Introduction:
IIH is increasingly recognized as a cause of spontaneous CSF-leak in the ENT literature. The diagnosis of IIH is classically made a few weeks after surgical repair of the CSF-leak, because of occurrence of IIH symptoms.

Methods:
Case reports and literature review: Two young obese women developed spontaneous CSF rhinorrhea. The CSF-leak occurred via an empty sella in one, and cribriform plate encephalocele in the other. Both underwent successful surgical repair of the CSF-leak with resolution of headaches. A few weeks later, they developed chronic headaches and eventually, bilateral papilledema was noted. Lumbar punctures showed elevated CSF-opening pressures with normal CSF contents, with temporary improvement of headaches. CSF-shunting procedures resulted in temporary improvement of headaches, but one patient remained visually impaired.

Results:
There is a definite association between IIH and spontaneous CSF-leak based on the following observations: 1) Spontaneous CSF-leak patients are often young obese women; 2) Spontaneous CSF-leak patients sometimes develop raised ICP once the leak is repaired; 3) The rate of leak recurrence after surgical repair is much higher with spontaneous CSF-leak than with other causes of leaks, and permanent simultaneous/subsequent CSF-shunting procedure may be necessary to eliminate the leak; 4) Some patients with intracranial hypertension secondary to tumors, or even IIH, develop CSF-leak, confirming that raised ICP can cause CSF-leak; 5) Skull-base encephaloceles and empty sella turcica are often found in IIH patients as well as in CSF-leak patients.

Conclusions:
CSF-leak may occasionally keep IIH patients symptom free; however, classic symptoms/signs of intracranial hypertension often develop after the CSF-leak is repaired, exposing these patients to a high-risk of recurrence of the leak unless a CSF-shunting procedure is performed. Early identification of these patients is warranted to prevent unnecessary repeat surgical repair of the CSF-leak and to prevent visual loss from papilledema (by CSF-shunting procedure when appropriate.

References:

Keywords: Spontaneous Cerebrospinal Fluid Leak, Skull-base Encephaloceles, Idiopathic Intracranial Hypertension

Financial Disclosures: This study was supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York, and by the NIH/NEI core grant P30-EY06360.
Teaching Ophthalmoscopy to Medical Students (TOTeMS)

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Introduction:
Learning direct ophthalmoscopy is challenging, and medical students and physicians often perform this skill poorly. The purpose of this study was to determine medical student preferences for learning the ocular fundus examination and assess their accuracy using different examination modalities.

Methods:
First-year medical students (M1s) received training in direct ophthalmoscopy using anatomically- and optically-correct simulators and human volunteers. Subsequently, M1s were randomized to receive vs. not receive specific training on interpreting fundus photographs prior to accuracy assessments. Students’ preferences for each of the three methods and recognition of normal and abnormal fundus features were assessed.

Results:
119/138 (86%) M1s completed all required elements. For learning ophthalmoscopy, 85 (71%) preferred humans to simulators. For learning relevant features of the ocular fundus, 92 (77%) preferred photographs to simulators. Regarding accuracy, M1s correctly answered 3.7 more questions (out of 48 questions per modality) when using fundus photographs vs. using direct ophthalmoscopy on simulators (p<0.001). M1s receiving specific teaching about reading fundus photographs before testing accurately answered 1.7 more fundus photography questions (p=0.02). Regarding patient examination preferences, M1s rated median ease of examination 5/10 for humans, 7/10 for simulators, and 9/10 for photographs (10=easiest); and frustration 4/10 for humans, 3/10 for simulators, and 2.5/10 for photographs (10=most frustrated). Half the M1s reported they would attempt direct ophthalmoscopy ≤40% of the time during clinical rotations, and 84 (70%) would prefer to have fundus photographs instead of using the ophthalmoscope.

Conclusions:
Students preferred fundus photographs for both learning and examining the ocular fundus. Identification of ocular fundus features was more accurate on photographs compared to examination by direct ophthalmoscopy; this was further improved by training in interpreting fundus photographs. The increasing availability of non-mydriatic ocular fundus photography may allow replacement of direct ophthalmoscopy in many clinical settings for non-ophthalmologists.

Keywords: Ophthalmoscopy, Undergraduate medical education, Fundus photography

Financial Disclosures: This study was supported in part by an unrestricted departmental grant (Department of Ophthalmology) from Research to Prevent Blindness, Inc., New York, and by the NIH/NEI core grant P30-EY06360.
Poster 76

A Pocket-Sized Screening Tool for Higher Visual Processing Deficits

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Introduction:
Testing protocols for higher visual processing deficits during routine neuro-ophthalmic consultation have not been standardised. In-depth neuropsychology testing protocols are available, but are time intensive. The aim of this project was to create an easy-to-use, pocket-sized booklet containing tests sensitive for detecting errors in higher visual processing.

Methods:
Available tests covering a range of higher visual processing pathways, not covered by copyright, were examined for inclusion. Each test had to be easy to reproduce in print and easy to administer. Efforts were made to minimize cultural or ethical bias. All images have been tested on patients with visual deficits, dementia and normal controls. Multiple iterations of each test were progressively altered, so that the majority of normal patients could perform the tests accurately, while still being sensitive enough to detect abnormalities in those with higher visual processing difficulties.

Results:
A set of 16 tests, within 4 major categories, was chosen. Early visual processing tests included are: shape discrimination, shape detection, hue discrimination and size discrimination. Tests of object perception used are: fragmented letters and unusual views. The unusual views are matched with the usual views of the objects to test for real object recognition. Spatial perception tasks chosen are: dot counting, line bisection, letter scanning and cube analysis. Tests of facial perception include age judgment and recognition of famous faces. Tests of reading include crowded and un-crowded letters, as well as prose reading.

Conclusions:
We hope that this pocket-sized manual will allow clinicians to perform quick screening tests for higher visual processing difficulties in their office. The easier access to these tests may increase the rate of early diagnosis of conditions such as posterior cortical atrophy, and assist in guiding rehabilitation after brain injury and stroke.

Keywords: Clinical testing, Higher visual processing, Cortical

Financial Disclosures: The authors had no disclosures.
Poster 77

**Pupillometry of the Swinging Flashlight Test Detects and Quantifies Relative Afferent Pupillary Defects**

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**Introduction:**
Our purpose was to investigate the use of a novel computerized pupillometer as an alternative to the clinical swinging flashlight test to more objectively detect and quantify relative afferent pupillary defects (RAPDs).

**Methods:**
Binocular pupillary response curves were recorded (RAPDx pupillometer, Konan Medical USA, Inc.) in neuro-ophthalmology patients (n=31) with RAPDs clinically graded by an examiner and normals (n=31) with RAPDs that were simulated using neutral density filters (NDFs) and quantifiable dimmed light intensities. In fourteen normals, testing was repeated within 1-2 months. Pupillary constriction amplitude (CA), velocity (CV), and onset latency (COL) were used to calculate RAPDs.

**Results:**
RAPDs in normal subjects were 0.16±0.11 log units (LU) (range=0-0.38). Significant correlations were found between RAPD values and dimmed light intensity (Pearson’s $r=0.94$, $p<0.001$) and NDF strength ($r=0.81$, $p<0.001$). RAPDs increased linearly with increasingly dimmed light intensities for all 31 normals. In retested normals, inter-exam variability was 0.21±0.12 LU (range=0-0.42). Significant correlation ($r=0.88$, $p<0.001$) was found between clinician and machine grading of RAPDs in abnormal patients. Using the one-sided 95% confidence interval of the ratio of percentage change in CA (LU) as determined from normals, 20/22 (91%) abnormal patients with RAPDs >0.5 LU were distinguished from normals. In 6/6 abnormals, RAPDs were successfully neutralized using NDFs. RAPDs calculated using CA and CV correlated more strongly with the clinician’s grading compared to COL (Steiger’s test $p<0.001$).

**Conclusions:**
RAPD values recorded in normals were consistent with previously reported findings, and inter-exam differences demonstrate long-term variability in intrasubject RAPD. CA and CV are the most sensitive parameters for automated pupillometry. Sensitivity of detecting RAPDs in abnormals can be improved using the ratio of percentage changes in CA instead of differences in CA. RAPD values generated by this novel pupillometer correlated strongly with an examiner’s clinical grading, suggesting this device can be used as an objective diagnostic clinical tool.

**Keywords:** Diagnostic tests, Pupil, Pupillometry, Relative afferent pupillary defect, Swinging flashlight test

**Financial Disclosures:** The authors had no disclosures.
Retinal Nerve Fiber Layer Analysis of Cupping in Children Born Prematurely

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Introduction:
We performed a quantitative study the retinal nerve fiber layer (RNFL) thickness in children born prematurely with isolated optic nerve cupping.

Methods:
In this observational case series of six otherwise healthy premature children with isolated optic nerve cupping, RNFL thickness was measured via Cirrus Optical Coherence Tomography (OCT) Optic Disc Cube 200x200 protocol, and compared to normative results using T-test for statistical significance with Bonferroni correction when appropriate.

Results:
Nerve fiber layer measurements showed a statistically significant decrease in overall RNFL. Superior quadrant RNFL thinning was statistically significant and inferior, nasal and temporal nerve fiber layer thickness was decreased, although results were not statistically significant.

Conclusions:
Nerve fiber layer thinning in children born prematurely showing enlarged cup to disc ratios appears more consistent with optic nerve hypoplasia than glaucoma, as reflected by the markedly decreased average nerve fiber layer thickness without significant segmental changes. Although the optic nerve morphology in these cases is similar to juvenile glaucoma, it can be differentiated from glaucoma by diffuse retinal nerve fiber layer thinning on OCT imaging.

References:

Keywords: Pediatric ophthalmology, Hypoplasia, Glaucoma, Optical Coherence Tomography, Prematurity

Financial Disclosures: The authors had no disclosures.
**Poster 79**

**Horizontal Gaze Palsy with Progressive Scoliosis and Characteristic MRI Appearance**

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**Introduction:**
Horizontal gaze palsy with progressive scoliosis (HGPPS) is an autosomal recessive disorder typically associated with scattered mutations of the ROBO3 gene. The gaze palsy is congenital and is often accompanied by horizontal pendular nystagmus; strabismus has also been reported. It has been described since 1970 in families descending from Europe, the Middle East, and Asia. MRI findings include hypoplastic basis pontis, pontine tegmentum, cerebellar peduncles, and abducens nuclei. Diffusion tensor imaging and evoked potentials demonstrate deficient hindbrain axonal crossing.

**Methods:**
In this case report we describe a 9 year-old female with clinical signs and MRI consistent with HGPPS.

**Results:**
An intellectually intact 9 year-old female presented with torticollis, scoliosis, and nystagmus. Visual acuity was 20/40 OD and 20/30-2 OS, no RAPD was noted, and confrontational fields and color vision were normal. No ptosis or retraction were present. Slit lamp and fundus exams were normal. Pendular horizontal nystagmus was present in the primary position and marked right and left horizontal gaze paralysis and exotropia were present. Vertical gaze was intact. Past MRIs were reviewed, showing slightly prominent ventricles, a rectangular medulla, 4\(^{th}\) ventricle tenting and a split pons sign. Diffusion tensor imaging showed a smaller right-sided medullary white matter track, but as standard clinical number of directions were employed, it did not demonstrate clear lack of decussation. Genetic testing was consistent with HGPPS, showing a 3-nucleotide deletion of the ROBO3 gene. Both parents were heterozygous for the same, despite having no known familial ties.

**Conclusions:**
This patient’s diagnosis is to our knowledge the first reported in a family from the Dominican Republic. Reviewing past MRIs with focused attention allowed us to discover imaging findings consistent with HGPPS. Identification of the syndrome through clinical and MRI findings helps guide diagnostic testing and prognostic conversation with the family.

**References:**
4. OMIM entry 607313: Gaze palsy, familial horizontal, with progressive scoliosis; HGPPS

**Keywords:** Horizontal Gaze Palsy with Progressive Scoliosis, ROBO3, Nystagmus, Magnetic Resonance Imaging

**Financial Disclosures:** The authors had no disclosures.
Poster 80

The Natural History of Ocular Tics in Children

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Introduction:
Children with ocular tics are often evaluated in pediatric neurology and ophthalmology clinics. However, there is a paucity of literature regarding the natural history of ocular tics.

Methods:
The charts of all patients (aged 1-18 years) in a pediatric neuro-ophthalmology clinic diagnosed with ocular tics were retrospectively reviewed. Ocular tics were defined as eye rolling, blinking, and eye widening. Children with known secondary causes for tics at presentation were excluded. Telephone contact information was abstracted from the medical record and used to contact patients, parents, and/or guardians to obtain follow-up information.

Results:
Forty-three patients were included in the retrospective cohort, with a mean age of 7.8 ± 4.8 years. Seven (16.28\%) of the children also had non-ocular motor tics at presentation, while four children (9.30\%) had vocal tics. Two children (4.65\%) had both non-ocular motor tics and vocal tics at presentation. None of the children carried a diagnosis of Tourette syndrome or obsessive-compulsive disorder (OCD) at presentation; one child had attention deficit disorder (ADD). Thirty-two patients participated in the follow-up survey, with an average follow-up of 6.07 ± 3.91 years. Fourteen children (43.75\%) had persistent ocular tics at follow-up. Three patients (9.38\%) reported new non-ocular motor tics, while five patients (15.63\%) reported new vocal tics. Four children (12.50\%) developed both non-ocular motor and vocal tics during follow-up. One patient was diagnosed with Tourette syndrome by a physician during the follow-up interval (3.13\%), and three were diagnosed with ADD (9.38\%).

Conclusions:
Almost half (43.75\%) of children with ocular tics at presentation had persistence of ocular tics upon follow-up. New non-ocular motor and vocal tics occurred in several patients. These findings suggest that ocular tics in children are not always self-limited and suggest the possibility of a future diagnosis of Tourette Syndrome in such patients. Neurological follow-up may be warranted in children with ocular tics.

References:


Keywords: Ocular tics, Tourette syndrome, Pediatric

Financial Disclosures: The authors had no disclosures.
Poster 81

Visual Recovery after Decompression of Pituitary Macroadenoma

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Introduction:
This study aimed to examine the relationship between tumour size and visual function in pituitary macroadenoma patients and to investigate the clinical course of visual recovery after decompression of the anterior visual pathway.

Methods:
The 113 patients who were diagnosed with pituitary tumours and underwent primary anterior visual pathway decompression by a transsphenoidal approach were included. Preoperative and postoperative best-corrected visual acuity (BCVA) was measured, and visual field (VF) was assessed using automated perimetry. A neuroradiologist measured tumour size using a method based on the Cavalieri principle. The postoperative follow-up was divided into 4 periods: phase 1 (Surgery to 1 week), phase 2 (1 week to 1 month), phase 3 (1 month to 3 months), phase 4 (3 months to 6 months).

Results:
Thirty-seven patients were included, and preoperative visual loss was found in 21 (56.76%) patients. A BCVA recovery of more than 3 lines compared with preoperative baseline BCVA was found in 80% of patients over the following 6 months. In recovery of visual acuity, the most noticeable improvement manifests in the early fast phase (surgery to 1 week). The improvement of total mean deviation in the visual field was 30.89%, 24.73%, 16.08% and 20.05% between phases. The visual field improvement lasted over 6 months, and the most dramatic improvement occurred after 1 week of surgery during phase 2. In the inferior temporal quadrant, early fast improvement appeared from phase 1 to phase 2, however, the superior temporal visual field exhibited early and late phase recovery over 6 months.

Conclusions:
Tumour volume is a useful predictor for preoperative visual deficits in the pituitary macroadenoma patient. Restoration of visual acuity occurred immediately within 1 month after surgery. However, recovery of the visual field defect persisted up to 6 months. This late improvement demonstrates a potential period of late visual restoration.

Keywords: Pituitary adenoma, Visual acuity, Visual field

Financial Disclosures: The authors had no disclosures.
Introduction:
To study the relationship between the ophthalmoscopic appearance of optic nerve pallor and retinal nerve fiber layer (RNFL) thickness by optical coherence tomography (OCT).

Methods:
A retrospective review of records from all patients (n=231) who underwent OCT imaging at a tertiary neuro-ophthalmology practice from 2006-2012 was performed. Excluded were eyes with glaucoma/glaucoma suspicion, macular/optic nerve edema, pseudophakia and with myopia of >-6D. OCT scan was a 3.2 mm diameter circular cross-section centered on the optic nerve. RNFL thickness was defined using software embedded within the OCT instrument. Optic nerve appearance was assessed by stereoscopic viewing via slit lamp examination by an experienced neuro-ophthalmologist.

Results:
68 patients (131 eyes) were included with average age (mean±SD) of 50±15 years. Of these, 50 eyes (38%) showed optic nerve pallor and 52 eyes (39%) showed RNFL thinning (either sectorial or total average). The finding of optic nerve pallor had a specificity of 78% for the detection of RNFL thinning, although the sensitivity of this sign was only 63%. Average RNFL thickness in patients with RNFL thinning and associated optic nerve pallor was 68mm compared to eyes without pallor (76mm,P<0.05) or to eyes with normal optic nerve appearance and normal RNFL thickness (98mm,P<0.05). Sectorial RNFL loss was greater in patients with optic nerve pallor (45% of normal mean thickness) compared to those with normal appearing optic nerves (62%,P<0.05).

Conclusions:
Optic nerve pallor is a specific but not as sensitive sign of RNFL thinning. There is a suggestion of a level of RNFL loss below which pallor becomes ophthalmoscopically evident. RNFL thickness measurements may prove to be a valuable complement to the interpretation of optic nerve pallor in neuro-ophthalmologic diseases although clinical correlation with visual function may be necessary.

Keywords: OCT, RNFL, Optic nerve pallor

Financial Disclosures: The authors had no disclosures.
Introduction:
Neuromyelitis Optica (NMO) is an immune-mediated neurodegenerative disease of the central nervous system characterized by frequent demyelinating/necrotizing attacks of optic nerves and spinal cord leading to significant morbidity and mortality. Treatments are highly toxic and only minimally effective. Fifty percent of patients are blind in one eye or require a mobility aide within 5 years, with a 5-year survival of 68%. The immunological features of NMO, coupled with its severity make it an ideal candidate for a trial of autologous hematopoietic stem cell transplantation (AHSCT) in an effort to achieve remission and freedom from immunosuppressive medications. Our goal was to determine if NMO patients experience a reduction in relapses and associated disability without need for chronic immunosuppressant after AHSCT. We hypothesize => 50% reduction in the proportion of patients relapsing 3 years post-transplant.

Methods:
Patients 18-65 years, with 1 relapse in 12 months and 2 in 24 months, despite immunotherapy and EDSS <6.5 are eligible provided there are no contraindicated medical conditions. Patients undergo non-ablative stem cell mobilization and infusion using cyclophosphamide and rituximab. Outcomes including Expanded Disability Status Scale (EDSS) score, NMO antibody titers, optical coherence tomography and MRI are done every 6-12 months for 5 years. Ten subjects are required to provide 80% power for our primary outcome.

Results:
Two NMO patients have undergone transplantation. A 28 year old female was transplanted in May 2011 with pre-transplant annualized relapse rate (ARR) of 5 and EDSS 4.5, now 0 and 2.0 respectively. A 36-year old female was transplanted in April 2012 with pre-transplant ARR of 5, now 0. Neither maintenance nor rescue NMO therapy has been required.

Conclusions:
Thus far, this transplant regimen is well tolerated with dramatic clinical improvement in the short-term. Additional patients and longer follow-up will determine if these benefits persist.

Keywords: Neuromyelitis Optica, Autologous Hematopoietic Stem Cell Transplantation, Expanded Disability Status Scale, Optical Coherence Tomography, Magnetic Resonance Imaging

Financial Disclosures: The authors had no disclosures.
The Role of Vitamin D Status in Optic Neuritis - An Update

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Introduction:
The optic nerve is commonly targeted in demyelinating disease, providing a model for assessment of antiinflammatory and neuroprotective therapies. Vitamin D is a known ameliorator of inflammatory activity. In-vitro evidence suggests it may also be neuroprotective, an examinable possibility using optic neuritis (ON) and optical coherence tomography (OCT) in which degeneration is seen as early as 6 months. We aim to study the relationship between vitamin D status and neurodegeneration in ON. OCT will be used to study optic nerve retinal nerve fiber layer thickness (RFNLT) acutely and 6 months after ON. We hypothesize that vitamin D sufficient patients (25(OH)D > 80 nmol/L) will have relatively less RFNL thinning at 6 months suggestive of axonal preservation.

Methods:
50 patients with acute ON will undergo OCT testing to measure RFNLT, macular volume and inter-eye RFNLT difference, neurological examination and 25(OH)D testing at onset and month 6. Comparisons will be made on OCT measures between vitamin D sufficient and insufficient patients at these times.

Results:
Currently 39 of 50 patients have been enrolled; 25 having completed the 6 month study. Twenty-four patients were vitamin D insufficient and 12 sufficient (mean 25(OH)D 60 nmol/L and 105 nmol/L respectively). Gender, age, disease category and steroid treatment did not differ between groups. Acutely, vitamin D insufficiency was associated with greater RFNLT (136 vs 98 μm, p=0.052) and macular volume (10.3 vs 9.8 mm³, p=0.032). At 6 months, mean inter-eye RFNL difference (IED) was 12 vs 2 μm in vitamin D insufficient vs sufficient patients.

Conclusions:
Results thus far show ON in the context of vitamin D insufficiency is associated with greater optic nerve head edema acutely, and worse RFNL thinning (as measured by IED) at 6 months. These results suggest that vitamin D may be protective against both inflammatory and degenerative effects of demyelination.

Keywords: Vitamin D, Optic Neuritis, Neuroprotection, Inflammation, Optical Coherence Tomography

Financial Disclosures: Dr. Jodie Burton and Dr. Fiona Costello received support from the endMS Training Network (MS Society of Canada) to support student involvement in this project.
Changes in Fixation in Leber's Hereditary Optic Neuropathy: Comparison of Fundus Photographs and Automated Visual Fields (AVF) (Humphrey 30-2)

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Introduction:
Leber's hereditary optic neuropathy (LHON) initially affects central vision with a central scotoma on automated visual field (AVF). As LHON progresses, AVF usually reveals a shift of the scotoma, possibly as a compensatory strategy. We sought to quantify the shift in eccentric fixation using both color fundus photograph and AVF in affected LHON patients.

Methods:
Seven affected LHON patients (2 eyes from 6 patients and 1 eye from 1 patient, 13 eyes total) with abnormal AVF had their 50-degree color fundus photographs taken with an eccentric fixation protocol. This protocol included fundus images taken while patients fixated at the tip of a fixation stick. The stick was incorporated in the photograph to identify the fixation point in the retina. AVFs were processed to make the fields transparent. The center of the flipped AVF was superimposed over the tip of the stick in the photograph. We modified the size of the AVFs to fit the fundus photographs.

Results:
In all 13 eyes from 7 affected LHON patients, the tip of the fixation stick corresponded to an area in the retina away from the fovea (eccentric fixation). The average shift from the fovea was 5.1 degrees (range 1 to 12) at a mean of 19.26 months (range 13.1 to 33.5) from vision loss. When color fundus photographs were superimposed over AVFs, 10 out of the 13 eyes demonstrated a shift of fixation away from the fovea (affected area) to a more favorable region of the AVF.

Conclusions:
Eccentric fixation was identified in affected LHON patients using AVFs and quantitated with color fundus photographs. These results suggest that LHON patients use eccentric fixation to a more sensitive area in the retina as a compensatory mechanism. Understanding this adaptation may lead to new approaches in the rehabilitation of LHON patients.

Keywords: Leber's hereditary optic neuropathy, Eccentric fixation, Fundus photograph, Humphrey visual field

Financial Disclosures: The authors had no disclosures.
Optic Nerve Sheath Meningioma: Retrospective Study About 24 Cases

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**Introduction:**
The aim of this study is to highlight the complexity of the therapeutic management of Optic Nerve Sheaths Meningioma (ONSM).

**Methods:**
Retrospective study of 24 patients who were followed between 1988 and 2012. Inclusion criteria were primitive meningioma of the optic nerve sheath. Exclusion criteria were secondary meningioma. Visual acuity, eyes fundus, color vision, visual field, oculomotricity and imaging were studied several times during the follow up.

**Results:**
Twenty-four patients (20 women and 4 men were followed for a mean of 76.3 months (range, 12-273 months). The mean age at diagnosis was 42 years old (range, 21–64 years). Patients complained about any kind of visual impairment including visual loss, amaurosis, diplopia or orbital pain. The variability of symptoms has led to high latency of diagnosis, 0-76 months (mean latency 11.5 months). All 24 patients had clinical signs of optic nerve dysfunction in the affected eye, characterized by reduced visual acuity (19), visual field defect (22), optic disc atrophy or oedema (20) and relative afferent pupillary defect (19). In addition, 2 patients had proptosis and 4 had diplopia. Central scotoma was the most common presentation of visual field defect. Of the patients, in first therapeutic intention, 6 were observed only, 5 were first observed and had a modification of treatment in the follow up, 6 had surgery (one biopsy, 5 canal resections), 5 had intravenous steroid therapy and 2 received radiation. Visual acuity remained unchanged in 10 patients, decreased in 8 patients and improved in 6 patients.

**Conclusions:**
Observation should be the first therapeutic issue in patients with stable and good vision. In cases of intracanalicular ONSM, surgery still have his place. Vision, MRI and complication outcomes are favorable for using fractionated conformal radiotherapy, but the optimum timing of this therapy is still a complex question.

**Keywords:** Meningioma, Optic nerv, Treatment, Radiotherapy

**Financial Disclosures:** The authors had no disclosures.
Poster 87

Callosal Conundrum: Conflicting Perimetry Results after Corpus Callosotomy

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Introduction:
A 49 y/o male had a corpus callosotomy for intractable seizures. The patient was started on Vigabatrin, necessitating visual field testing. Confrontation visual field (CVF) testing revealed a left homonymous hemianopia (LHH). Paradoxically, automated visual fields (HVF 30-2) showed a dense right homonymous hemianopia (RHH). CVF requiring the patient to mimic the examiner’s fingers produced a crossed hemianopia — RHH when he used his left hand, and LHH when he used his right hand. These conflicting visual field defects were suspected to be an artifact of the patient’s disconnection syndrome. Additional visual field tests were performed to explore these “switching” homonymous defects. (The patient consented to videotaping. Portions of his examination will be shown.)

Methods:
HVF’s repeated with the response push-button in the right hand, left hand, and with verbal response all produced a RHH. Kinetic perimetry (Goldmann) performed on each eye and with both eyes open also produced a RHH. CVF with finger counting or hand comparison produced a LHH, both with a verbal response and push-button. However, CVF with a penlight, using either verbal or push-button response in either hand, showed a RHH.

Results:
The “switching” homonymous hemianopia did not appear to be the result of the response method (verbal vs. push-button) or the hand used (except CVF hand-mimicking). However, a light stimulus always produced a RHH, whereas hand comparison and finger counting produced a (less dense) LHH. We found only one report of this phenomenon in the literature (Kamaki, 1993).

Conclusions:
The side of the resultant homonymous hemianopia was dependent on the stimulus type. This suggests that the right hemisphere has some advantage in processing a light stimulus, whereas the left hemisphere does better with hand comparison and finger counting. Given the limitations of HVF in this patient, ERG and OCT are also being used to monitor retinal toxicity of Vigabatrin.

References:


Keywords: Visual field testing, Disconnection syndrome, Vigabatrin, Homonymous hemianopia

Financial Disclosures: The authors had no disclosures.
Poster 88

Ruptured Paraclinoid Aneurysm Presenting as Retrobulbar Optic Neuropathy

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Introduction:
Paraclinoid internal carotid artery (ICA) aneurysms arise between the roof of the cavernous sinus and the posterior communicating artery. Their rupture rate is estimated at 11.4-25%¹⁻³. Ruptured paraclinoid aneurysms typically present with subarachnoid haemorrhage; a new visual deficit as a presenting complaint is distinctly uncommon.

Methods:
A 48-year-old woman presented with sudden “darkening” of vision of the upper nasal aspect of her right eye vision and a generalized headache. She had poorly controlled hypertension. Neurological examination demonstrated an upper nasal field defect in the right eye and mild right-sided weakness (of which she did not complain). She was found to have subarachnoid haemorrhage from a ruptured right paraclinoidal aneurysm. After immediate therapy with dexamethasone, phenytoin, nimodipine, and hydrochlorothiazide, she was transferred to our institution where she noted further deterioration of her vision. Exam revealed no light perception in the right eye, with an amaurotic right pupil. Vision in the left eye was 20/20, and the fundus exam was normal in both eyes. Optic discs were normal, without edema or pallor.

Results:
Digital subtraction angiography (DSA) revealed bilateral paraclinoid ICA aneurysms. The right, symptomatic aneurysm measured 15x9x9 mm, arising distal to the ophthalmic artery and proximal to the posterior communicating artery. A pseudoaneurysmal component was seen. Endovascular therapy of the right-sided aneurysm was performed emergently with delivery of sixteen detachable microcoils. Post-treatment angiography showed complete aneurysm obliteration. Eight months later, vision in the right eye was hand motions in the temporal field of vision with right optic nerve pallor.

Conclusions:
The formation of a large pseudoaneurysm secondary to a ruptured non-traumatic saccular aneurysm, although rare, has been known to occur. By direct compression of the small peripheral vessels coming off the ophthalmic artery, the pseudoaneurysm was the most likely cause of the retrobulbar optic neuropathy.

References:

Keywords: Paraclinoid aneurysm, Pseudoaneurysm, Retrobulbar optic neuropathy, Subarachnoid hemorrhage, Vision loss

Financial Disclosures: The authors had no disclosures.
Bladder Cancer Metastases to the Extraocular Muscle

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Introduction:
To describe a case of an extraocular metastasis as the first presentation of metastatic bladder cancer.

Methods:
An 80-year-old male presented with complaints of diplopia for a period of two months. His vision was 20/30 bilaterally with normal intraocular pressures and no pupillary abnormality. His extraocular movements were measured at: -2 supraduction, -1 infraduction, -2 adduction, -3 abduction for the right eye, and full in all directions for the left eye. The right eye was proptotic, with an otherwise normal anterior and posterior ocular exam. His medical history was significant for bladder cancer and prostate cancer two years prior for which he had underwent radical cystectomy, prostectomy and chemotherapy.

Results:
A CT revealed thickening of the right lateral rectus muscle. The thickening appeared to extend into the tendinous insertion upon the globe with some resultant displacement of the optic nerve. An orbital biopsy was performed and immunohistochemical stains for CK-7 and CK-20 were found to be positive, consistent with metastatic bladder cancer. The patient developed increasing injection, proptosis, and discomfort of the right eye. He was subsequently treated with radiation to the right orbit with adjuvant chemotherapy. Several months later, he was found to have multiple sites of metastasis to his humerus, sternum, lung, and liver.

Conclusions:
Bladder cancer usually metastasizes to the lymph nodes, liver, lung, and bone. It is rarely noted to metastasize to the orbit. It is more prevalent in men and the prognosis is typically dismal. Treatment is palliative radiotherapy to reduce the orbital mass.

References:

Keywords: Orbital metastasis, Extraocular muscle, Bladder Cancer

Financial Disclosures: The authors had no disclosures.
Optic Chiasmal Abnormality On MRI In Ethambutol Induced Bitemporal Hemianopia

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Introduction:
Ethambutol induced optic neuropathy is known to cause central and cecocentral scotomas, but bitemporal visual field defects are also reported[1,2]. The exact pathogenesis and location of the lesion in bitemporal hemianopia is poorly understood. Some authors suggest chiasm toxicity [3], while retinal lesions are also proposed based on abnormalities on multifocal electoretinogram[4]. One study has shown focal axonal swelling without demyelination in the optic chiasm in rats exposed to ethambutol[5]. But, chiasmal abnormalities on imaging are not reported in cases of bitemporal hemianopia with ethambutol use.

Methods:
A 72 year old lady presented with a six-week history of progressive, painless, loss of vision in both eyes. Her visual symptom was noticed 4 months after commencing antitubercular treatment (ATT) for pulmonary atypical mycobacterial infection. Her regimen consisted of Ethambutol, Isoniazid, Rifampin, and Azithromycin. Her ATT was stopped immediately after she reported the visual loss. On examination her visual acuity was 20/200 in both eyes, with significant dyschromatopsia; sluggish, but symmetric pupillary light reflex and normal optic discs. Automated perimetry (Humphrey visual field analyzer) disclosed a bitemporal hemianopic defect, which prompted an MRI brain specifically to exclude a chiasmal lesion.

Results:
MRI of brain showed a symmetric hyperintensity within the chiasm on axial and coronal T2-weighted images, without any enhancement or diffusion abnormalities. Other investigations including CBC, serum electrolytes, autoimmune screen, Vitamin B12, Syphilis, VZV, HSV and HIV were unrevealing. CSF analysis is pending.

Conclusions:
Bitemporal visual field loss associated with Ethambutol toxicity and an abnormal signal in the optic chiasm on MRI signifies toxicity within the chiasm. To our knowledge, this is the first report of a chiasmal abnormality confirmed by MRI to be caused by Ethambutol.

References:


Keywords: Ethambutol, Chiasma, Bitemporal hemianopia, MRI, Toxicity

Financial Disclosures: The authors had no disclosures.
Introduction:
Optic disc anatomy is important. This is a study using OCT to measure the 12-6 dimensions in one neuro-ophthalmology practice and the implications.

Methods:
385 consecutive patients provided their demographic information by race first (Black, White, Asian), then they were asked whether they had Native American ancestry, whether they knew which tribe, then after that, what other nationality they were, from greatest to least. This was self-description with few genetic studies blood or mouth scrapings. The OCT (optical coherence tomography) machine used was the Zeiss Stratus in all cases, and the same technician did all of the studies. We used the 12 to 6 vector from “rapid optic disc” diameter. The largest of the 2 disc diameters was used. Papilledema and acute NAION data were excluded until the swelling was gone.

Results:
Diameters were seen from 1.1mm to 2.66mm. Diameters peaking at 1.9mm were seen in Blacks (50) and Whites (85) who had native American ancestries. The most common tribe represented was Cherokee. The 250 patients with no Native American ancestry were: 50 mostly German (peaked at 1.6mm), 30 mostly English (peaked at 1.4mm), 9 each Italian and Scottish, 7 Irish, 5 each German Jews, Dutch, Welsh and Polish.

Conclusions:
Disc diameters have many determinants, including Native American ancestry in Americans. This was not foreseen in German literature in the 1800s and early 1900s. It has an impact on glaucoma and neuro-ophthalmology conditions.

Keywords: Population genetics, Optic disc anatomy, Prognosis with disease entities

Financial Disclosures: The authors had no disclosures.
Etiology of Recurrent Isolated Sixth Nerve Palsies in Older Adults

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Methods:
Methods: In a retrospective review of the medical records database over a 10-year period in a neuro-ophthalmology clinic, patients ≥ 50 years of age with a diagnosis of isolated recurrent 6th nerve palsy were identified. Medical history and laboratory studies for risk factors for microvascular disease, including hypertension, diabetes, hypercholesterolemia, and tobacco use were noted. Laboratory data for thyroid disorders, myasthenia gravis, and other infectious and autoimmune disorders were also noted. MRI of brain and orbits w/ and w/o gadolinium and CTA or MRA of brain were reviewed.

Results:
Results: Five males and 2 females presented with isolated 6th nerve palsy that recurred over an average period of 6-12 months: 1) 4 had meningiomas located in the parasellar, petrous apex/cavernous sinus regions; 2) 1 had ophthalmoplegic migraine; 3) 1 had intracavernous carotid artery aneurysm; 3) 1 had hypertension and diabetes in microvascular disease.

Conclusions:
Conclusion: In this case series of 7 patients from review of a neuro-ophthalmic database over a 10-year period, structural lesions located in the parasellar or petrous apex cavernous sinus regions were associated with recurrent isolated 6th nerve palsies. Compression by a structural lesion was hypothesized to be related to the regeneration of the nerve to re-innervate the lateral rectus muscle for recovery of normal function. Hypertension/diabetes and ophthalmoplegic migraine were relatively less common etiologies of recurrent 6th nerve palsy in the older population.

References:

Keywords: Recurrent, 6th nerve palsy

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Poster 93

Long-Chain 3-Hydroxyacyl-CoA Dehydrogenase (LCHAD) Deficiency and Retinopathy. A Case Report: Over a 14-Year Period of Follow-up

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Introduction:
LCHAD deficiency is a rare disorder of mitochondrial long-chain fatty acid oxidation inherited as a recessive trait. Affected patients can present with hypoglycemia, liver dysfunction, cardiomyopathy, failure to thrive. Most patients suffer from retinopathy.

Methods:
A sixteen-year-old girl was initially diagnosed with LCHAD deficiency at seven months of age, when she suffered a coma with hypoglycemia and hepatomegaly. Appropriate diet modifications and fatty acid supplementations were instituted and regularly maintained. Retinopathy was noted when she was two years old. Physical, biochemical and ophthalmological assessments including visual acuity and ERG recordings were conducted over a fourteen-year period of follow-up. She underwent Zeiss OCT recordings when she was sixteen years of age.

Results:
Visual acuity was approximately 20/30 until she was eleven years old, and decreased gradually afterward. At the age of two, her fundi had showed a chorioretinopathy that remained stable until age eleven. At sixteen years of age, her retina appeared atrophic. Photophobia was noticed when she was eight whereas her photopic-ERG was nearly normal. Four years later, at age eleven, her Goldmann visual field was normal, whereas her photopic-ERG was decreased, her scotopic-ERG normal and her EOG flat. At sixteen, her visual acuity was decreased to 20/60, and her Goldmann visual field showed an annular scotoma. OCT showed a normal macular thickness with an abnormal transparency of retinal pigment epithelium and an atrophic aspect of choriocapillaris. Photopic and scotopic-ERGs were barely recordable.

Conclusions:
Despite regular dietary treatment and a good quality of life, there was progression of her retinopathy. Dysfunction of cones and retinal pigment epithelium appeared earlier than that of rods. The mechanisms of the progressive retinopathy have not yet been elucidated. They may be related to retinal ATP deficiency with cones having greater energy requirements than rods and/or possible RPE toxicity due to accumulated acylcarnitines.

Keywords: LCHAD-D, Fatty acid beta-oxidation defect, Progressive cone-rod retinopathy, RPE deficiency, L-carnitine

Financial Disclosures: The authors had no disclosures.
Is Pediatric Idiopathic Intracranial Hypertension on the Rise?

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Introduction:
Idiopathic intracranial hypertension (IIH) in children is an uncommon but significant cause of morbidity, including permanent visual loss. It is important to understand if, like obesity, IIH in children is on the rise and is related to that increase. The aim of this study is to compare the recent incidence rate of pediatric IIH in a tertiary care hospital to earlier data published from that same hospital.

Methods:
Charts of all children aged 2 to 16 diagnosed with IIH at the IWK Health Centre in Halifax between 1997 and 2007 were retrospectively reviewed.

Results:
Twelve cases (5 males, 7 females) fulfilling all diagnostic criteria for IIH were identified, for an annual incidence of 0.6 cases per 100,000 children with no gender predilection (p=0.51). Obesity was noted in 75% of patients examined (9/12). Children older than 12 years were more likely to be obese (6/6, 100%) compared to those under 12 years (3/6, 50%).

Conclusions:
The calculated incidence is lower than that found in an earlier study for the same geographic region (0.9 cases per 100,000 children) despite increasing obesity. However, a higher association of obesity after puberty suggests that post-pubertal IIH more closely resembles the adult disease and therefore could represent yet another significant problem of the obesity epidemic. The decreasing rate of pediatric IIH could be explained by better diagnostic tools that continue to narrow the diagnostic criteria. The true association between pediatric IIH and obesity may best be elucidated by a multicenter prospective study.

References:

Keywords: Idiopathic intracranial hypertension, Obesity, Pediatrics

Financial Disclosures: The authors had no disclosures.
**Poster 95**

**Multi-Modal Imaging of Nasal Hypoplasia of the Optic Disc**

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**Introduction:**
Nasal optic nerve hypoplasia (NOH) is a rare anomaly characterized by a wedged shaped inferior temporal visual field defect emanating from the blind spot, hypoplasia of the nasal disc and variable acuity. It can be unilateral or bilateral and is non-progressive\(^1\). No association has been recognized with intrauterine drug exposure or neurologic abnormality. In the literature, there are four articles reporting 15 patients with NOH, so it is rarer than other forms of optic nerve hypoplasia\(^2,3,4\).

**Methods:**
Case report with spectral domain OCT (sdOCT).

**Results:**
A 30 year-old female noted the onset of a fixed blind spot in the temporal visual field of her left eye following a flu-like illness. Acuity was 20/20 OU. AO-HRR color plates were 6/6 OU and the pupils were normal. Humphrey perimetry OS revealed the left blind spot was enlarged. These findings suggested acute idiopathic blind spot enlargement syndrome (AIBSES). However a wedge- shaped inferior temporal visual field defect emanating from the blind spot and breaking out to the temporal periphery was seen on kinetic perimetry (Goldmann visual field). Re-examination of the fundus revealed nasal optic nerve hypoplasia OS and a mildly anomalous right disc. Full-field and multi-focal ERG was normal. Superior nasal peripapillary retinal nerve fiber layer thinning was seen OS on sd-OCT with the integrity of the outer retina layers maintained.

**Conclusions:**
Nasal optic nerve hypoplasia (NOH) was first described by Buchanan and Hoyt in 1981, when they reported the characteristic visual field defect in three patients with normal visual acuity who were noted to have NOH\(^5\). Our patient's presentation initially suggested AIBSES because of the recognition of the visual field defect following a flu-like illness and the enlargement of the blind spot on central threshold perimetry. However, sdOCT in this case ruled out AIBSES and demonstrated the absence of the superior nasal nerve fiber layer.

**References:**

**Keywords:** Optic Disc Hypoplasia, Nasal Optic Disc Hypoplasia, Visual field defect

**Financial Disclosures:** Donald Hood PhD: Grant from Topcon, Inc.
Ethambutol Optic Neuropathy in a Hemodialysis Patient on Guideline-Recommended Doses

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Introduction:
Ethambutol is a first-line treatment in tuberculosis. It is primarily excreted by the kidneys and is minimally cleared by hemodialysis. International tuberculosis treatment guidelines recommend adjusting the dosing regimen to 15-25 mg/kg 3 days/week in patients receiving maintenance hemodialysis.

Methods:
Case report.

Results:
A 79 year-old woman with pulmonary tuberculosis and end-stage renal disease on hemodialysis presented with progressive visual difficulty occurring over 4 weeks approximately 6 months after starting ethambutol 20mg/kg 3 days/week. The diagnosis of ethambutol toxic optic neuropathy was based upon bilateral decreased visual acuity, impaired color vision, central scotomas on formal visual field testing, and increased latency of the P100 waves on visual evoked potential testing.

Conclusions:
We report the first known case of ethambutol toxic optic neuropathy occurring in a patient receiving maintenance hemodialysis despite receiving guideline adjusted doses.

References:

Keywords: Ethambutol, Optic neuropathy, Hemodialysis

Financial Disclosures: The authors had no disclosures.
Poster 97

From William Harvey to Harvey Cushing: Incorporating Emissary Veins on the Cusp of the Final Frontier

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Introduction:
To explain recently described ophthalmic and cerebral findings, characterized by mild papilledema, globe flattening, choroidal folds and hyperopic shift, in astronauts after long duration space missions.

Methods:
Review of cephalic venous anatomy and cerebrospinal fluid physiology.

Results:
Overlooked in proposals to date has been the emissary and valveless nature of the cephalic venous system. Due to the head’s position above the heart, and the lack of contractile elements within the cranium, the return of cephalic blood to the heart is ordinarily assisted by gravity. Since the head contains different compartments, an increase in overall cephalic venous pressure in the absence of gravitational assist will manifest in different ways.

Conclusions:
Under conditions of so-called microgravity in space, relative pooling of blood will occur in the dural sinuses with corresponding increases in cerebrospinal fluid pressure. While the resulting pressure elevation remains uniform within a rigid cranium and causes minimal anatomic distortion, anterior to the optic canal where the central nervous system is not fully encased by rigid structures, but anteriorly is exposed to surrounding atmosphere, pressure-gradients can develop with expansion and distortion of both ocular and orbital contents. The pressure gradient between venous lumen and surrounding tissues reaches its peak at the interface of the head with surrounding atmosphere. Venous transudate will be greatest, whilst lymphatic drainage will be reduced by slowed external jugular venous drainage, manifesting as chemosis as well as epicranial swelling. Upon return to normal gravity environment, cephalic venous return to the heart promptly normalizes, with corresponding changes in cephalic pressures.

Keywords: Intracranial hypertension, Space flight, Emissary veins, Hyperopic shift, Neuroanatomy

Financial Disclosures: The authors had no disclosures.
Introduction:
Pediatric idiopathic intracranial hypertension (IIH) may be identified in patients with clinical symptoms including headache and blurry vision and signs of papilledema on exam. Little is known about the clinical features and outcomes in asymptomatic IIH patients. The purpose of this study was to define the clinical features of asymptomatic patients.

Methods:
A ten year retrospective chart review of all patients with a diagnosis of IIH (ICD-9 348.2) at a single, tertiary care center was performed. Data including clinical characteristics and complete ophthalmic findings were recorded.

Results:
The incidence of asymptomatic IIH was 19% (13/68) and 77% (10/13) of these patients were male. Asymptomatic patients on average were younger than symptomatic patients (9 yrs versus 13 yrs, P=0.05) and male gender was associated with the absence of symptoms (P=0.002). Only two patients had a BMI > 25 kg/m². Mean opening pressure did not differ significantly between symptomatic and asymptomatic groups (37 cm H20 versus 34 cm H20, P=0.17). Chronic optic nerve edema was noted in 54% (7/13) of asymptomatic patients and permanent vision loss in one. One asymptomatic patient was treated with serial lumbar punctures and two required definitive surgical intervention.

Conclusions:
Asymptomatic IIH can be associated with significant morbidity including vision loss, chronic optic nerve edema, and the need for surgical intervention. Pediatric patients with IIH and papilledema may be asymptomatic, young and male. The clinician should not assume in these patients that the diagnosis is pseudopapilledema but rather pursue an evaluation to confirm the diagnosis.

References:


Keywords: Idiopathic intracranial hypertension, Papilledema, Pediatric

Financial Disclosures: The authors had no disclosures.
Poster 99

The Paroxysmal Ocular Tilt Reaction - Jerk See-Saw Nystagmus: A Delayed Sequela of Meso-Diencephalic Hemorrhage

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Introduction:
The ocular tilt reaction (OTR) consists of skew deviation with both conjugate ocular torsion and head tilt toward the lower eye [1-8]. It is thought to be secondary to injury of the otolith-pathway which runs from the ear to the midbrain, crossing in the pons [1,2]. Paroxysmal OTR (POTR) follows hemorrhagic meso-diencephalic lesions after a delay and appears to be irritative, responding to drugs that suppress abnormal neural firing. We report a case of delayed onset of POTR following a meso-diencephalic hemorrhage from a presumed cavernous malformation.

Methods:
Clinical case presentation which includes video recording of POTR, fundus imaging and neuro-imaging.

Results:
A 51 year-old male suffered a hemorrhagic stroke of the right midbrain and thalamus two years prior to presentation. Six months later, he developed abnormal eye movements and oscillopsia. He exhibited paroxysms of left hypertropia accompanied by conjugate clockwise ocular torsion (patient viewpoint) and variable head tilt. During paroxysms, he exhibited small amplitude contraversive torsional nystagmus. Occasionally, his paroxysms would occur rhythmically as in jerk see-saw nystagmus. Vertical saccades were slowed between paroxysms. Neuro-imaging revealed a right-sided thalamic-midbrain lesion with features typical of cavernous malformation and old hemorrhage, extending inferiorly to the level of the red nucleus. Symptoms were mildly improved with oral baclofen.

Conclusions:
This case demonstrates delayed development of POTR/jerk see-saw nystagmus following meso-diencephalic hemorrhage. These observations support a previously presented theory: an excitatory lesion in the interstitial nucleus of Cajal produces an ipsiversive POTR via episodic hyperactivity of neurons involved in vertical and torsional eye movements [1-6]. Inhibition of neuronal activity with baclofen provides some relief, thereby supporting the theory that POTR involves excitatory irritation of neurons.

References:

Keywords: Ocular tilt reaction, Nystagmus, Skew deviation, Interstitial nucleus of Cajal

Financial Disclosures: The authors had no disclosures.
Vasospastic Amaurosis Fugax Due to Ophthalmic Artery Vasospasm

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Introduction:
Vasospastic amaurosis fugax (VAF) is a rare clinical entity in which there is recurrent transient unilateral visual loss due to presumed retinal artery vasospasm (RAV). The visual loss can last from seconds to few hours and most patients have negative visual phenomena. Treatment with Calcium-channel blockers may be an effective treatment in these cases (1, 2). We will present a unique case of VAF due to ophthalmic artery vasospasm (OAV) that has different clinical features then RAV, and responded to different treatment. Review of literature shows it to be the first angiography proven case of VAF due to OAV.

Methods:
Case presentation of a healthy fifty seven years old male with sudden recurrent episodes of reduce vision and multiple faint speckles in his right eye that lasted between 5-10 hours.

Results:
Clinical examination during visual attacks showed right eye vision to decrease from 20/20 to 20/50 and a right eye RAPD appeared. Fundus examination was normal without retinal artery spasm but a fluorescing angiography (FA) showed much delayed segmental choroidal and central retinal artery filling. The exact FA features were seen in repeated visual attacks. Laboratory evaluation for inflammatory diseases and hypercoagulability were normal. Carotid doppler was normal. Brain MRI, CT angiography and a cerebral angiography did not show a structural defect. There was increased right ophthalmic artery velocity on transcranial doppler and mild hypertension was found. Treatment with anti-platelet or increasing doses of calcium-channel blockers had only partial and temporary effect. Treatment with inhalation of oxygen stooped the visual attacks.

Conclusions:
We describe the first imaging proven case of vasospastic amaurosis fugax due to ophthalmic artery spasm. The difference in clinical features and respond to treatment suggest a different mechanism then of retinal artery vasospasm.

References:

Keywords: Vasospastic, Amaurosis fugax, Ophthalmic artery, Vasospasm, Fluorescing angiography

Financial Disclosures: The author had no disclosures.
Introduction:
Central nervous system cavernous malformations affect roughly 0.5% of the population and can occur anywhere in the brain and spinal cord. Cavernous malformations affecting the optic pathway are extremely rare. Patients with these lesions typically present with acute headaches and sudden loss of vision from chiasmal apoplexy. We present a case of sudden vision loss due to a cavernous malformation affecting the optic nerve, optic chiasm and optic tract. Only 9 similar cases have been reported in the literature.

Methods:
Case report.

Results:
A 21 year-old male presented with sudden loss of vision and acute headaches. Goldmann perimetry demonstrated an incongruous left homonymous heminaopia. His CT head showed a hyperdense lesion involving the proximal right optic nerve, optic chiasm and right optic tract. An MRI brain demonstrated mixed T2 signal and enlargement of the right optic nerve, optic chiasm and right optic tract with minimal enhancement after administration of gadolinium. Pituitary function testing was within normal limits. His inflammatory and infectious work-up was negative. The patient was initially managed with intravenous steroids in hospital for one week without a change in his vision occurring. Open biopsy of the posterior aspect of the chiasm revealed a surface hemorrhage overlying a cavernous hemangioma affecting the right optic nerve, optic chiasm and right optic tract. The patient noted improvement of his vision after the biopsy.

Conclusions:
Apoplexy from cavernous malformations affecting the optic pathway is a rare cause of acute vision loss, but should be considered in the differential diagnosis of lesions that affect the optic nerve, optic chiasm and optic tract.

Keywords: Cavernous hemangioma, Optic Nerve, Optic Chiasm, Optic Tract

Financial Disclosures: The authors had no disclosures.
Ophthalmic Ischemia Secondary to Moyamoya Syndrome in Neurofibromatosis Type 1

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Introduction:
Moyamoya is a syndrome described of stenosis of the large intracerebral arteries and resultant formation of fine vascular collateral vessels at the base of the brain. Ischemia results from hyperplasia of smooth muscle cells and luminal thrombosis of the proximal internal carotid arteries thereby affecting the middle and anterior cerebral and ophthalmic arteries. Moyamoya exists as a primary or secondary disorder from a variety of etiologies including Neurofibromatosis type I (NF1), as illustrated in the following case. Ophthalmologic manifestations of Moyamoya include morning glory disc anomaly, optic nerve hypoplasia, chorioretinal coloboma, sphenopharyngeal meningocele, ocular ischemic syndrome, anterior ischemic optic neuropathy, and central retinal vein and artery occlusions.

Methods:
We present the case of an infant with known NF1 who was able to fix and follow with either eye independently but objected more strongly to occlusion of the left eye. An afferent pupillary defect was present in the right eye. Fundoscopic exam demonstrated a pale optic nerve, blunted foveal reflex, geographic chorioretinal scarring and atrophy nasally and infranasally extending to the optic disc with peripheral vascular obliteration beyond arcades in a 360 degree fashion in the right eye. The left eye demonstrated small corkscrew retinal vessels, normal foveal reflex and normal optic nerve.

Results:
Fluorescein angiography (FA) of the right eye showed delayed filling with extensive vascular tree pruning beyond the arcades. Retinal atrophy was evident nasally with prominent choroidal vessels. No neovascularization is noted. The FA of the left eye demonstrated corkscrew vessels in the superior periphery with no leakage. Cerebral angiography showed distal right internal carotid artery and proximal middle cerebral artery stenosis with early collateral vascularization. MRI showed hemiatrophy of the right hemisphere from ischemia.

Conclusions:
Findings are consistent with ophthalmic artery vascular occlusion secondary to Moyamoya syndrome, a known complication of NF1.

Keywords: Neurofibromatosis type 1, Moyamoya Syndrome, Ocular Ischemia

Financial Disclosures: The authors had no disclosures.
Poster 103

Idiopathic Intracranial Hypertension In An Adult Female Following A Minor Head Trauma

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Introduction:
Idiopathic intracranial hypertension (IIH) is a clinical syndrome characterized by elevated intracranial pressure in the absence of space occupying lesions or other brain disorders. The elevated pressure typically results in headaches, pulsatile tinnitus, and visual symptoms. Similarly minor head injury can lead to non specific headaches, tinnitus and visual blurriness. Intracranial hypertension (IH) is more commonly a sequel of moderate and severe head injury.

Methods:
We discuss a case of progressive IIH with papilledema (PE) following minor traumatic head injury

Results:
A 24-year-old- female presented with headaches, visual disturbances, and “whooshing sound” after she was involved in a low speed motor vehicle accident resulting in minor head injury. The clinical presentation was correct for a IIH with bilateral PE. Body Mass Index was 19.29 kg/m2. Magnetic resonance imaging (MRI) supported IIH. Magnetic Resonance Venogram (MRV) was unrevealing. After failing Acetazolamide she was treated with serial lumbar punctures and a temporary lumbar drain with complete resolution of headache. Her PE resolved. Optical coherence tomography showed no evidence of nerve fiber loss.

Conclusions:
IIH can result from trauma related venous sinus thrombosis resulting in venous flow obstruction. No such findings were appreciated on imaging. Perhaps the minor head injury resulted in disruption of the fluid dynamics in the brain. Interestingly spontaneous remission was achieved with temporary lumbar drain, further supporting role of previous head injury. We underscore importance of recognizing signs and symptoms of IIH in patients with headache after minor head injury, even though whiplash injury symptoms and tension headaches are higher on the differential. We highlight that MRI and MRV of the brain are valuable in recognizing common secondary causes of secondary IH after traumatic brain injury. Early recognition of IIH through a thorough clinical evaluation is paramount in preventing permanent visual impairment.

References:

Keywords: Idiopathic intracranial hypertension, Papilledema, Traumatic head injury

Financial Disclosures: The author had no disclosures.
Extra-Axonal Ocular Damage Quantification by Multifocal Electroretinography in Chronic Progressive Multiple Sclerosis

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Introduction:
Immune mechanisms in multiple sclerosis (MS) are more complex than previously thought, are mediated by both T and B cells, and produce demyelination and axonal pathology in both grey and white matter.¹ Previous studies have focused on optical coherence tomography (OCT) measurements of retinal nerve fiber layer (RNFL) thinning corresponding to visual deterioration in MS.² More recent findings, however, indicate retinal pathology involving the inner and outer nuclear layers as well (e.g., bipolar and Muller cells, and photoreceptors).³,⁴ Because multifocal electroretinogram (mfERG) measures inner and outer retinal layer function, this may be indicated as a useful technique to detect and follow inner and outer retinal dysfunction in MS.

Methods:
This is a case report with an English PubMed literature search.

Results:
A 60-year old white female with a 15-year history of MS was referred for routine ocular examination (asymptomatic from a visual standpoint). Best corrected visual acuity was 20/20 in both eyes with 1+ left relative afferent pupillary defect. Dilated ophthalmoscopic examination showed normal appearing optic nerves, maculae, and vasculature in both eyes. OCT revealed overall RNFL thinning in left eye (79 μm) greater than right eye (85 μm), as well as maculopapillary bundle thinning greater in left eye (51 μm) than right eye (61 μm (borderline)). MfERG in both eyes revealed attenuated foveolar and peri-foveolar responses in both eyes.

Conclusions:
The mechanism of visual loss may arise from secondary axonal degeneration after an optic nerve insult, or from primary retinal involvement.⁵,⁶ MfERG detects discrete areas of macular retinal dysfunction and has only been briefly reported in select MS patients as a diagnostic technique to demonstrate inner and outer retinal damage, but has not been widely studied in this population.⁷ This report demonstrates the utility of mfERG for studying retinal pathology in MS patients and highlights that larger, prospective studies are needed.

References:

Keywords: Multiple Sclerosis, Chronic Progressive Multiple Sclerosis, Optical Coherence Tomography, Multifocal Electroretinogram

Financial Disclosures: The authors had no disclosures.
The Prognosis of Functional Visual Loss in Children and efficacy of bottom up refraction

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

Methods:
Retrospective medical record review was performed in 45 patients who were 4 to 16 years old, diagnosed with FVL between April 2006 and April 2012. The Bottom-up refraction is a visual acuity (VA) test starts from the smallest letter with suggestion.

Results:
Eighteen male patients and 27 female patients included and the mean age was 9.62±0.42 years old. The mean best corrected VA at baseline was 0.48±0.04 (Snellen acuity). The Bottom-up refraction was performed in 37 patients and 31 patients (83.8%) were proved to have normal vision. The 40 patients were followed up and 36 patients (90%) of them were recovered to normal vision or lost the symptoms.

Conclusions:
The bottom up refraction is very useful in proving non-organicity in children with FVL. The prognosis of FVL in children is excellent and most of them are recovered spontaneously.

Keywords: Functional visual loss, Non-organic visual loss, Bottom up refraction, Prognosis

Financial Disclosures: The authors had no disclosures.
Ocular Neuromyotonia: Inducible Superior Oblique Myotonia

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Introduction:
Superior oblique myotonia is a rare disorder known to occur following skull-base radiation therapy, most commonly related to pituitary/parasellar malignancy. We present a case of inducible right-sided superior oblique myotonia starting one month after radiation therapy for right posterolateral frontal lobe anaplastic oligodendroglioma with video documentation. A similar case was not able to be found in the literature.

Methods:
Retrospective case report.

Results:
32 year old man with history of seizure disorder now status-post resection of right-sided posterolateral frontal lobe glioblastoma multiforme followed by 2 months of external-beam radiation therapy (60 Gy total over 30 fractions) and concurrent temozolamide who noted intermittent binocular vertical and torsional diplopia which was precipitated by looking down and left. The attacks lasted less than 30 seconds but were interfering with his ability to work as a store manager. On exam his vision was 20/20 OU, equal pupils without RAPD. No ptosis or exophthalmos. EOMs and confrontational visual fields were full. The patient was initially orthotropic with trace exophoria on alternate cover. After looking down and left for a few seconds, the patient induced binocular diplopia with left hypertropia made worse by looking up and left. The episode was captured on video using a cell phone in the clinic. Recent MRI demonstrated normal post-operative changes to right frontal lobe with normal course of CN IV. The patient was previously taking levetiracetam for seizure control which was changed to oxcarbazepine with significant improvement in frequency of episodes.

Conclusions:
Clinical exam was consistent with intermittent, inducible superior oblique myotonia OD secondary to posterolateral frontal lobe radiation therapy. Video documentation provides a clear demonstration of motor deficits. Oxcarbazepine is useful for both seizure control and improvement in ocular myotonia.

References:

Keywords: Neuromyotonia, Superior oblique, Diplopia, Ocular myotonia, Oxcarbazepine

Financial Disclosures: The authors had no disclosures.
Poster 107

Significance of Anisocoria in the Inpatient Setting

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Introduction:
Anisocoria and other pupillary abnormalities are common findings among inpatients, particularly in the setting of trauma. This review evaluated the consults that requested assessment of pupillary abnormalities within a 13 month time period at a level I trauma center. Specifically we sought to determine the frequency in which anisocoria heralded a catastrophic neurological event.

Methods:
A review of all adult and pediatric inpatient consults placed to the ophthalmology and neurosurgery services at a level 1 trauma center between the dates of 6/1/10 and 6/30/11 was done. Patients for whom consults were placed included those in the inpatient adult, pediatric and psychiatric hospitals as well as those in the emergency department. All consults that mentioned any pupillary abnormality were reviewed. The etiology for all pupillary abnormalities was obtained through review of the consult note and any additional follow up visits the patient may have had.

Results:
There were 51 consults requested from ophthalmology and 31 consults requested from neurosurgery to evaluate pupillary abnormalities. Of the consults placed for ophthalmology, the most common cause of pupillary abnormality was traumatic mydriasis. Of the consults placed for neurosurgery the most common cause of pupillary abnormality was shift or herniation secondary to brain injury. There were no cases of a catastrophic neurological event causing a pupillary abnormality in the absence of altered mental status or other focal neurological deficits.

Conclusions:
The presence of a pupillary abnormality can portend a catastrophic neurological event. Inpatient primary care providers including emergency medicine, internal medicine and trauma physicians routinely request evaluation of anisocoria. The patient’s neurological status and historical information can help the primary team to determine the etiology and significance of a pupillary abnormality.

Keywords: Anisocoria

Financial Disclosures: The authors had no disclosures.
Cyclic Oculomotor Nerve Paresis Plus

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Introduction:
Cyclic oculomotor nerve paresis is extremely rare and its exact pathophysiology remains mysterious. Based on electromyographic and electron microscopic evidence in two cases, Kommerell et al. suggested in 1988 that chronic damage to the oculomotor nerve may play a role in its pathogenesis. A well-documented example of this entity is being presented in a boy who in addition was diagnosed with hereditary motor and sensory neuropathy (Charcot-Marie-Tooth disease type 1).

Methods:
We describe a 14-year-old boy with cyclic oculomotor nerve paresis who has been followed at our institution since early childhood. Repeated orthoptic, photographic and video documentations were performed during follow-up.

Results:
The patient developed first signs of left oculomotor nerve paresis at the age of ten months. The paresis slowly progressed to become virtually complete by the age of four years. Five years later first cyclic spasms were observed: Each spastic episode lasting 10-15 seconds consisted of co-contraction of all affected extraocular muscles, miosis and accommodation and was preceded by involuntary twitching of the upper lid with a periodicity of 60 to 100 seconds. A film will be shown that illustrates the characteristics of this patient’s rare disorder.

Conclusions:
To our knowledge this is the first report of a concurrent cyclic oculomotor nerve paresis and hereditary motor and sensory neuropathy in the same individual. We cannot rule out a pure coincidence albeit it is conceivable that the underlying neuropathy renders the oculomotor nerve more susceptible to damage.

References:


Keywords: Cyclic oculomotor nerve paresis, Charcot Marie Tooth Disease, Hereditary motor and sensory neuropathy

Financial Disclosures: The authors had no disclosures.
Primary CNS Lymphoma Initially Presenting as Progressive Visual Loss

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Introduction:
Primary intraocular lymphoma (PIOL) is a rare subset of primary CNS lymphoma (PCNSL).

Methods:
A 53-year-old male carpenter complained sequentially progressive bilateral vision loss for 6 months.

Results:
Eye examination showed BCVA 20/30 OD and 20/200 OS, with bilateral vitreous opacity, KP and an unsatisfied fudus examination. Patient was diagnosed as “uveitis” and was treated with topical steroid. Left limb weakness developed and patient was referred to Neurology Department. Patient was found disorientated, barely cooperative, a central facial, tongue, left limb paralysis and bilateral Babinski’s sign (+). Blood test showed a normal panel except slightly increased WBC. Brain MRI showed diffuse, multiple lesions affecting brainstem, basil ganglion and hemisphere. Lumber puncture showed elevated pressure, protein, MBP and lymphocyte counts. Patient was put on high dose IV-MP pulse and did not show any improvement. Biopsy of temporal lobe and vitreous were performed and the diffuse Large-B cell lymphoma was diagnosed. Patient quit treatment and died after a few weeks.

Conclusions:
Vitritis/vitreous opacity are common findings which could be easily misdiagnosed as uveitis in PIOL. Even it is usually diagnosed by biopsy, it could possibly diagnosed clinically by slit lamp/eye exam. As a high-grade, malignant tumor with frequently diffuse Large B-cell lymphoma histologically, early detection and dedicated treatment may improve vision and survival.

References:


Keywords: Primary intraocular lymphoma (PIOL), Primary CNS lymphoma (PCNSL), Vitreous

Financial Disclosures: The authors had no disclosures.
The Effect of Visual Field Loss on Driving Ability

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Introduction:
In many places, persons with hemianopias are not allowed to operate motor vehicles or obtain a driver’s license. However, there is controversy in this legislation; some studies have shown that people suffering from a hemianopia can in fact drive safely.¹ This prospective study aims to characterize how driving ability is affected by different types of hemianopia and quadrantanopia.

Methods:
We used an A+B Software Driving Simulator: Simuride Home Edition with 10 test subjects. Subjects used spectacles with parts of the visual field blocked out in each lens. The visual field defect glasses included right superior, right inferior, left superior, and left inferior quadrantanopias as well as right and left hemianopias. The control glasses had no visual field defects. The subjects became accustomed to the driving simulator by driving a specific test course twice without the study glasses. They then repeated the course with each pair of glasses in random order. Driving infractions (crashing into other cars or curbs, running red lights or stop signs, speeding, etc.) were recorded as well as the amount of time to complete the test course. A subjective score from 1 (apprehensive) to 3 (comfortable) was also recorded after each drive.

Results:
Driving ability (number of infractions, time elapsed, and subjective score) was most affected by left sided visual field defects, especially the left superior quadrant.

Conclusions:
We believe the left visual field, especially the left superior quadrant, may be the most vital part of the visual field concerning driving. This may be because the driver is sitting on the left side of the vehicle (USA). In this way, opposing traffic moves into the driver’s left field of vision. The small number of subjects in this study may limit these results. Further experimentation should be conducted to confirm these results using larger numbers of subjects.

References:


Keywords: Hemianopia, Quadrantanopia, Driving Ability, Visual Field Defects

Financial Disclosures: The authors had no disclosures.
Optic Atrophy as a Presenting Sign of Suprasellar Germinoma in a Patient With Klinefelter's Syndrome

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Introduction:
Klinefelter’s syndrome is the most common chromosomal disorder associated with male hypogonadism and infertility, occurring in approximately 1 in 1000 male births.

Methods:
A 29 years old male diagnosed with Klinefelter’s syndrome was admitted to the hospital because of severe painless visual deterioration in his left eye. Upon admittance his visual acuity was 20/20 in his right eye and HM on his left. The fundus examination demonstrated a normal optic disc in the right eye and a pale disc on the left.

Results:
Brain CT and MRI images revealed a large sellar and suprasellar lesion involving the chiasm and the left optic nerve. Biopsy was taken from the lesion and the pathological exam confirmed the diagnosis as Germinoma. He was surgically treated followed by chemotherapy and radiation.

Conclusions:
Although it is known that patients with Klinefelter syndrome are predisposed to the development of extragonadal intracranial tumors, the appearance of germinomas in CNS is extremely rare. In the literature there are only few reported, associated with Klinefelter’s syndrome. The uniqueness of our case is that the presenting symptoms was of painless decreased vision for only two months, and upon examination there was left optic atrophy with bitemporal visual fields defects.

Keywords: Optic atrophy, Germinoma, Klinefelter syndrome

Financial Disclosures: The authors had no disclosures.
Poster 112

Bilateral Internuclear Ophthalmoplegia in Neuromyelitis Optica Spectrum Disorder

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Introduction:
Neuromyelitis Optica (NMO) and NMO spectrum disorders are increasingly recognized clinical entities. Outside of typical cases, the physician must use clinical clues to determine whether it is appropriate to evaluate for NMO. Recent studies show that some NMO brain lesions tend to be located in the hypothalamus and around the third and fourth ventricles. This distinctive localization provides an opportunity to assist in the diagnosis of NMO, as is seen in the case we present here.

Methods:
A 36 year old female with a history of cervical transverse myelitis presented with several days of horizontal binocular diplopia. Examination revealed bilateral internuclear ophthalmoplegia. Initial serum studies were unremarkable. CSF examination showed 13 leukocytes (97% lymphocytes), 660 erythrocytes, normal glucose and protein, and greater than 5 oligoclonal bands. MRI brain showed gadolinium enhancement and T2 signal hyperintensity in the hypothalamus and along the floor of the fourth ventricle.

Results:
She received methylprednisolone 1000mg IV daily for 5 days, followed by plasma exchange. Towards the end of her course of plasma exchange her internuclear ophthalmoplegia improved. Serum NMO antibody testing was positive.

Conclusions:
Bilateral INO has been described infrequently in NMO. Yet, the high density of aquaporin channel expression near the floor of the fourth ventricle, adjacent to the medial longitudinal fasciculus, provides a neuro-anatomic rationale for bilateral INO in NMO spectrum disorders. NMO, in addition to multiple sclerosis, should be considered in cases of bilateral INO.

References:


Keywords: Neuromyelitis Optica, Internuclear Ophthalmoplegia

Financial Disclosures: The authors had no disclosures.
Reversing Vision Loss and Preventing Optic Atrophy in Leber’s Hereditary Optic Neuropathy

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Introduction:
Leber’s hereditary optic neuropathy (LHON) causes profound loss of central vision and optic atrophy. A few reports exist of reversal of vision loss with idebenone or EPI-743 but almost invariably associated with optic atrophy. We report a young man presenting with central vision loss due to LHON. Vision and optic disc “pseudoedema” improved during treatment over a year without the development of optic atrophy.

Methods:
Case Report

Results:
A 17-year-old man presented in April 2011 with progressive painless vision loss over 6 months and visual acuities of 20/70-2 in both eyes. Color vision was normal, tested with HRR plates. Ophthalmoscopy and fluorescein angiography showed optic disc “pseudoedema”, engorged retinal vessels and abnormal ‘telangiectatic’ peripapillary arterioles. Cirrus OCT showed thickened peripapillary RNFL, averaging 174 microns OD and 202 microns OS. Automated visual fields (Humphrey) showed central scotomas with mean deviations of -5.9 dB OD and -8.9 dB OS. Genetic testing revealed 60-70% heteroplasmy for the 14484/ND6 mtDNA mutation. He was started on idebenone 900 mg daily, brimonidine drops 0.15% qid, N-acetylcysteine 600 mg tid, Vitamin B, C, E and liberal consumption of flax seed (phytoestrogens). By April 2012, visual acuities had improved to 20/25+2 OD and 20/20-1 OS, mean deviations were -2.3 dB OD and -3.4 dB OS and OCT RNFL measurements were 111 microns OD and 112 microns OS.

Conclusions:
Reversal of progressive vision loss and RNFL swelling in LHON without the development of optic atrophy has been reported only once previously. Treatment with a combination of idebenone, N-acetylcysteine, brimonidine eye drops, multivitamins and phytoestrogens may reverse ganglion cell dysfunction and prevent ganglion cell death in LHON.

References:

Keywords: Leber's hereditary optic neuropathy, Idebenone, brimonidine, Phytoestrogens, Vitamins

Financial Disclosures: The authors had no disclosures.
Introduction:
Idiopathic Intra-cranial Hypertension (IIH) is characterized by elevated intracranial pressure in the absence of a cranial mass or other disease process. Elevated intracranial pressures are hypothesized to result from an increased cerebrospinal fluid production, decreased reabsorption, or cerebral edema. Our patient illustrates a paradoxical change in intracranial pressure after diagnostic lumbar puncture for evaluation of IIH.

Methods:
Case Report: A 37-year-old caucasian, obese male with a history of hypertension and OSA was referred to ophthalmology for evaluation of blurred vision and headache. He was found to have optic disc swelling bilaterally. A diagnostic LP revealed an opening pressure of 440 mm H2O. The patient was discharged home but returned six hours later with complaints of pain nine out of 10 (VAS), throbbing postural headache. MRI revealed prominent cortical veins and diffuse posterior fossa, epidural and subdural enhancement throughout the spine. The patient’s headache did not improve despite intravenous fluid, caffeine, and NSAIDs over the next six days. An autologous epidural blood patch was performed without difficulty. The following day the patient noted significant improvement in his symptoms.

Conclusions:
We present a male patient that developed PDPH after LP for IIH. There have been many different contributors for development of PDPH reported in the literature including patient BMI, patient positioning, post-procedure ambulation, previous PDPH, and needle technique. IIH is most common in post-pubertal, pre-menopausal females and is rare in men with reported incidences of nine to 10%. Men often present with visual field impairment. There is also a strong gender component associated with risk of developing a PDPH, with women having two to three times the risk compared to men. We present a male patient that developed PDPH after LP for IIH. To our knowledge, this is the first case reporting a male with OSA and obesity being his only risk factors for IIH.

References:
3. Fraser, Bruce, Rucker, Risk Factors for Idiopathic Intracranial Hypertension in Men: A Case Control Study. Journal of the Neurological Sciences, 290(1), 86-89, 2010

Keywords: Idiopathic Intracranial Hypertension, Post dural Puncture Headache

Financial Disclosures: The authors had no disclosures.
Pelizaeus-Merzbacher Disease Presenting as Infantile Pendular Nystagmus: Potential for Masquerade as Spasmus Nutans

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Introduction:
Pelizaeus-Merzbacher Disease (PMD), a rare hereditary dysmyelinating disorder, may rarely masquerade as spasmus nutans (SN) in infancy presenting as isolated nystagmus.

Methods:
Case report with video recording

Results:
A 1 month-old boy presented with eye shaking and head bobbling for 2 weeks. Perinatal and family histories were unremarkable. He had a slow conjugate, sometimes elliptical, pendular nystagmus, head bobbling, shaking and turning, all extinguished during sleep. Pupils, fundus, vestibulo-ocular response, electroretinogram, CT and MRI of the brain were normal. SN was diagnosed. At 7 months, poor neck control and inability to sit unsupported, led to suspicion of PMD. A Proteolipid protein-1 (PLP-1) mutation was confirmed.

Conclusions:
In isolated infantile pendular nystagmus, after excluding retinal dystrophy with an ERG and fundus exam to rule out optic atrophy the patient undergoes scanning to exclude an intrinsic or extrinsic chiasmal mass or other intracranial abnormality, than needs continual follow-up for the development of further neurological deficits or amblyopia. Our patients nystagmus frequency was slower than classic SN. If the nystagmus goes away and no other deficits develop SN is diagnosed. PMD should be suspected when hypotonia, nystagmus and head bobbing comingle. An X-linked recessive PLP-1 mutation, (normally encoding a myelin sheath component), causes PMD, which has a wide spectrum of clinical severity. The Connatal form presents within the first 2 weeks of life with stridor, seizures, and hypotonia. Cognitive impairment and early mortality follow. Classic PMD presents within the first year of life with nystagmus and head titubation. Dysarthria, spastic quadriplegia, and rarely, seizures develop; lifespan may be shortened to mid-adulthood. MRI reveals hypomyelination in the hemispheres, cerebellum, and/or brainstem, with cortical atrophy. The corticospinal tracts may be spared in classical PMD. However, radiologic features may not be apparent until age 2.

References:


Keywords: Nystagmus, Dysmyelination, Genetic

Financial Disclosures: The authors had no disclosures.
Extreme Intracranial Hypertension Associated with Abnormal Ocular Motility and Ptosis

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Introduction:
Few cases of unusual eye movements associated with intracranial hypertension are reported.1-5 Infrequently, cranial nerve palsies, including third nerve palsies are reported as well.6-9 To our knowledge, ptosis with gaze palsy associated in the setting of increased intracranial pressure has not been previously reported.

Methods:
Two cases of intracranial hypertension with unusual eye movements and associated ptosis are reported.

Results:
Two women presented with profound bilateral vision loss and headache. Examination, documented with videography, showed bilateral gaze palsies, ptosis, optic nerve fiber layer edema, and elevated intracranial pressure (ICP) > 55 cm H2O. One woman had a venous sinus thrombosis; the other did not have radiologic evidence of intracranial pathology accounting for increased ICP. Optic nerve sheath fenestration with cerebral spinal fluid diversion was recommended to both patients. One underwent both and recovered vision to better than 20/400; the other refused neurosurgical intervention and remained NLP. Eye movements and ptosis improved over time in both patients.

Conclusions:
Eye movement abnormalities, coupled with ptosis, are ominous signs of diffuse neurologic compromise from raised intracranial pressure. Emergent pressure reduction of the optic nerve and cerebral subarachnoid spaces to salvage neurologic function should be considered.

References:

Keywords: Intracranial hypertension, Ophthalmoplegia, Ptosis

Financial Disclosures: The authors had no disclosures.
Poster 117

“Localize This” Lesion

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Introduction:
Poster to discuss workup steps not to miss in diagnosing atypical causes of a Lambert Eaton Myasthenic Syndrome.

Methods:
Case presentation of an African American female with a Myasthenia-like presentation from a neuroendocrine tumor that led to a positive work up for LEMS after an exhaustive workup for myasthenia gravis.

Results:
Discussion of treatment strategy differences for this cause of LEMS.

Conclusions:
When a patient presents with classic myasthenia symptoms that do not fit a temporal pattern, maintain a high suspicion for LEMS and consider imaging the abdomen in addition to the chest.

Keywords: Lambert Eaton Myasthenic Syndrome, Neuroendocrine tumor, Atypical Myasthenia

Financial Disclosures: The authors had no disclosures.
Traumatic Avulsion of the Optic Nerve

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Introduction:
Traumatic avulsion of the optic nerve is an unusual form of traumatic optic neuropathy characterized by disruption of the axons at the level of the optic nerve due to penetrating or non-penetrating trauma. These patients generally have a poor prognosis and are often seen late with scarring. Recognition at the time of injury is often difficult because of associated damage to the anterior segment or blood within the vitreous.

Methods:
Three patients with traumatic avulsion of the optic nerve are presented documenting the poor visual result and the classical findings seen on funduscopy and B-scan.

Results:
Traumatic avulsion of the optic nerve is often recognized late with scarring over the discs but sometimes acutely with variable disruption of the vascular supply over the optic nerve head leading to local hemorrhage around the disc but also into the vitreous. Visual results remain poor and no therapeutic intervention has been successful. As the blood supply to the anterior segment is not usually disrupted in spite of traumatic reduction in afferent system function these patients usually maintain their globe unless there is additional associated injury.

Conclusions:
Traumatic optic nerve avulsions are a rare syndrome associated with hemorrhage at the disc and into the vitreous leading to late scarring over the disc.

Keywords: Trauma, Optic neuropathy, Avulsion, Vitreous hemorrhage

Financial Disclosures: The authors had no disclosures.
Alport's Syndrome: A Diagnosis Made Easier to Swallow by the Retinal Lozenge

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Introduction:
Alport Syndrome is a genetically and clinically heterogeneous disorder characterized by progressive renal failure, sensorineural hearing loss, and ocular abnormalities. These clinical findings have been recently attributed to a disorder of basement membrane type IV collagen (ref 1). Many affected individuals are unrecognized, because the clinical features are atypical, renal biopsy is unhelpful, and genetic testing not readily available (ref 2). The ocular manifestations can be diagnostic.

Methods:
Case Report

Results:
A forty-seven year old male was referred for loss of vision in his right eye for six months. History was notable for bilateral sensorineural hearing loss since age 12, end-stage renal failure requiring kidney transplantation at age 19, when diagnosis of Alport Syndrome was suspected. Slit-lamp showed anterior lenticonus OU. Fundus demonstrated a dull macular reflex with a temporal confluent area of retinal flecks, i.e., a retinal lozenge OU. OCT showed thinning of the temporal macula in the region of the lozenge.

Conclusions:
Alport Syndrome is a progressive hereditary disease characterized by renal failure, sensorineural hearing loss, anterior lenticonus, corneal posterior polymorphous dystrophy, macular lozenge, peripheral retinal flecks, and macular holes. Red-free photography increases the sensitivity of the retinal exam for diagnosis (ref 2). In forty percent of cases, ocular manifestations may be the earliest sign of Alport’s prior to the onset of the renal disease. Drusen and retinal flecks have been seen in other glomerulonephropathies, especially, membranoproliferative glomerulonephritis type II, however, the retinal lozenge has been only reported in Alport’s (ref 3). The classic findings of anterior lenticonus and retinal “lozenge” as seen in our patient are specific and diagnostic for the syndrome. (ref 4) and associated with severe retinopathy, early onset renal failure, and is found in both X-linked and autosomal recessive disease. (ref 2).

References:

Financial Disclosures: The authors had no disclosures.
Introduction:
Silicone breast prosthesis has been associated with a variety of medical conditions or autoimmune diseases, which has coincidental relation with the implants insertion; it’s loomed as a new and unknown pathology of the modern times. More than 87% of symptomatic patients developed demyelination axonal neuropathy demonstrated by nerve and muscle biopsy; 22% to 25% have evidence of autoimmune thyroid disease. An a small group of patients (10%-12%) have primary central nervous system demyelination disease as, multiple sclerosis. The diagnosis of multiple sclerosis was corroborated by magnetic resonance imaging and cerebrospinal fluid analysis. Also, an other wide spectrum of immunological diseases have been observed, such as fibromyalgia, Hashimoto’s thyroiditis, polymyositis, dermatomyositis, lupus erythematosus, rheumatoid arthritis, scleroderma, and the presence of autoantibodies. Finally, for symptomatic patients, an adjuvant syndrome of silicone breast prosthesis or implant is proposed as a unitary diagnosis. The authors presented four patients whom illustrated this entity.

Methods:
Neuroophthalmic examination was doing in four patients with contracture and extracapsular rupture of silicon breast implants. Subjects underwent a complementary test as brain and breast magnetic resonance imaging (MRI), immunological test, tumor markers, silicon level and biopsy.

Results:
Optic neuropathy and scleritis were the visual manifestations. Three patients developed autoimmune diseases “like” multiple sclerosis, rheumatology syndrome, thyroiditis and one patient developed cancer.

Conclusions:
The human adjuvant disease isa controversial autoimmune disease as well as with non-specific autoimmune manifestations. Silicon breast implant can cause serious local and systemic complications, including death. Patients with neuroophthalmic manifestations and implant silicone breast should be study to discard human adjuvant diseases

References:

Keywords: Siliconosis, Human Adjuvant disease, Breast silicon implant, Augmentation mammoplasty, Autoimmune disease

Financial Disclosures: The authors had no disclosures.
Should We Consider Pilocarpine as Part of Therapy in Patients With Third Nerve Aberrant Regeneration?

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Introduction:
To determine the utility of topical diluted pilocarpine as adjuvant therapy for patients with internal ophthalmoplegia secondary to third nerve aberrant regeneration.

Methods:
Topical 0.125% pilocarpine was installed in 13 patients with pseudo Argyll-Robertson pupil (internal ophthalmoplegia) secondary to third nerve regeneration. The parameters studied were visual acuity, refraction, pupillary reaction and size, prismatic measurement, visual field, photophobia and astenopia symptoms.

Results:
Thirteen patients with third nerve aberrant regeneration were studied (ten unilateral, two bilateral). As a result of the pharmacological meiosis: the visual acuity and refraction improved for near vision in 1.5 to 2 diopters; the prismatic values decreased, as well the photophobia and astenopia symptoms.

Conclusions:
Topical pilocarpine by pupillary cholinergic supersensitivity cause meiosis in patients with pseudo Argyll-Robertson pupil with consequent improvement the visual acuity, refraction and diminution of the photophobia and astenopia symptoms. Treatment with dilute pilocarpine should be considered in patients with third nerve aberrant regeneration.

References:

Keywords: Third nerve aberrant regeneration, Supersensitivity denervation, Diluted pilocarpine, Internal ophthalmoplegia, pseudo Argyll-Robertson pupil

Financial Disclosures: The authors had no disclosures.
Poster 122

Posterior Cortical Atrophy – Case Report and Retrospective Review

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Introduction:
Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative disease with prominent cortical visual dysfunction, first described by Benson in 1988. Studies have shown visual field defects on formal testing, increased phosphorylated-tau protein in the CSF, and hypometabolism in the parieto-occipital regions on PET imaging. We describe one patient with PCA whose symptoms are classic but whose progression has been atypically slow. To further investigate this disease entity, we retrospectively reviewed 8 additional charts and report the commonalities in all 9 cases.

Methods:
We reviewed the index subject’s symptoms and findings on presentation and follow up. We reviewed 8 additional cases of PCA evaluated by Neuro-Ophthalmology. We analyzed the signs and symptoms in, as well as the demographics of, these cases to look for common themes at presentation.

Results:
A 58 year old woman presents with a progressive 10 year history of difficulty processing spatial information, reading handwriting, and recognizing faces. She described visual hallucinations, photophobia, poor depth perception, and visual recall. Visual field testing found an incomplete hemianopsia. CSF analysis detected reduced Aβ (1-42) Tau Index and elevated phosphorylated-tau. Poor visuospatial skills were found on neuropsychological testing and occipito-parietal hypometabolism on PET imaging. Furthermore, 7/9 patients reported trouble reading and 4/8 depth perception difficulty, which were the most recurrent complaints. Recurrent signs included abnormal color plates (9/9), simultanagnosia (6/8), hemianopsia (3/7), decreased stereopsis (4/4), and abnormal Amsler’s grid (4/5).

Conclusions:
Posterior cortical atrophy is an uncommon atypical variant of Alzheimer’s disease, sometimes termed “Benson’s syndrome”. Patients commonly report difficulty reading despite normal visual acuities. Our observations and previous studies describe visual signs and symptoms attributable to the parieto-occipital cortex. More recently, CSF phosphorylated-tau has been utilized as a bio-marker. A systematic approach using clinical, laboratory, and radiographic data can aid in diagnosis of PCA. This may help clinicians with similar diagnostic dilemmas.

References:

Keywords: Posterior cortical atrophy, Neurodegenerative, Cortical visual dysfunction, Diagnosis, Bio-marker

Financial Disclosures: The authors had no disclosures.
Breast Cancer Metastases to Bilateral Optic Nerve Sheaths in Leptomeningeal Carcinomatosis

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Introduction:
Tumor metastasis to the optic nerve sheath is rare and is associated with significant morbidity and a poor prognosis. Bilateral optic nerve sheath involvement is exceedingly uncommon, but one possible route for this devastating complication is through the cerebrospinal fluid in cases of leptomeningeal carcinomatosis.

Methods:
We report a case of metastatic breast cancer that presented with bilateral vision loss secondary to metastases to the optic nerve sheaths in the setting of leptomeningeal carcinomatosis.

Results:
A 50-year-old female with a history of metastatic breast cancer (in remission) presented with rapid-onset, painless, sequential vision loss in both eyes. Visual acuity was 20/50 and hand motion in the right and left eyes, respectively, and a relative afferent pupillary defect was noted in the left eye. Magnetic resonance imaging demonstrated hyperintensity of the intra-orbital optic nerve sheaths. Cerebrospinal fluid cytology demonstrated breast tumor cells. Whole brain radiation resulted in a subjective improvement in visual acuity.

Conclusions:
The ophthalmologist can play a key role in the diagnosis of leptomeningeal carcinomatosis when the optic nerve sheaths are involved. However, bilateral involvement is very uncommon, so a high level of clinical suspicion and coordination with multiple medical disciplines are needed for its diagnosis.

References:

Keywords: Optic Nerve Sheath, Leptomeningeal Carcinomatosis, Metastatic Breast Cancer

Financial Disclosures: The authors had no disclosures.
Complete Bilateral Horizontal Gaze Paralysis as Only Presenting Symptom of Stroke: A Case Report

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Introduction:
Complete bilateral horizontal gaze paralysis has been reported from causes including hemorrhage, masses, multiple sclerosis, and stroke. However, a complete horizontal gaze palsy in the absence of other neurological symptoms from a single, isolated pontine infarction has not previously been reported. We report a case of complete bilateral horizontal gaze palsy as the only presenting sign of inferior pontine infarction.

Methods:
Case report.

Results:
A 77 year old woman with longstanding hypertension and hyperlipidemia presented to the hospital with bilateral gaze disturbances. Her neurological exam revealed a woman who was alert and oriented. She had complete paralysis of horizontal pursuit bilaterally. Doll’s head maneuver was ineffective in driving horizontal eye movement. She had only trace limitation of vertical gaze bilaterally. Facial sensation and movement were intact, and she was otherwise neurologically intact. Diffusion-weighted MRI one day after the onset of symptoms demonstrated restricted diffusion involving the dorsal aspect of the inferior pons, in the region of the nucleus of the sixth cranial nerve. T2/FLAIR images revealed a high intensity signal in the same area.

Conclusions:
Horizontal gaze relies upon the function of motor neurons of the abducens nerve as well as internuclear neurons of the contralateral MLF and oculomotor nucleus. This patient likely had bilateral damage to PPRF and abducens nucleus which are anatomically commingled. Previous reports of pontine infarction demonstrate a range of clinical severity with neurological deficits including hemiparesis and ataxia. While this patient is fortunate to have limited neurological deficits, patients with brainstem infarction of any kind have a stroke recurrence and mortality rate of 30% within 3 years.

References:

Keywords: Ophthalmoplegia, Horizontal Gaze Palsy, Pontine Infarction

Financial Disclosures: The authors had no disclosures.
Syphilitic Optic Neuropathy Mimicking Amarusis Fugax and Ischaemic Optic Neuropathy

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Introduction:
Syphilitic optic neuritis typically presents with acute visual loss associated with swelling of the optic disc. However, the myriad presentations of this condition is recognized and the term ‘great imitator’ should always be remembered. We present 2 cases of syphilitic optic neuritis, one mimicking giant cell arteritis, and the other ischaemic optic neuropathy. These cases may highlight the vasculitic nature of this condition.

Methods:
Retrospective case series

Results:
Two patients with neurosyphilis are presented. Case 1: A 69 year-old Chinese male who complained of transient visual obscurations for a month. On initial presentation, ophthalmological examination was normal. A month later, he returned with the same complaint but this time right optic disc swelling was noted although visual function was normal. He was investigated for giant cell arteritis but temporal artery biopsy was negative. Case 2: A 53 year old Chinese female complained of decreased vision in her right eye for a week. Visual field examination revealed inferior field loss in the right eye, the left was normal. She had bilateral disc swelling. In both cases the final diagnosis was neurosyphilis.

Conclusions:
Syphilis should always be considered in the differential diagnosis of a patient with optic neuritis. We believe that this is the first reported case of syphilitic optic neuritis presenting with transient obscuration of vision mimicking giant cell arteritis.

References:

Keywords: Syphilis, Amarusis fugax, Giant cell arteritis, Optic neuritis, Papillitis

Financial Disclosures: The author had no disclosures.
Diagnostic Criteria for the Pseudotumor Cerebri Syndrome

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Introduction:
Problems with the term IIH, over- and misuse of the diagnosis "IIH without papilledema," greater awareness of the associated radiologic abnormalities, and a better understanding of this condition in children and what constitutes a normal CSF opening pressure in this age group, necessitate a revision to the nomenclature [pseudotumor cerebri syndrome (PTCS)] and diagnostic criteria for all age groups.

Methods:
We revised the 2002 classification and diagnostic criteria.

Results:
Criteria: 1. Required for diagnosis of the PTCS: A. Papilledema B. Normal neurological examination except for cranial nerve abnormalities C. Neuro-imaging: Normal brain parenchyma without evidence of hydrocephalus, mass, or structural lesion and no abnormal meningeal enhancement on magnetic resonance imaging (MRI), with and without gadolinium, for obese females, and MRI, with and without gadolinium, and MR venography for others. If MRI is unavailable or contraindicated, contrast-enhanced computed tomography may be used. D. Normal CSF composition 2. Diagnosis of PTCS without papilledema: In the absence of papilledema, a diagnosis of PTCS can be made if B-D from above are satisfied, and in addition A and B below are satisfied. In the absence of papilledema or sixth nerve palsy, the diagnosis is "suggested" but not established if B-D from above are satisfied, in addition to A and C below. A. Elevated LP opening pressure (> 250 mm CSF in adults and > 280 mm CSF in children (250 mm CSF if the child is not sedated and not obese) in a properly performed lumbar puncture B. Unilateral or bilateral abducens nerve palsy C. Neuroimaging signs (at least three): (i) Empty sella; (ii) Flattening of the posterior sclera; (iii) Distention of the perioptic subarachnoid space +/- a tortuous optic nerve; (iv) Protrusion of the optic nerve papillae into the vitreous cavity; (v) Transverse venous sinus stenosis

Conclusions:
Further study will determine the specificity and sensitivity of these criteria.

References:

Keywords: pseudotumor cerebri syndrome, idiopathic intracranial hypertension, diagnostic criteria

Financial Disclosures: Drs. Friedman and Digre have received research support from the National Eye Institute (Idiopathic Intracranial Hypertension Treatment Trial). Dr. Liu serves as a consultant to Ipsen.
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Xq26.3 Microdeletion in a Male with Wildervanck Syndrome

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Introduction:
Wildervanck syndrome (cervico-oculo-acoustic syndrome) consists of the Klippel-Feil anomaly, Duane retraction syndrome, and congenital deafness. It is much more common in females than males and could be due to an X-linked mutation that is lethal to hemizygous males. We present the genetic evaluation of a male with Wildervanck syndrome and his family.

Methods:
Clinical evaluation, neuroimaging, sequencing of candidate genes, and array comparative genomic hybridization.

Results:
The patient had bilateral type 1 Duane retraction syndrome, fusion of almost the entire cervical spine, and bilateral severe sensorineural hearing loss due to bilateral cochlear dysplasia; he also had congenital heart disease requiring surgery. His parents were unrelated, and he had eight unaffected siblings. The patient had no mutation found by Sanger sequencing of HOXA1, KIF21A, SALL4, and CHN1. He had a 3kB deletion in the X-chromosome at Xq26.3 that was not found in his mother, one unaffected sibling, or 56 healthy controls of matching ethnicity. This deletion encompassed only one gene, Fibroblast Growth Factor Homologous Factor 2 (FGF13), which encodes a 216-amino acid protein that acts intracellularly in neurons throughout brain development.

Conclusions:
Analysis of this patient’s phenotype and genotype opens the possibility that X-chromosome deletions may be a cause of Wildervanck syndrome with larger deletions being lethal to males and that FGF13 mutations may be a cause of Wildervanck syndrome.

Keywords: Wildervanck syndrome, Duane retraction syndrome, Medical genetics, Congenital eye movement abnormality, Congenital deafness

Financial Disclosures: The authors had no disclosures.
**Severe Bilateral Retrobulbar Optic Neuritis In The Elderly with Urinary Tract Infection**

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**Introduction:**
Bilateral optic neuritis in the elderly is rarely reported due to urinary tract infection (UTI) when multiple systemic disorders are often present. Parainfectious optic neuritis or NMO like syndrome has been more often seen in children or due to viral, mycoplasma or mycobacterium infection. We present two elderly cases of isolated severe bilateral retrobulbar optic neuritis recovered after IV Steroid whereas bacteria UTI is the sole finding in the history or workup.

**Methods:**
Summary of case reports. A 91 year-old White woman with myelodysplasia and pancytopenia treated periodically with transfusion, Procrit and Neupogen, developed rapid progressive painless visual loss over one week. At visit, she had NLP for 5 days OD, and 2 days OS. The second case is a 61 year-old Filipino man, s/p colon and partial bladder resection for colon cancer one month prior, who developed progressive bilateral visual loss with minimal pain to CF in OD, LP in OS about one week after treatment for urosepsis. No Flu like symptoms in either case. Both cases showed mild optic disc edema without hemorrhage OU. Orbit MRI revealed mildly enhanced optic nerves in the orbital segment; brain MRI, mild microvascular deep white matter change. Temporal artery biopsy, CSF study, NMO IgG, etc. were all negative. Urinalysis and culture revealed 100 K colonies of G (-) rod and enterococcus in the first case.

**Results:**
Both patients were treated with IV Methylprednisolone 500-1000 mg /day for 3 days followed by oral prednisone 40-60 mg/d with rapid tapering off in week in the first case besides antibiotics for UTI, and prolonged oral prednisone for 4 weeks in the second case. Both had visual recovery in 2 months to 20/30- 20/25, and no recurrence for 5-12 months respectively.

**Conclusions:**
Bacterial UTI in the elderly can result in bilateral severe inflammatory demyelinating retrobulbar optic neuritis while the underlying compromised immune status may be a risk factor. Urinalysis and culture are warranted in the work up of cause of optic neuropathy in this population, and to try IV steroid while treating UTI.

**References:**

**Keywords:** Bilateral Retrobulbar Optic Neuritis, Bacteria Urinary Tract Infection, Elderly

**Financial Disclosures:** The authors had no disclosures.
Poster 129

Chiari I Malformation and Idiopathic Intracranial Hypertension

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Introduction:
Chiari I malformation and low-lying cerebellar tonsils are more common in patients with idiopathic intracranial hypertension (IIH) than in the general population. It is not clear what factors predispose people with IIH to have low lying tonsils. One possibility is that they have small posterior fossas.

Methods:
I reviewed the charts of four of my patients with both IIH and Chiari I. I compared the size of their posterior fossas and the opening pressures on spinal taps to those of four patients with IIH whose tonsils were above the foramen magnum. The posterior fossas were measured using the method of Milorat (Neurosurgery 44(5): 1005-1017, May, 1999).

Results:
There was not difference in size of the posterior fossa or intracranial pressure between the two groups.

Conclusions:
Small posterior fossa size does not predispose patients with IIH to develop low-lying cerebellar tonsils or Chiari I syndrome.

Keywords: Chiari I malformation, Idopathic intracranial hypertension

Financial Disclosures: The authors had no disclosures.
Multicenter Inter-rater Reliability of Retinal Layer Segmentation Using Spectral-domain OCT

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Introduction:
Optical coherence tomography (OCT) measures of different retinal layers correlate with structural and functional markers of disease severity in multiple sclerosis (MS), making intra-retinal layer segmentation of high-resolution OCT scans an increasingly important tool for the assessment of clinical progression in MS patients. While automatic intra-retinal layer segmentation is not yet widely available, semi-automatic segmentation of scans is necessary. The aim of our study was to evaluate reliability of semi-automatic and assisted intra-retinal layer segmentation in a multicentre setting and to identify sources of disagreement.

Methods:
Macular volume scans (25 vertical B-scans á 1024 A-scans, scan angle = 20°x20°, ART=49) were generated with a spectral-domain OCT device (Spectralis, Heidelberg Engineering) from 3 MS patients and 3 healthy controls. After agreeing on a common procedure in preceding video sessions, intra-retinal segmentation was performed using a semi-automatic algorithm with manual correction. Standard deviation (SD) between graders was visualized in spatial maps and interclass correlation coefficients (ICC) were used to analyse the inter-rater disagreement for all retinal layers.

Results:
SD of segmentation lines between graders were reaching a maximum of 6 pixel (=23.4μm). However, high SD values were strongly correlated with the locations of retinal blood vessels. The ganglion cell layer yielded the lowest ICC, still mean ICCs were very high for all segmentation lines (ICC > 0.9 for all layers). Spatial distribution of ICCs was different for the retinal layers: the retinal nerve fiber layer showed more disagreement in superior and inferior macular areas while ICCs were lower for the central macular region for the ganglion cell layer.

Conclusions:
Retinal layer segmentation is reliable and can be applied in multicentre settings. Training graders before assisted segmentation guarantees high reliability among centres. However, blood vessels running mainly through the retinal nerve fiber and ganglion cell layers are an important source of disagreement.

References:

Keywords: Multiple sclerosis, Optical coherence tomography, Retinal segmentation, Inter-rater reliability

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Orbital Cerebrospinal Fluid Leak Following Pterional Orbitozygomatic Craniotomy

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Introduction:
In this report we describe orbital cerebrospinal fluid (CSF) collections as a complication of orbitozygomatic pterional craniotomy. Pathophysiology and management are assessed.

Methods:
In this retrospective interventional case series the medical records were reviewed of two patients (18 year-old male, 57 year old female) with orbital CSF leaks following orbitozygomatic pterional craniotomy. Main measures were mechanism and management.

Results:
Case 1: An 18 year-old woman underwent exploratory orbitozygomatic pterional craniotomy. On post-operative day five, following removal of a lumbar drain, proptosis and a compressive optic neuropathy developed. Computed tomography (CT) demonstrated a CSF collection contiguous with the craniotomy site. Resolution followed percutaneous aspiration and replacement of the lumbar drain.

Case 2. A 57 year-old woman underwent an orbitozygomatic craniotomy for removal of a left anterior clinoid meningioma, complicated by a large left hemorrhagic stroke requiring decompressive hemicraniectomy. Extracranial CSF collections accumulated in both the orbit and subgaleal spaces. Resolution followed placement of an external ventricular drain.

Conclusions:
We describe orbital CSF collections as a complication of pterional-orbitozygomatic craniotomy. Based these cases the mechanism appears to be the combination of an iatrogenic communication with the sub-arachnoid space and elevated ICP. Successful management is achieved simply by normalizing ICP. In vision threatening cases, more rapid resolution is facilitated by conjunctival percutaneous drainage.

Keywords: Orbit, Cerebrospinal fluid, Craniotomy, Surgical complication, Neurosurgery

Financial Disclosures: The authors had no disclosures.
Vertical Diplopia from Skew Deviation in a Patient with Thalamic Infarct

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Introduction:
Skew deviation has been reported in thalamic infarcts but is not as common as vertical gaze paresis. We describe a patient with chronic thalamic and pontine infarcts who presented with a transient new onset vertical diplopia due to skew deviation.

Methods:
Single case report and review of literature

Results:
A 55 year old female with uncontrolled diabetes mellitus, hypertension, hyperlipidemia and a past history of stroke presented with a 12 hour history of sudden onset vertical diplopia. Initial neurologic examination showed left hypertropia and chronic left spastic hemiparesis. Neuro-ophthalmic exam showed skew deviation. She had 2 prism diopter left hypertropia in primary gaze, excyclotorsion of right eye and normal ocular movements. Diplopia and vertical deviation resolved within 24 hours. MRI head showed an acute infarct in the right frontal periventricular white matter and a chronic right paramedian thalamic infarct. Ischemic gliotic changes were seen in the right caudal pons. She was also noted to have atherosclerotic lesions of both internal carotid arteries which suggested an atheroembolic process.

Conclusions:
We believe our patient developed a skew deviation due to transient ischemia of the right paramedian thalamus adjacent to the prior thalamic infarct from a new thromboembolic process. Thalamic lesions rarely present with vertical skew deviation. In patients with thalamic stroke, the skew is believed to be secondary to an extension of peri-infarct edema into the territory of the medial longitudinal fasciculus and interstitial nucleus of Cajal in the brainstem. An acute onset of skew deviation prompted neuro-imaging, which revealed additional right frontal infarcts from an atheroembolic process that resulted in aggressive risk factor management.

References:

Keywords: Skew deviation, Thalamic infarct, Vertical diplopia

Financial Disclosures: The authors had no disclosures.
A Case of Kaleidoscopic Vision

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Introduction:
Visual allesthesa is a poorly understood, pathological distortion of visual perception. The classic description is that of tilting or rotation of the visual field. There are case reports of more complex phenomenon where the retinotopic visual field is rotated, flipped, or inverted independent of head position. We report a case of a patient who described typical symptoms of visual allesthesa followed by a more complex visual phenomenon in the context of a posterior circulation stroke.

Methods:
An otherwise healthy 51 year-old right-handed female presented with report of transient visual changes as the first symptoms of a stroke two months earlier. At the time of her evaluation she was asymptomatic, her neuro-ophthalmologic and general neurologic examination were normal. At the time of her stroke, she described a tilting of her visual field by “seventeen degrees”. This was followed by a brief episode of “kaleidoscope” vision where she reported a fractionation of her visual field with segments misplaced or jumbled, like ‘pieces of a misarranged puzzle.’ These initial visual symptoms were then followed by more persistent motor pulsion, nausea and vomiting. Subsequent MRI/MRA showed a small residual stroke at the right superior vermis, with no evidence for atherosclerotic disease or vessel occlusion.

Results:
The MRI confirmed a stroke in the posterior circulation. At the time of imaging, the patient was asymptomatic so more precise localization of her visual symptoms could not be established.

Conclusions:
The patient’s symptoms were likely the result of an emboli to the posterior circulation that then autolysed leaving the small residual asymptomatic infarction, seen on MRI. Her initial visual symptoms were consistent with typical descriptions of visual allesthesa. We propose that the second phenomenon was a more complex form of visual allesthesa. To our knowledge, similar cases of disordered segmentation of vision have not previously been described in brainstem/cerebellar events.

References:
3. Audiovisual recording to be included with poster

Keywords: Kaleidoscopic Vision, Visual Allessthesa, Posterior Circulation Stroke

Financial Disclosures: The authors had no disclosures.
Poster 134

Bilateral Blindness from a Metastatic Abdominal Neuroblastoma

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Introduction:
Very few cases of adult metastatic neuroblastoma with vision loss without accompanying optic nerve compression or orbital metastases have been described. We present a case of a metastatic neuroblastoma presenting with headache and bilateral visual loss from increased intracranial pressure. Clinical findings included skull base involvement without optic nerve compression.

Methods:
Retrospective case report.

Results:
29-year-old woman presented with headache and significant visual loss for one month. Work up revealed pelvic neuroblastoma with intracranial metastasis. She did not have opsoclonus-myoclonus; serum homovanillic acid and vanilmandelic acid were elevated. Dilated fundus exam showed significant bilateral optic disc edema with confluent, diffuse hemorrhages, multiple cotton wool spots and retinal striae. Magnetic resonance imaging (MRI) showed dural and calvarial involvement and abnormal flow within both transverse sinuses. Opening pressure was greater than 55 cm H2O. Cerebral spinal fluid (CSF) did not show carcinomatous meningitis nor infection. Visual acuity improved after treatment with acetazolamide and CSF diversion, suggesting increased pressure as the source of her initial visual loss. Her vision then again decreased to HM and NLP. Subsequent MRIs showed narrowing of the superior sagittal and transverse sinuses, extensive sinus disease, and ultimately Posterior Reversible Encephalopathy. The patient underwent sinus debridement, optic nerve sheath fenestration, and revision of CSF diversion without improvement of visual symptoms. Her initial CSF protein was normal, but rose to 354. She was treated with radiation and chemotherapy.

Conclusions:
Neuroblastoma is an extremely rare tumor in adults, with only 10% of cases occurring in patients older than 10 years. Visual loss at presentation is uncommon, and is usually due to compression or direct involvement of the optic nerves, or orbital metastases. In this patient, the unusual presentation of metastatic neuroblastoma with visual loss was found to be related to increased intracranial pressure rather than compression, overt optic nerve infiltration, or orbital metastases.

References:


Keywords: Metastatic Neuroblastoma, Vision loss, Papilledema, No optic nerve compression, Adult

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Cavernous Sinus Syndrome: Where Do You Go From Here?

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Introduction:
It is well recognized that squamous cell carcinoma of the skin can go on to perineural invasion of the surrounding tissue. However, this diagnosis can be quite challenging in the face as the carcinoma can spread along branches of the trigeminal nerve to invade the cavernous sinus.

Methods:
A series of two patients was reviewed.

Results:
The first case is a 70 year old man with history of cutaneous basal and squamous cell carcinomas who presented with left eye pain and forehead numbness. He went on to develop vertical diplopia with a normal MRI and was diagnosed with a fourth nerve palsy, however his pain continued. A supraorbital biopsy was performed that showed no malignant cells, however when he developed a 3rd and 6th nerve palsy, repeat MRI showed thickening of the trigeminal nerve. Neurosurgical consultation for cavernous sinus exploration was entertained and eventually showed squamous cell carcinoma. The second case is a 59 yo man with pain in the right eye for one year. He, too, had a history of a basal cell carcinoma excised from the opposite side of the forehead 4 years prior. An initial MRI was normal, however he went on to develop numbness in V1, and later cavernous sinus syndrome with ophthalmoplegia. A repeat MRI showed an enlarged cavernous sinus with extension along the fifth nerve. A supraorbital nerve biopsy was performed which showed squamous differentiated carcinoma cells surrounding the nerve.

Conclusions:
These clinical scenarios are consistent with perineural spread from cutaneous carcinoma. Often, radiologic imaging is helpful in the diagnosis however may often be negative especially early in the clinical course. Numbness and involvement of additional cranial nerves led to repeated neuroimaging in both patients. In some cases, biopsy of the supratrochlear nerve and/or exploratoration of the cavernous sinus may be the only way to diagnose this.

References:


Keywords: Trigeminal nerve, Cavernous sinus syndrome, Squamous cell carcinoma

Financial Disclosures: The authors had no disclosures.
Poster 136

Diffusion-Weighted Imaging Findings in a Case of Malignant Optic Glioma

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Introduction:
Anterior visual pathway malignant gliomas are rare, often simulate optic neuritis clinically and radiographically, and progress to blindness and death within a year. We report a patient with a typical presentation, in whom biopsy was deferred because the diagnosis was thought to be Tolosa-Hunt syndrome (THS). The DWI findings in this case have not previously been reported, to the best of our knowledge, and highlight the cytotoxic edema/infarction often seen in this condition.

Methods:
Case report.

Results:
A 54-year-old man lost vision OS over 6 weeks, then developed pain, eyelid swelling, and limitation of eye movement. Visual acuity was 20/20 OD and NLP OS. Funduscopic examination revealed florid disc edema with CRVO and CRAO OS. Orbit CT findings were consistent with inflammation. Blood tests were unremarkable. Oral corticosteroids resulted in pain relief. MRI showed non-enhancing enlargement of the left optic nerve and chiasm, with T2 and DWI hyperintensity, and hypointensity on ADC mapping. Within days, he developed a temporal hemianopsia OD. LP was unrevealing. A left intraorbital optic nerve biopsy showed necrosis without evidence of neoplasm. Repeat MRI showed significant reduction in previous enlargement and T2 signal, with new enhancement. Neurosurgery diagnosed THS, and biopsy was deferred because of resolved pain and radiographic improvement. One month later, he was NLP OU, with altered mental status. MRI showed an infiltrating optic chiasm mass extending into both optic tracts; the pathologic diagnosis was glioblastoma, WHO grade IV.

Conclusions:
Proximal optic nerve involvement with malignant optic nerve glioma is associated with distinctive funduscopic findings of progressive optic disc edema, venous stasis/occlusion, followed by arterial occlusion, and infarction. The DWI findings in this patient were consistent with and due to post-infarction changes. The correct interpretation of this radiographic finding, in addition to the lack of optic chiasmal involvement in THS, may have led to earlier diagnostic biopsy.

References:
1. Hoyt WF, Meshel LG, Lessell S, Schatz NJ, Suckling RD.

Keywords: Malignant optic glioma, Diffusion-weighted imaging, ADC mapping

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Poster 137

More Than Skin Deep

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Introduction:
We present a case of an elderly patient with multiple medical problems with a painful right third nerve palsy and progressive vision loss. Diagnosis of perineural spread of squamous cell carcinoma was challenging due to his concomitant medical issues limiting diagnostic testing. In many cases diagnosis is further delayed as patients are unaware of prior dermatologic history.

Methods:
An 86-year-old man with end-stage congestive heart failure with ICD, atrial fibrillation, chronic obstructive pulmonary disease, hypertension, cerebral infarction, renal failure and forehead skin cancer of unclear etiology was admitted for a painful ophthalmoplegia and right eye vision loss. Four months prior, he was worked up at an outside hospital for a painful third nerve palsy where imaging was reported to be unremarkable. He was given the diagnosis of Tolosa Hunt syndrome. He was not treated and eventually progressed to no light perception and complete ophthalmoplegia.

Results:
On examination, his vision was no light perception on the right and 20/30 on the left. His right pupil was fixed with a reverse afferent pupillary defect. He had proptosis, ptosis, facial numbness and ophthalmoplegia on the right but no disc edema or pallor. Intravenous steroids were initiated without improvement. A repeat CT with contrast showed an extensive soft tissue mass throughout the right pterygoid palatine fossa, orbital apex and right cavernous sinus. Biopsy revealed metastatic squamous cell carcinoma. He underwent radiation therapy which significantly improved his pain and prevented further vision loss in the left eye. The past excised forehead skin lesion was later confirmed as squamous cell carcinoma with clear margins.

Conclusions:
The incidence of perineural spread with SCC is rare. Although imaging is revealing in the majority of cases, there have been reports were imaging has been inconclusive when patients have presented early. In addition, since the disease typically presents later in life, the presence of concomitant medical issues can limit diagnostic testing. However, in an elderly patient with painful ophthalmoplegia and visual loss malignancy should be highly considered. Therefore, obtaining correct imaging is critical to make the diagnosis.

References:


Keywords: Squamous cell carcinoma, Ophthalmoloplegia, Orbital apex, CT scan, Tolosa Hunt syndrome

Financial Disclosures: The authors had no disclosures.
Pach Man: IgG4-Related Pachymeningitis as a Cause of Severe, Bilateral Vision Loss

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Introduction:
A patient presenting with severe, bilateral vision loss demonstrated retrobulbar optic neuropathy and leptomeningeal enhancement on MRI. Dural biopsy revealed IgG4 positivity, suggesting a diagnosis of IgG4-related pachymeningitis.

Methods:
Case report.

Results:
A 54-year-old male reported four months of progressive, bilateral vision loss, gait instability, and generalized weakness. He was no light perception in both eyes with amaurotic pupils. External, slit lamp, and cranial nerve exams were normal. Dilated fundoscopic exam revealed bilateral mild optic disc pallor. MRI revealed diffuse leptomeningeal enhancement. Fat saturated orbital images showed perineural enhancement of the left intracanalicular optic nerve. Dural biopsy revealed an infiltrate composed predominantly of lymphocytes and mature-appearing plasma cells. Immunostains for IgG4 showed 40% positive plasma cells and 15-20 IgG4 cells per high power field. IgG4 to IgG ratio was increased. Serum IgG showed elevated IgG4 subclasses.

Conclusions:
In the past, cases with similar findings received a diagnosis of idiopathic hypertrophic pachymeningitis (IHP). However, this entity may represent a type of IgG4-related disorder. Many previously identified conditions have been classified as IgG4-related diseases, including autoimmune pancreatitis, retroperitoneal fibrosis, and autoimmune cholangitis. They share strikingly similar histopathologic features despite varied sites of presentation. They have been linked to severe vision loss. Current literature does not mention this complaint in IgG4-related disease. However, review of cases previously diagnosed as IHP revealed a high percentage met the criteria for IgG4-related pachymeningitis. Therefore, previous reports of severe vision loss in IHP may represent unrecognized IgG4-related disease.

References:

Keywords: Pachymeningitis, IgG4, Optic neuropathy

Financial Disclosures: The authors had no disclosures.
Paraneoplastic Dermatomyositis Associated with CNS Chondrosarcoma

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Introduction:
Dermatomyositis is a cutaneous disease with a 30% association with malignancy (Callen 1993). A CNS location for malignancy is rare, with pulmonary and gynecological tumors being more commonly implicated (Stockton 2001). Furthermore, there is only one prior report of dermatomyositis secondary to chondrosarcoma (Mol 1986) which involved the tibia. We present the first case of paraneoplastic dermatomyositis secondary to chondrosarcoma of the central nervous system.

Methods:
Case report.

Results:
A 51 year old woman developed dermatomyositis, involving joint pain and rashes on the hands, face, and buttocks. No underlying cause was determined. Six weeks later, she presented with diplopia and right eyelid ptosis. Clinical evaluation revealed a right pupil-sparing partial third nerve palsy and sixth nerve palsy. There was also decreased sensation along the trigeminal nerve territory on the right. There was no evidence of optic neuropathy. MRI revealed a homogenous enhancing mass 5.5x 4.8x 5.5 cm in the right petroclival fissure, extending into the right middle cranial fossa and right cavernous sinus. Pertinent labs included: transaminits (AST 71, ALT 87), and elevated aldolase (10.9), ACE slightly elevated at 59, and ANA positive at 1:40. Creatinine kinase was normal and Mi-2 and anti-Jo antibodies were negative. She was treated with prednisone which improved her diplopia and rash. A craniotomy for debulking and biopsy revealed a myxoid chondrosarcoma, grade II. Her dermatomyositis is currently controlled with methotrexate.

Conclusions:
Laboratory findings for dermatomyositis typically include elevated transaminases, aldolase, creatinine kinase, and the presence of anti-Mi-2 or anti-Jo antibodies. In paraneoplastic dermatomyositis, as in this case, normal creatinine kinase (Fudman 1986) and negative anti-Mi-2 or anti-Jo antibodies (Love 1991) are typical. Paraneoplastic dermatomyositis should be included in the differential diagnosis for a patient with a rash and cavernous sinus involvement, and the appropriate imaging should be obtained to evaluate for malignancy.

References:

Keywords: Paraneoplastic dermatomyositis, Chondrosarcoma, Cavernous sinus

Financial Disclosures: The authors had no disclosures.
Case Report of Bilateral Compressive Optic Neuropathy Due to Idiopathic Hyperostosis of the Skull

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Introduction:
A 13 yo Filipino boy presented with progressive painless vision loss in his left eye x 3 months. In August, 2011, visual acuities were 20/40 OD and 20/80 OS. By December, 2011, his visual acuities were 20/60 OD and light perception OS. Funduscopic exam by a retina specialist and fluorescein angiogram were reportedly normal.

Methods:
Case report.

Results:
The patient was admitted in December, 2011. Our exam showed visual acuity 20/30- OD and CF OS. There was left RAPD, Ishihara color plates 11/14 OD. There was no proptosis or ptosis, and ductions were full OU. Dilated fundus exam showed trace pallor of the left optic nerve and normal fundus OD. CT head and orbits showed diffuse thickening of the skull and walls of the orbits resulting in narrowing of the optic foramina. There were no typical radiologic signs of fibrous dysplasia or neoplasm. MRI brain and orbits showed diffuse thickening of the skull and enhancement of the inner and outer tables, thickening of the anterior clinoids, and narrowing of the optic foramina. The patient was started on dexamethasone 4mg QID and underwent left pterional craniotomy with unroofing of the left optic canal. Labs: hypocalcemia 6.9, elevated PTH 70.8 Vitamin D was low 20.1 GNAS1 gene (McCune –Albright) – negative GH & IGF – WNL. Plain films of limbs: normal. The skull pathology showed fragments of sclerotic bone with marrow showing focal fibrosis and unremarkable cellular hematopoietic elements. There were no nodules or fibrous tissue.

Conclusions:
The cause of the hyperostosis of the skull was unclear. The treatment for his right optic neuropathy was not clear. Neurosurgery favored orbital decompression of the right optic canal. We recommended close follow up and surveillance. Followup nine months later shows visual acuity of 20/20- OD and HM OS with stable HVF OD.

Keywords: Optic neuropathy, Hyperostosis of the skull, Pediatric

Financial Disclosures: The authors had no disclosures.
Limited Wegener's Granulomatosis Presenting with Abduction Deficit in a Teenager

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Introduction:
Necrotizing granulomatous inflammation has a broad differential diagnosis, including inflammatory, infectious, and neoplastic processes (1). The classic histopathologic triad in Wegener’s granulomatosis is granulomatous inflammation, tissue necrosis, and vasculitis (2). We report a rare case of limited Wegener’s presenting with an isolated abduction deficit.

Methods:
Case report.

Results:
A 14 year-old girl presented with acute, painless binocular horizontal diplopia and was found to have an isolated abduction deficit of the right eye. Initial MRI brain with contrast was unremarkable, but suboptimal due to artifact from her braces. Diplopia resolved spontaneously but recurred six weeks later, when she developed painful periorbital edema, proptosis, and a nodule at the right lateral canthus. Braces were removed for repeat MRI, which showed enlargement of extraocular muscles, enhancement of the preseptal orbit and superior orbital fissure, and expansion of the right cavernous sinus. An extensive systemic inflammatory and infectious workup was negative. ANCA was negative. She was treated with oral prednisone for 4 months and improved dramatically, but developed recurrent facial and oral nodules acutely after stopping the steroid. Biopsy of the nodules showed necrotizing granulomatous inflammation with multinucleated giant cells. The granulomas had a linear and branching appearance, but were not clearly associated with vessels. In consultation with several treating services, the diagnosis of limited Wegener’s granulomatosis was reached and she ultimately remained free of relapses on methotrexate.

Conclusions:
Wegener’s granulomatosis has a limited form, which can present with orbital inflammation and can be ANCA-negative (3). This diagnosis should be considered, even in the absence of the full histopathologic triad (2). When not organ- or life-threatening, this disease can be treated with less toxic immunosuppressants such as methotrexate.

References:

Keywords: Wegener's granulomatosis, Orbital inflammation, Abduction deficit

Financial Disclosures: The authors had no disclosures.
Suprasellar Salivary-Gland Like Tumor: An Unlikely Location

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Introduction:
Salivary gland-like tumors in the sellar region are rare. They arise from salivary gland rests as remnants of Rathke’s pouch which is embryonically derived from the oropharyngeal cavity.

Methods:
We present a 71 y.o black female with sudden onset of vision loss in the right eye in April 2009. She has hypertension, gastro-esophageal reflex and history of left submandibular gland adenoid cystic carcinoma treated in 1974 with radical left neck dissection and external beam radiation. On exam, best visual acuity was 20/40 OD and 20/20 OS with a right RAPD, and asymmetric pupils of 4.5mm and 3.5mm. Color plates and IOPs were normal OU. VF to confrontation showed bitemporal defects confirmed with automated fields. Anterior exam showed right upper lid ptosis. Motility revealed a right 3rd nerve palsy. The left was normal. Fundus examination showed pale optic nerves, right more than left.

Results:
MRI of the orbits revealed a cavernous sinus lesion with extension into the sellar and parasellar areas with encasement of the right optic nerve. The lesion had an appearance consistent with meningioma. Her pituitary function panel was normal. The patient underwent right craniotomy on 7/2009 with decompression of right optic nerve, which improved her extraocular motility. Metastatic work-up was normal. Sixteen months after completing radiation therapy, she developed panhypopituitarism, and a right cavernous sinus syndrome. MRI brain showed tumor regrowth into the right cavernous sinus. She underwent radical dissection with subsequent Cyberknife radiotherapy. The second biopsy revealed the same histopathology. The histopathology was initially obscure. Further analysis and immunohistochemical stains showed that the tumor was immunopositive for keratin, p63 (myoepithelial marker) and S-100 and was negative for pituitary hormones, among others. The Ki67 malignancy index was moderately high.

Conclusions:
The diagnosis was salivary-gland like myoepithelial neoplasm with prominent myxoid stroma. These tumor types are quite rare and should be considered in sellar-based tumors.

References:

Keywords: Salivary, Gland-like, Tumors, Sellar region

Financial Disclosures: The authors had no disclosures.
Progressive Visual Loss: The Plot Thickens

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Introduction:
We describe the case of an 82-year-old male with a progressive optic neuropathy associated with Propionibacterium acnes pachymeningitis.

Methods:
Case presentation.

Results:
An 82-year-old male veteran presented with a 4-day history of painless vision loss in the OD. His ophthalmic history included sequential NAION in the OS 20 years prior and in the OD four months prior. He had a 10-year history of Churg-Strauss syndrome treated with oral steroids. On examination, acuity was 20/50 OD, down from 20/40 two months prior. Automated perimetry showed a new inferior arcuate defect in addition to his previous superior altitudinal defect OD. There was stable optic nerve pallor bilaterally without edema. Laboratory studies showed only a stable, elevated IgE level. Fluorescein angiogram was non-diagnostic. Temporal artery biopsy was negative. MRI showed diffuse pachymeningeal enhancement. LP showed normal opening pressure and CSF indices. No organisms were cultured. He received IV steroids over three days without change in vision. Dural biopsy showed evidence of chronic meningeal inflammation. Culture grew 2+ Propionibacterium acnes. Oral doxycycline was started. Post-operative MRI showed an abscess at the biopsy site and persistent pachymeningitis. The abscess was drained and broad-spectrum IV antibiotics were started. MRI one month later showed decreased inflammation at the biopsy site and resolution of pachymeningeal enhancement. His vision remained stable.

Conclusions:
Propionibacterium acnes was the only organism cultured from the dural biopsy. Our patient’s immunosuppression and the indolent nature of P. Acnes likely masked the signs and symptoms of his infection. The post-biopsy abscess grew Klebsiella oxytoca, and has no causal relationship to the optic neuropathy or pachymeningitis. Pachymeningeal enhancement persisted while our patient was on steroids, and resolved only after broad-spectrum antibiotic therapy. This clinical picture is most consistent with an infectious etiology, making this case a unique presentation of progressive optic neuropathy in the setting of P. acnes pachymeningitis.

References:

Keywords: Hypertrophic Pachymeningitis, Propionibacterium Acnes, Optic Neuropathy, Dural Biopsy

Financial Disclosures: The authors had no disclosures.
Optic Neuropathy in the Flesh

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Introduction:
A healthy male presented with acute vision loss consistent with a non-arteritic ischemic optic neuropathy. Although asymptomatic, a work up was initiated revealing systemic sarcoid. Sarcoid causing an ischemic optic neuropathy is rare, but should be considered in affected patients lacking typical vasculopathic risk factors.

Methods:
A 42 year-old healthy male presented with acute field loss in his right eye. He denied other symptoms except for mild retrobulbar discomfort. Acuity was 20/20 in both eyes. He had slight color desaturation with an afferent pupillary defect on the right. Visual field testing revealed an inferior, altitudinal field defect on the right. Fundus exam demonstrated a diffusely swollen, hyperemic optic nerve and a cotton wool spot, extending from the optic nerve to the macula. The left eye was unremarkable.

Results:
He underwent additional testing including blood work and imaging, significant for a low white count (3,300), ACE 113 (normal 0-67 u/L) and lysozyme 31 (normal 0-17 ug/mL). MRI of the brain and orbits with gadolinium was unrevealing but chest CT showed extensive pulmonary interstitial abnormalities and mediastinal lymphadenopathy. Biopsy revealed non-necrotic granulomas suggestive of sarcoidosis. He received five days of high-dose intravenous steroids followed by a slow prednisone taper over 14 months by his pulmonologist. At two months the optic nerve edema and cotton wool spot resolved, but the field defect improved minimally.

Conclusions:
Sarcoidosis has numerous ophthalmic manifestations but affects the optic nerve in only 1-5% of cases in the form of retrobulbar optic neuritis, perineuritis, neuroretinitis, non-arteritic ischemic optic neuropathy, or an optic nerve head granuloma. Due to the various optic nerve manifestations, one should consider sarcoid when patients present with the above findings. A work up is recommended in asymptomatic patients, as in this case, since multi-organ disease can occur with minimal symptoms.

References:

Keywords: Optic neuropathy, Sarcoidosis, Optic disc edema

Financial Disclosures: The authors had no disclosures.
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Chronic Idiopathic Demyelinating Polyneuropathy with Ocular Motor Findings

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Introduction:
We present a case of recurrent abduction deficit in setting of chronic inflammatory demyelinating polyneuropathy.

Methods:
A 32 year-old man experienced double vision on horizontal gaze, hand paresthesias and bilateral foot drop fifteen years ago. He was diagnosed with Guillain-Barre. He was treated with IVIG and plasma exchange with no response and only improved on steroids. Two years later, his diplopia returned while off steroids. Nine years later, he had parasthesias in his feet and fingers. He had no other neurological dysfunction and no family history of neurological diseases.

Results:
His visual acuity was 20/20 in both eyes. There was small esodeviation in primary gaze that increased in left lateral gaze and a small exodeviation in right gaze. He had a marked saccadic delay and limitation of the excursion of the left abducting eye. He had loss of deep tendon reflexes, but otherwise no sensory or motor dysfunction. MRI showed areas of swelling along all three divisions of both trigeminal nerves and extensive cervical to sacral nerve root enlargement. Electroneuromyography showed reduced conduction velocity in sensory and motor nerves. An auricular nerve biopsy showed marked loss of myelin with prominent “onion bulb” formation. Given this information, a presumed diagnosis of chronic inflammatory demyelinating polyneuropathy was made.

Conclusions:
Chronic inflammatory demyelinating polyneuropathy (CIDP) is a symmetric polyradiculoneuropathy that affects motor and sensory fibers of proximal and distal limbs but less than 5% develop external ophthalmoparesis. In 2002, a case of CIDP reported an isolated ptosis and adduction deficit, whereas our patient had abduction deficit. Usually ocular manifestations occur symmetrically, but in our patient and in this case the oculomotor deficit was relatively unilateral despite bilateral symmetrical MRI enhancement along the nerves. The severity and symptoms experienced vary considerably between patients; therefore, making it difficult to diagnose CIDP early.

References:


Keywords: CIDP, Onion Bulb, Polyneuropathy, Abduction Deficit, Saccadic Delay

Financial Disclosures: The authors had no disclosures.
Deficient Contrast Visual Acuity in Patients with Multiple Sclerosis Degrades Gait Performance Under Conditions of Low Illumination

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Introduction:
To determine the effect of deficient contrast visual acuity on gait performance under conditions of bright and dim illumination. Contrast visual acuity is deficient in MS and associated with optic nerve fiber depletion, demonstrated by optical coherence tomography. Studies on functional implications on motor performance particularly in conditions of low illumination are limited.

Methods:
Twenty two MS subjects versus matched controls. Tests: EDSS scoring, contrast sensitivity acuity (SLOAN) 100%, 2.5%, and 1.25% efficiency; Spectral optical coherence tomography; randomized analysis of multiple gait parameters (GAITRite FAP) in conditions of simple and maze walking in high/low illumination (>80 & <4 candela) with/without blue blocking lenses. Exclusion criteria: VA <20/50, comorbid ophthalmologic disorders, EDSS >6.

Results:
Significantly poorer contrast visual acuity was found in the MS group at 2.5% and 1.25% visual efficiency. MS patients had reduced nerve fiber bundle thickness and thinner ganglion cell layers (GCL+IPL) compared with matched controls, a finding that correlated directly with reduced performance on the Sloan chart (Correl >50%). Such visual deficiency significantly degraded motor performance under low (p <0.02) but not high contrast conditions. A correlation existed between disease severity (EDSS) and FAP scores; those with EDSS score of 4 or greater performed significantly poorer than those with a score of less than 4. In an effort to improve ambulatory function, blue blocking lenses, reported to improve contrast sensitivity, showed improvement in dim illumination in more severely affected MS patients compared with controls, especially for complex walking tasks (p-value 0.002). Neither group improved in high illumination. Multivariate analysis of performance vs. visual function and OCT findings will be discussed.

Conclusions:
Reduced contrast sensitivity acuity degrades motor performance in MS in dim illumination. In analyzing motor performance, blue blocking lenses produced significant improvement in individuals with higher EDSS scores (>4).

Keywords: Multiple Sclerosis, Contrast Visual Acuity, ambulatory function, Blue blocking lenses, Optical Coherence Tomography (OCT)

Financial Disclosures: The authors had no disclosures.
Correlation of Peripapillary Retinal Nerve Fiber Layer Thickness (prNFLT) and Macular Thickness (MT) as Measured by spectral domain OCT (SD-OCT) in Patients with Severe Vision Loss in End Stage Optic Neuropathy (ESON)

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Introduction:
Optical Coherence Tomography (OCT) of the optic nerve head and macula is a useful adjunct test in monitoring optic nerve disease. Few studies have looked at the utility of OCT in severe vision loss states. Visual acuity in patients with ESON may be classified using the semi-quantitative scale of counting finger (CF), hand motion (HM), light perception (LP), and no light perception (NLP). Correlating prNFLT or MT with severe vision loss in patients with optic neuropathies may help determine threshold for change, prognosis for change and help guide treatment to prevent further vision loss.

Methods:
A retrospective chart review was performed of patients diagnosed with optic neuropathy and best corrected visual acuity (BCVA) of CF, HM, LP, and NLP. Inclusion criteria were a stable BCVA, disease state present for at least 9 months, and SD-OCT of the MT or prNFLT. The central volume of retinal nerve fiber layer, ganglion cell layer, and inner plexiform layer was measured with a manual segmentation procedure. Exclusion criteria were a Q number of <15 and confounding ophthalmic pathology. Non-parametric Spearman Correlation was used to analyze the data.

Results:
68 eyes in 61 patients were reviewed. Of the 68 eyes, 47 had SD-OCTs of the PRNFL and 19 had SD-OCTs of the central macula and met all inclusion and exclusion criteria. Statistical analysis indicated there was no significant difference in PRNFLT or inner macular volume among the four SVL groups.

Conclusions:
Our data suggests that there is no significant difference in SD-OCT prNFLT or MT among low vision states in ESON. Possible explanations include SD-OCT may be detecting non-axonal content when measuring prNFLT, such as glial content and blood vessels. Signal-Noise ratio may be suboptimal in end stage disease. Recruitment of a larger dataset is needed to confirm these findings.

Keywords: Optic Neuropathy, OCT, Low Vision

Financial Disclosures: The authors had no disclosures.
Homonymous Hemianopia with a Normal MRI: When to Consider Retinal Mimics of Neurologic Disease

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Introduction:
A 34 year old man with history of retinal detachment presented with progressive, painless vision loss in his left eye over 5 days. He noticed “pulsating” in his missing visual field, but no flashes or floaters. Examination by a retina specialist was remarkable for nasal hemi-field loss in his left eye.

Methods:
Neuro-ophthalmologic examination showed visual acuity of 20/15 OU, normal color vision, a 0.6 log unit relative afferent pupillary defect OS and normal posterior examination. Humphrey visual fields showed a nasal defect OS and temporal defect OD, suggestive of an incongruous homonymous hemianopia. Over the next five days, his vision OS worsened, with photopsias, headache, and photophobia. Repeat examination showed normal visual acuity and color vision OD and 20/800 and no color vision OS, with a >1.8 log unit RAPD OS. There was mild attenuation of the arteries OS with mottling of the macula, and normal disc appearance.

Results:
MRI and laboratory testing were unremarkable. Full-field electroretinogram showed global cone receptor dysfunction OU. Red-free photographs showed a white outer retinal irregular annular band of deep retinal opacification corresponding to his central scotoma OS. He was started on valacyclovir.

Conclusions:
Despite his examination and visual fields being initially concerning for a post-chiasmal lesion, his eventual diagnosis was acute annular outer retinopathy, a suspected variant of acute zonal occult outer retinopathy (AZOOR). Bilateral involvement is rare and it is an important consideration in the differential diagnosis of sudden unilateral or bilateral vision loss.

References:

Keywords: Visual Field Defect, Retinal Mimic of Neurologic Disease, Vision Loss

Financial Disclosures: The author had no disclosures.
Characteristics of Patients with Primary Divergence Insufficiency

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Introduction:
Divergence Insufficiency (DI) is a comitant esotropia at distant fixation, with minimal or no deviation at near and full ductions. When this occurs in patients with no associated neurologic disease, the deviation is considered primary. The purpose of this study was to characterize the patients with this disorder in terms of demographics, clinical presentation, ocular examination, treatment and prognosis.

Methods:
We reviewed all cases diagnosed with primary divergence insufficiency in our neuro-ophthalmology unit between the years 2005 and September, 2012. Patients with known neurologic diseases or positive findings in neurologic examination or findings on imaging explaining divergence weakness were considered secondary DI, and therefore were excluded. We collected demographic data as well as data on background diseases, duration of symptoms, visual acuity, refraction, angle of deviation at near and distance, treatment, duration of follow up and patient satisfaction.

Results:
Thirteen patients were classified with primary DI, of whom 10 were men. The mean age was 71 years. Ten patients suffered from at least one condition considered a risk factor for micro vascular disease, i.e. Diabetes mellitus, or hypertension. Mean duration of symptoms was 32 months ranging from 4 -144 months. Mean BCVA in both eyes was 20/30 with refraction between minus 2.5 spherical equivalent to plus 2.00. Mean horizontal deviation on far fixation was 10 PD (range: 6-14PD) and on near fixation 1 PD (range: 0-6PD). Eight patients were treated with prismatic correction, one patient improved spontaneously and the remaining four patients were offered prisms but chose not to use them. Follow-up lasted between 1-22 months (mean: 6 months). There was high patient satisfaction in all those treated with prisms.

Conclusions:
Primary divergence insufficiency is a condition in the older population which has an insidious onset and runs a benign course. High patient satisfaction is achieved with prismatic treatment.

Keywords: Primary divergence Insufficiency, Comitant esotropia

Financial Disclosures: The authors had no disclosures.
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**Microperimetric Assessment of Scotopic Relative to Photopic Sensitivity to Assist in the Differential Diagnosis of Occult Retinopathy Versus Optic Neuropathy**

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**Introduction:**
Bilateral, paracentral scotomata, without optic disc pathology or gross retinal pathology on color fundus photographs, may be due to bilateral retrobulbar optic neuropathy or occult retinopathy. Here we present a case where point-by-point differential visual field assessment of day and night vision suggested a preferential reduction of rod photoreceptor function and therefore a retinal location of the disease process.

**Methods:**
Observational case report.

**Results:**
During a study leave abroad, a 22 year-old woman noticed bilateral blurring of central vision upon awakening. Flu-like symptoms had begun a few days earlier. She was on no medication except for oral contraceptives. The patient was initially seen elsewhere and found to have dense bilateral paracentral scotomata but best-corrected visual acuity 20/20 in both eyes. Ophthalmoscopic findings were described as normal, which prompted the suspicion of bilateral retrobulbar optic neuritis and MRI of the brain and orbits, which was normal. The patient was referred to our institution three weeks after the onset of visual symptoms. Visual field examination registered to fundus locations on a reference image (microperimetry) was made at both scotopic and photopic background illumination levels and revealed relative paracentral scotomata in both eyes that were roughly twice as deep in the dark-adapted as in the light-adapted state. Corresponding wedge-shaped areas of hyperreflectivity and photoreceptor layer irregularity were found on confocal laser-scanning ophthalmoscopy and optical coherence tomography. The patient was diagnosed with acute macular neuroretinopathy. At 15 months follow-up little functional or morphological improvement was seen.

**Conclusions:**
Of the three modalities that documented a retinal origin of the visual symptoms in acute macular neuroretinopathy, the semipreferential affection of rod photoreceptors may have the advantage of being applicable also in other occult retinopathies. This may assist the differential diagnosis vis-à-vis retrobulbar optic neuritis and toxic-/nutritional optic neuropathy.

**Keywords:** Microperimetry, Acute Macular Neuroretinopathy, Paracentral Scotomata, Occult Retinopathy, Optic Neuropathy

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A Case of Bacterial Meningitis Complicating Unilateral Vestibular Dysfunction and Bilateral Deafness

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Introduction:
Previous studies have reported several cases of hearing loss and gait disturbance during the meningitis. But there have been few reports focusing on the vestibular dysfunction associated with the meningitis. We report a case of meningitis, which had the concurrent unilateral vestibular dysfunction and bilateral cochlear dysfunction.

Methods:
A 59-year-old woman visited our emergency department with fever and drowsiness. She had been suffering from headache and vomiting for a month before admission. Neurologic examination revealed drowsiness, neck stiffness and forced conjugate eye deviation to right. MR imaging suggested diffuse leptomeningeal enhancement. CSF examination revealed pleocytosis with neutrophil dominance and elevated protein content. CSF latex agglutinin test was positive in Streptococcus pneumoniae antigen. We performed electroencephalography, but there was no epileptiform discharge despite of the persistent rightward eyeball deviation. Under the clinical diagnosis of bacterial meningitis, intravenous antibiotics and steroid were administered. And then she showed marked attenuation of neurological impairments. Two days after the administration of intravenous antibiotics, she became alert and eyeball deviation disappeared. On neurological examination she showed spontaneous left beating nystagmus, catch-up saccade at head impulse test to the right side. And at her audiologic examination showed sensorineural-type hearing impairment of both ear. Right ear was 49% weaker at caloric test. Subjective visual vertical and fundoscopic exam suggested clockwise ocular torsion. Those findings were compatible with right vestibulopathy. Also, bilateral hearing impairment or cochleopathy was suggested.

Conclusions:
We described a bacterial meningitis patient with a concurrent vestibulocochlear dysfunction. We documented vestibulocochlear dysfunction with neurootologic/neuroophthalmologic evaluation. We suggest neurootologic/neuroophthalmologic assessment is necessary in addition to the other tests for meningitis in case of the patient with vestibular dysfunction suspected, especially with forced conjugate eyeball deviation during impaired consciousness.

Keywords: Meningitis, Vestibulopathy, Deafness

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A Case Report of Four and a Half Syndrome

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Introduction:
Being able to localize a lesion based on the physical exam findings is critical in neuro-ophthalmology. Crossed localizing signs are particularly helpful in brainstem lesions. We report a patient with a IV nerve palsy ipsilateral to an internuclear ophthalmoplegia. This combination localized the lesion to the dorsal midbrain.

Methods:
We present a 69-year-old man who awoke with blurred vision, disequilibrium, and vertical diplopia. He denied any nausea, vertigo, or other symptoms of central nervous system dysfunction. He reported a history of controlled diabetes, hypertension, hyperlipidemia and coronary artery disease.

Results:
His physical exam was unremarkable except for the oculomotor evaluation. This showed a primary position left beating nystagmus and a left hypertropia that was greatest looking down and to the right. It also increased on left head tilt. Double Maddox rod testing showed a baseline excyclotorsion. During head tilt to the right, extorsional saccades were normal in the right eye but the patient had no intorsional saccades with the left eye. Torsional saccades were normal bilaterally with head tilting to the left. Although versions were normal, there was marked slowing of adducting saccades to the left. These findings can be localized to a small right-sided paramedian lesion in the right dorsal midbrain involving the right IV nerve nucleus, causing a left IV nerve palsy, and the right medial longitudinal fasciculus, causing an ipsilateral internuclear ophthalmoplegia. MRI of the head confirmed the predicted abnormality. His sensation of imbalance resolved quickly. On follow-up after 5 months, the nystagmus had nearly resolved. Although the hypertropia had improved slightly, the diplopia persisted.

Conclusions:
We were able to find only one report from 1992 of a similar lesion. To aid in remembering this syndrome, we suggest calling this combination a “four and a half syndrome.”

Keywords: Internuclear Ophthalmoplegia, Fourth nerve palsy, Brainstem lesion, Diplopia, Saccades

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Granulomatous hypophysitis masquerading as hemorrhagic pituitary adenoma

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Introduction:
Optic neuropathy from granulomatosis with polyangiitis (GPA), formerly termed Wegener's, is rare. Even more unusual is blindness related to hypophysitis in this vasculitis. We encountered a perplexing case of acute vision loss ultimately found to be result from GPA

Methods:
A retrospective case presentation.

Results:
19 year old, obese, young woman presented with acute vision loss OS associated with left sided headache. Slight optic nerve swelling was detected OS by her ophthalmologist and neuro-imaging performed. Past medical history is significant for migrane, polycystic ovarian syndrome and hypertriglyceremia. Review of symptoms is notable only for increased lethargy. Exam showed BP 135/91, weight 97 Kg, height 5’9”, BMI 31.5. Vision was 20/20 OD and NLP OS. Pink and healthy nerve OD and pink nerve with only subtle nerve fiber layer edema OS. Neuroimaging showed enlargement of pituitary consistent with macroadenoma and chiasmatic compression. The pituitary mass was resected and showed paucicellular degenerative material containing fragments of squamous epithelium but no histologic evidence of a pituitary adenoma. Lumbar puncture had opening pressure of 35 cm H20. Ventrivoperitoneal shunt was inserted to protect the right eye from the raised intracranial pressure. Cerebral spinal fluid protein 27 mg/dL (15-45), glucose 73 mg/dL (45-75) normal cell count and differential, no bacteria, no oligoclonal bands, angiotensin converting enzyme 1.4 U/L (0.0-2.5). Blood work showed positive antinuclear antibody with titer >1:160, antinuclear cytoplasmic antibody negative with antmyeloperoxidase and anti protease 3 negative, Sedimentation rate 47 mm/hr (0-20), RPR non-reactive, c-ANCA negative, and NMO IgG negative. A complex medical course then evolved. Histopathology from second pituitary resection revealed necrotizing inflammation and neutrophilic microabscesses with abundant acute and chronic inflammation.

Conclusions:
GPA can be a challenging diagnosis to establish particularly in the setting of a grossly enlarged sella mass and obesity. Steroid responsive, variable neurologic compromise can overlap with the presentation of neurosarcoid. Ultimately histopathologic data can confirm clinical suspicion in an atypical presentation.

References:

Keywords: Wegeners, Hypophysitis, Optic Neuropathy, Visual Field Defect

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