North American Neuro–Ophthalmology Society  
**Poster Presentations**  
Columbia Room  
Tuesday, February 8, 2011  
6:00 p.m. – 10:00 p.m.  
*Authors will be standing by their posters at the hours indicated below:*  
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**Even-Numbered Posters:** 7:30 p.m. – 8:15 p.m.  
*Tour of Posters: 8:30 p.m. – 10:00 p.m. (Pacific Ballroom)*  
*Tour Moderators: Peter Savino, MD and Steve Galetta, MD*

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North American Neuro–Ophthalmology Society
Poster Presentations
Columbia Room
Tuesday, February 8, 2011
6:00 p.m. – 10:00 p.m.
Authors will be standing by their posters at the hours indicated below:
Odd–Numbered Posters: 6:45 p.m. – 7:30 p.m.
Even–Numbered Posters: 7:30 p.m. – 8:15 p.m.

*Tour of Posters: 8:30 p.m. – 10:00 p.m. (Pacific Ballroom)
Tour Moderators: Peter Savino, MD and Steve Galetta, MD

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Multifocal Objective Pupil Perimetry in Multiple Sclerosis

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1The Canberra Hospital, Canberra, ACT, Australia, 2The Australian National University, Canberra, ACT, Australia

Introduction:
An earlier study using multifocal visual evoked potentials1 indicated that sparse multifocal stimuli diagnosed relapsing remitting multiple sclerosis (MS) patients well whether or not they had experienced optic neuritis (ON). The purpose of this study was to examine the diagnostic power of multifocal pupillographic objective perimetry (mfPOP) in MS.

Methods:
85 subjects with MS (49.8 ± 11.3 yr, 62 women) and 35 normal controls (47.9 ± 16.8 yr, 22 women) were enrolled. Two MS patients had primary and 11 secondary MS (PS) while the remainder had relapsing remitting MS (RR). The TFA stimuli tested 44 regions/eye within the central 60º delivered as dichoptic sparse stimuli at a mean rate of 1/s/visual field region. The total stimulus duration was 4 min. Receiver operator curves (ROC) were calculated for RR and PS groups vs. controls. Subgroup analyses of RR patients with and without a history of ON were performed.

Results:
EDSS scores were 3.53 ± 1.04 (mean ± SD) for RR and 5.90 ± 1.43 for PS. ROC plots revealed that %AUC was 75.0 ± 3.31 (mean ± SE) for the 144 RR eyes and 94.8 ± 3.55 for the 26 PS eyes. %AUCs were 75.7 ± 4.48 and 75.4 ± 3.84 for RR patients with and without ON. %AUC for RRMS with EDSS >= 5 (5.29 ± 0.57) was 91.4 ± 8.1.

Conclusion:
mfPOP performed well, particularly in patients with a higher EDSS scores in both RR and PS patients. A past history of ON had no significant effect, suggesting that the results were more dependent on "secondary" degeneration than a history of inflammation. Using mfPOP it was possible to examine the visual fields of both eyes in just 4 min. This technology is potentially useful in the diagnosis and follow-up of MS patients.

References:

Key Words: Multiple Sclerosis, Pupil, Perimetry, Multifocal Pupillographic Objective Perimetry

Financial Disclosure: Maddess and James hold IP, share options in Seeing Machines Pty.
Alemtuzumab Improves Contrast Sensitivity in Relapsing-Remitting Multiple Sclerosis Patients

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¹University of Pennsylvania, Philadelphia, PA, United States, ²Genzyme Corporation, Cambridge, MA, United States

Introduction:
Alemtuzumab is a humanized monoclonal antibody that alters the circulating lymphocyte pool. In a 3-year, phase 2 trial, alemtuzumab was superior to subcutaneous IFNB-1a in patients with early, active relapsing-remitting MS (RRMS), significantly reducing relapse rate, risk for sustained accumulation of disability, and mean disability. Contrast sensitivity is a test that may distinguish MS patients from controls and effectively measure visual function. We evaluated alemtuzumab’s effects on contrast sensitivity in patients with RRMS.

Methods:
Patients (n=334) were randomized 1:1:1 to IFNB-1a (44mcg SC 3x/week) or 24 mg or 12 mg/day alemtuzumab (IV-administered, 2 or 3 brief annual cycles). Pelli-Robson contrast sensitivity was measured for each eye at baseline and quarterly by a masked rater. Clinically significant improvement in contrast score was defined as ≥0.30 log unit change, sustained for ≥3 months. Time to sustained improvement was analyzed with proportional-hazards models and Kaplan-Meier estimation, accounting for within-patient inter-eye correlations. A 5-category ordinal scale ranging from improvement in score for both eyes to decrease for both eyes was evaluated with repeated-measures proportional odds models.

Results:
On an ordinal scale, alemtuzumab patients had a higher probability of achieving better responses compared to IFNB-1a patients (HR: 1.73; 95% CI: 1.18, 2.55; p=0.0054). Probability of sustained visual improvement was greater among alemtuzumab patients compared to IFNB-1a: Kaplan-Meier estimate of proportion of eyes with improvement at 36 months was 39.3% (95% CI: 24.7, 53.9) for pooled alemtuzumab patients vs. 27.7% (95% CI: 18.6, 36.7) for IFNB-1a. Alemtuzumab patients trended toward sustained improvement through 36 months compared with IFNB-1a patients (hazard ratio [HR]: 1.53; 95% CI: 0.98, 2.38; p=0.062).

Conclusion:
Alemtuzumab was associated with improved contrast sensitivity in patients with RRMS. Tests of low-contrast vision are effective measures of visual outcome for MS trials.

References:

Key Words: Contrast Sensitivity, Pelli-Robson, Multiple Sclerosis (MS), Vision, Alemtuzumab

Financial Disclosure: Dr. Balcer has received consulting fees and honoraria from Biogen-Idec and Bayer for development of visual outcomes for MS trials. Dr. Galetta has received speaking honoraria from Biogen-Idec and Teva. Dr. Maguire has nothing to disclose. Drs. Palmer, Margolin, and Rizzo are employees of Genzyme Corporation.
Introduction:
Fractionated stereotactic radiotherapy (FSRT) is an accepted adjunct in the treatment of meningiomas threatening
the anterior visual pathways (AVP). FSRT can be delivered using several devices including the Gamma
knife, Cyberknife, Tomotherapy, and linear accelerator (LINAC) systems. Comparative long-term follow-up for these
techniques is currently being compiled.

Methods:
A comparison of the success and complication rates for all patients with AVP meningiomas following LINAC FSRT
using the multiple, non-coplanar, dynamic conformal rotation paradigm with those reported for other FSRT techniques.

Results:
Of 87 patients with AVP meningiomas, 17 were referred for FSRT, 16 of who completed > 12 months' follow-up (mean
39 mo). Tumor control was achieved in 14/16 cases, with three meningiomas shrinking in size after radiation. Two
meningiomas progressed, one in an area outside of the radiation field. Visual functions improved or stabilized in 14
(88%) patients and worsened in two (12%).

Conclusion:
LINAC FSRT using the multiple non-coplanar dynamic rotation conformal paradigm may be offered to patients with
AVP meningiomas with results comparable to those reported with other FSRT techniques.

Key Words: Meningioma, Fractionated Stereotactic Radiotherapy, LINAC, Anterior Visual Pathways, Radiation

Financial Disclosure: None
Pattern of Retinal Ganglion Cell Loss in Dominant Optic Atrophy due to OPA1 Mutations

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Introduction:
Autosomal dominant optic atrophy (DOA) is the most common inherited optic nerve disorder and about 60% of patients harbor pathogenic mutations in the OPA1 gene.1-3 Although the pathological hallmark of DOA is the preferential loss of retinal ganglion cells (RGC), up to 20% of patients with OPA1 mutations will develop a more severe disease variant (DOA+) with additional neuromuscular features.4 In this prospective, observational case series, optical coherence tomography (OCT) was used to define the pattern of retinal nerve fiber layer (RNFL) loss in patients with both the pure and complicated forms of DOA, and to document the changes in RNFL thickness seen with disease progression.

Methods:
Forty patients with a molecular diagnosis of DOA due to OPA1 mutations were prospectively recruited from our neuro-ophthalmology clinic: 26 patients with isolated optic atrophy, and 14 patients manifesting DOA+ features. Peripapillary RNFL thickness was measured with the Fast RNFL (3.4) acquisition protocol on a time-domain OCT platform.

Results:
There was a statistically significant reduction in average RNFL thickness in the OPA1 group compared with normal controls (P < 0.0001). The percentage decrease was greatest in the temporal quadrant (59.0%), followed by the inferior (49.6%), superior (41.8%), and nasal (25.9%) quadrants. Patients with DOA+ features had worse visual outcomes compared with patients with pure DOA (P = 0.0018). Except in the temporal quadrant, RNFL measurements were significantly thinner for the DOA+ group. There was an inverse correlation between average RNFL thickness and LogMAR visual acuity (P < 0.0001), and this relationship was also significant for the inferior, superior, and nasal quadrants on subgroup analysis.

Conclusion:
Patients with OPA1 mutations exhibit a distinct pattern of RGC loss, with early severe involvement of the papillomacular bundle and relative sparing of the nasal fibers. RNFL thinning is progressive and is more pronounced in patients with DOA+ phenotypes.

References:

Key Words: Autosomal Dominant Optic Atrophy, OPA1, Mitochondrial DNA, Optical Coherence Tomography, Retinal Ganglion Cells

Financial Disclosure: None
Poster 5*

Postintervention Status in Ocular Myasthenia Gravis: Effects of Treatment

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Introduction:
Evidence-based strategy of treatment of ocular myasthenia gravis (OMG) is still not sufficient. The role of thymectomy is in controversy, especially in non-thymomatous OMG. This study was performed to assess the effect of different therapies on post-intervention status in patients with OMG.

Methods:
Post-intervention status of OMG patients with onset age ≥15 years old who had one of the following four therapeutic interventions were evaluated according to MGFA standard at 3, 6, 12 months, 1, 2 and 5 years after treatment: only cholinesterase inhibitor (o-CH), only cortical steroid (o-CS), only thymectomy (o-T), and thymectomy combined with cortical steroid (T-CS). Patients who at least reached Minimal Manifestations (MM) were considered effective. The percentages of effectiveness were statistically compared among four different therapeutic groups at different time points to see the trend of effect of different treatments on OMG.

Results:
141 OMG patients with mean age of 42±18 yrs were included. There were 76 male and 65 female patients. At baseline, except the mean age of o–CH group was statistically higher than the other three groups, the other clinical features were similar. The effectiveness was similar between all groups at 3 months after treatment. The o-T and T-CS groups showed ascending trends of effectiveness, reaching 37.0% and 64.3% respectively at 2 years, 46.7% and 71.4% respectively at 5 years after treatment, while the o-CS groups showed a highest effectiveness at 6 months but decreased after that. At 6 months, the effectiveness of o-CS(42.9%) and T-CS group(50%) were significantly higher than the other two groups(p=0.01, p=0.005) while at 5 years only T-CS group kept the significantly higher effectiveness(71.4%,p=0.02).

Conclusion:
Combination of steroid and thymectomy showed a better and longstanding effectiveness in postintervention status of OMG, suggested being a better choice of OMG treatment.

References:

Key Words: Myasthenia Gravis, Ocular, Treatment, Thymectomy, Postintervention

Financial Disclosure: None
Progression of Visual Field Defects in Patients with Optic Nerve Drusen: Follow-up with Automated Perimetry

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Introduction:
The majority of patients with optic nerve drusen (OND) manifest visual field defects\(^1\), which, rarely, can be blinding. Descriptions of the rate of visual field loss progression in OND are lacking from the literature. A literature search revealed a single case series from 1957 that detailed the change in Goldmann visual fields in 14 patients with an unspecified average time of follow up\(^2\).

Methods:
We reviewed 274 charts with a diagnosis of OND (ICD9 377.21). We excluded eyes with an incorrect diagnosis, fewer than two reliable 24-2 visual fields, confounding diagnoses (AION, glaucoma, retinal pathology affecting visual fields, brain pathology affecting visual fields), and eyes with less than 6 years follow-up. We calculated the change in mean deviation (MD) on Humphrey visual field (HVF) 24-2. To be considered reliable, a field had to contain fewer than 15% fixation losses, 15% false positives, and 30% false negatives.

Results:
We included 23 eyes of 16 patients. Mean follow up time was 9.7 years. Average change in MD was -0.78 dB (standard deviation 2.54 db). The majority of eyes did not display clinically significant decreases in MD and only 3 eyes lost more than 3 db over the course of their follow-up. Only one eye, followed for 12 years, lost more than 5 db.

Conclusion:
This is the first attempt to use automated perimetry to document the natural history of visual field defects in OND. The vast majority of eyes with OND in our study had stable visual fields over a nearly 10-year period. We will be reviewing additional charts between now and NANOS 2011 and will have a larger data set to report at that time.

References:

Key Words: Optic Nerve Drusen, Automated Perimetry, Visual Field Defects

Financial Disclosure: None
Correlating MRI Texture Measures of Tissue Integrity with Structural and Functional Assessments of Vision in Optic Neuritis

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Introduction:
Optic nerve function and structure can be reliably quantified, which makes it an ideal model to study the consequences of central nervous system (CNS) injury in multiple sclerosis (MS). The objective of this study was to correlate lesion texture with structural and functional measures of vision after acute optic neuritis (ON).

Methods:
Fourteen patients with acute ON were evaluated within 15 days of symptom in an ongoing prospective study. Optic nerve lesion length, gadolinium (Gd)-enhancing activity, and optic nerve area ratio was evaluated on 3T MRI at baseline and after 6-months. Lesion texture was analyzed based on a spatial-frequency algorithm concurrently on the coronal STIR images showing maximal lesion area. Retinal nerve fiber layer thickness (Zeiss Stratus III) and logarithm of minimum angle resolution (logMAR) visual acuity were assessed at the same time points.

Results:
Acute lesions were seen in all ON eyes (mean length=14.2 mm) and in one fellow eye (4 mm); 9/15 lesions had Gd-enhancement. Relative to the fellow eyes, there was a tendency for texture to be coarser (0.46 vs 0.38, p=0.07) and RNFL to be thicker (148 mm vs 86 mm, p=0.09) in ON eyes at baseline. Visual acuity was markedly decreased in ON eyes (0.5 vs 0.05, p<0.01), which correlated with acute lesion texture (p<0.01). At month 6, the difference between ON and fellow eyes was smaller in each assessment (p>0.05) and none correlated with logMAR visual acuity. Vision improved ON eyes by 0.49 (p<0.01); RNFL decreased by 26 mm (p=0.03); and lesion length decreased by1.3 mm (p=0.09) at month 6 compared to baseline.

Conclusion:
MRI texture measured tissue integrity is a sensitive marker of visual function associated with an acute ON event. Future work will test the predictability of texture to visual outcome, which recovers despite residual structural damage in the optic nerve.

References: None

Key Words: Optic Neuritis, Magnetic Resonance Imaging, Texture Analysis, Retinal Nerve Fiber Layer

Financial Disclosure: None
Poster 8*

Leber Hereditary Optic Neuropathy Gene Therapy Clinical Trial for G11778A Mutation: Serial Pretreatment Evaluation from Baseline to One Year

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Bascom Palmer Eye Institute, University of Miami, Miami, FL, United States

Introduction:
The aim of the preparatory phase of the LHON gene therapy clinical trial is to identify and characterize affected patients and carriers for planned gene therapy studies that will utilize “allotopic expression” by delivering copies of normal nuclear-encoded ND4 gene into the nuclei of retinal ganglion cells via an adeno-associated virus vector. The normal ND4 protein expressed in the cytoplasm is then imported into the mitochondria.

Methods:
LHON patients with acute visual loss, chronic visual loss and optic atrophy as well as their asymptomatic maternally-related relatives undergo ocular examination, visual fields, pattern electroretinogram (PERG), Cirrus OCT and fundus photography every 6 months. Blood samples for phosphorylated neurofilament heavy chain (NfH) quantitation of axonal loss are obtained.

Results:
A total 75 persons with G11778A have been recruited and more are needed. The one year results of serial evaluations of 19 patients and 16 asymptomatic carriers are available. LHON ETDRS acuity was stable at one year (mean 13.00 letters to 11.87 baseline). HVF mean defect (MD) was -24.66 one year and -25.31 baseline. PNF-H (square root) levels were unchanged (0.44 ng/ml one year to 0.41 baseline). Average RNFL was 56 μm one year and 61 μm baseline. PERG amplitudes were 0.47 μV one year and 0.49 μV baseline. In carriers, acuity was 86.84 one year and 87.41 baseline. MD was -1.37 one year and -1.82 baseline. PNF-H levels were 0.67 ng/ml one year and 0.59 baseline (p=0.13). PERG was 0.78 μV one year and 1.00 μV baseline (p=0.001).

Conclusion:
For affected LHON patients, clinical measures were stable through one year, and spontaneous improvement was rare (5%), thus making gene therapy improvements with injection of AAV containing a normal ND4 easy to detect within a year. Contact information for patient recruitment is available by googling “BPEI LHON”.

References:

Key Words: Leber Hereditary Optic Neuropathy, Gene Therapy

Financial Disclosure: None
Poster 9*

Long-Term Results of Optic Nerve Sheath Fenestration for Idiopathic Intracranial Hypertension: Earlier Intervention Favors Improved Outcomes

Stacy Pineles, Nicholas Volpe

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Introduction:
Idiopathic intracranial hypertension (IIH) is a common disorder affecting predominately young obese females. While weight loss and medical management are often sufficient treatments, surgical intervention is occasionally required. Optic nerve sheath fenestration (ONSF) is often employed therapy in patients with severe/progressive vision loss, yet its use is controversial.

Methods:
Records of all patients undergoing ONSF for IIH were reviewed. Data recorded included age, gender, symptoms, medical treatments, time to surgery, complications, subsequent procedures, and results of serial measures of visual acuity (VA), color vision, visual fields (VF), and optic nerve evaluations.

Results:
37 patients underwent 50 ONSFs. The mean age was 33±11 years (range 19-74) and 5 (13%) patients were male. Eight subjects underwent subsequent ventriculo-peritoneal shunts. The mean time to surgery was 36±54 weeks (range 0.5-226) and the mean post-operative follow-up 210±200 weeks (range 4-700). Complications included anisocoria (5%), diplopia (3%), and conjunctival abscess (3%). VA improved (<-0.2 logmar) in 22% of surgical and 17% of fellow eyes, was stable in 53% of surgical and 74% of fellow eyes, and deteriorated (>0.2 logmar) in 25% of surgical and 9% of fellow eyes. Mean deviation stabilized in 88% of surgical and 76% of fellow eyes followed with automated VFs. VF patterns more likely to resolve included enlarged blind spots and generalized constriction, while arcuate defects, nasal steps, and central scotomas were less often resolved. Multivariate analysis revealed that better pre-operative VA (p=0.01) and color vision (p=0.002) and earlier intervention (p=0.04) were associated with VA and VF stabilization. In general, optic nerve edema resolved first temporally and last nasally, with the earliest resolution at 2 weeks.

Conclusion:
ONSF is a useful surgical technique to stabilize VA and VF. Our results are comparable with previous series, and were best in patients with better pre-operative VA, color vision, and in those with earlier surgical intervention.

References: None

Key Words: Idiopathic Intracranial Hypertension, Optic Nerve Sheath Fenestration, Optic Nerve Edema

Financial Disclosure: None
Poster 10*

Is Review of Video Taped Exams of Ocular Motility Reliable for Surveys of Eye Movement Disorders in Populations with Neurodegenerative Disease?

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¹University of Illinois College of Medicine, Chicago, IL, United States, ²University of Pennsylvania School of Medicine, Philadelphia, PA, United States, ³University of Iowa College of Medicine, Iowa City, IA, United States

Introduction:
Neuro-ophthalmic findings are useful for characterizing neurological disease because of the multitude of diverse anatomical regions involved in vision and ocular-motor control. Objective characterization of ocular-motor findings typically relies on sophisticated eye movement recording devices that are not widely available. We measured the agreement between expert clinicians reviewing video taped examinations to test the hypothesis that this method is reliable for defining ocular motility disorders in a population with neurodegenerative disease.

Methods:
Subjects underwent a standardized ocular motility examination that included gaze fixation, voluntary saccades, reflex saccades, smooth pursuits, eyelid function, and Bell's phenomenon. Two senior neuro-ophthalmologists independently reviewed videos of the examinations and rated each task using ordered categories. Ten randomly chosen videos were presented to each reviewer a second time. Intra-rater and inter-rater correlations were assessed using Cohen’s Kappa statistic (K).

Results:
100 subjects (age 58.6 +/- 15.4 years, 63 with amyotrophic lateral sclerosis) were enrolled. All tasks except for vertical smooth pursuit had good intra-rater agreement. Gaze impersistence (K=0.68), upgaze restriction (K=0.66), horizontal smooth pursuit abnormalities (K=0.64), eyelid apraxia (K=0.65) and impaired Bell’s phenomenon (K=0.74) had good inter-rater agreement. Findings with less than 5% prevalence had poor agreement. Challenges identified by the raters included eyelids obscuring movements, lack of a standardized rating scale, and not being able to see the examiner.

Conclusion:
Experts reviewing videos of an ocular motility examination protocol agree regarding findings of gaze impersistence, upgaze limitations, horizontal smooth pursuit abnormalities, eyelid apraxia and impaired Bell’s phenomenon. This methodology is applicable to studies of the prevalence of these findings in populations for which quantitative eye movement recordings are prohibitive. Alterations in the exam protocol may improve expert agreement and expand the applications of this methodology. Limitations of this study include lack of comparison to a gold standard and rating by only two experts.

References: None

Key Words: Clinical Examination, Inter-Rater Reliability, Supra-Nuclear Ocular Motility

Financial Disclosure: None
Intracranial Arachnoid Cyst Causing Papilledema

Michael Vaphiades, Shannon Cox
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Introduction:
Papilledema is a term reserved for those patients who have optic disc edema caused by increased intracranial pressure. Arachnoid cysts are rare accounting for less than one percent of all intracranial lesions and are a very rare cause of papilledema. We present a patient with increased intracranial pressure from a cerebellar arachnoid cyst.

Methods:
A 43 year old Indian woman presented with a two month history of transient visual obscurations in both eyes. Her symptoms were worse with bending over, associated with headache and pulsatile tinnitus. She travelled to India frequently but did not report any history or symptoms of parasitic disease. Examination showed a slightly obese woman (5'4", 200 lbs), vision was 20/25 OU, color vision was 9/10 plates OU, confrontational fields were full, automated fields were mildly constricted OU. Ocular motility was normal. She had moderate papilledema in both eyes. The patient was prescribed acetazolamide 250 mg three times / day. Cranial and orbital MRI demonstrated a 4.5 by 4 centimeter right sided non-enhancing cerebellar cyst. There was mild displacement of the fourth ventricle. An empty sella and mild flattening of the posterior sclera indicated increased intracranial pressure. An ultrasound of the abdomen was normal. The patient was referred to neurosurgery and a right retromastoid craniectomy with resection of the cyst, intraoperative placement of a right parietal ventriculostomy and cranioplasty was performed.

Results:
Pathologic examination revealed the cyst to be a benign arachnoid cyst. After surgery the patient’s papilledema improved and the acetazolamide was tapered.

Conclusion:
There have been only five reported cases of posterior fossa arachnoid cysts in adults and only one was associated with papilledema and it was due to obstructing hydrocephalus. Papilledema is an unusual symptom of an arachnoid cyst and prompt surgical therapy is necessary to preserve vision.

References:

Key Words: Arachnoid Cyst, Papilledema, Posterior Fossa, Cerebellum, Increased Intracranial Pressure

Financial Disclosure: None
Zinc Deficiency Optic Neuropathy

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Introduction:
Eye tissues are rich in zinc (Zn) which converts vitamin A to retinol involved in the formation of cones.¹ Zn chelation occurs as an uncommon side effect of cimetidine, but more often seen in elderly with hepatic or renal dysfunction. Five patients were previously reported to have cimetidine-related optic neuropathy.² We present 3 patients who took cimetidine and developed Zn deficiency optic neuropathies.

Methods:
Case series

Results:

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<th>Cimetidine 800 mg/day for duodenal ulcers/Treatment duration</th>
<th>Initial visual acuity</th>
<th>Humphrey 30-2 visual fields</th>
<th>Fundus findings</th>
<th>Initial serum Zn levels (60-120 ug/dL normal)</th>
<th>MRI brain/orbits</th>
<th>CSF analysis</th>
<th>Final visual acuity 3 mos later</th>
<th>Serum Zn levels (ug/dL) after drug withdrawal</th>
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<td>82yo Male</td>
<td>8 days</td>
<td>20/40 OU</td>
<td>Paracentral scotoma OD Central scotoma OS</td>
<td>Optic disc edema OU</td>
<td>54</td>
<td>Pt refused MRI's</td>
<td>Pt refused LP</td>
<td>20/20 OD 20/30 OS</td>
<td>91</td>
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<tr>
<td>64yo Male</td>
<td>36 days</td>
<td>20/80 OD 20/100 OS</td>
<td>Central scotomas OD Diffuse depression OS</td>
<td>Mild optic disc pallor OU</td>
<td>37</td>
<td>normal</td>
<td>CSF normal; MS panel normal</td>
<td>20/50 OD 20/100 OS</td>
<td>58</td>
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<tr>
<td>67yo Male</td>
<td>17 days</td>
<td>20/80 OD 20/60 OS</td>
<td>Central scotomas OU</td>
<td>Optic disc edema OU</td>
<td>40</td>
<td>normal</td>
<td>CSF normal; MS panel normal</td>
<td>20/50 OD 20/40 OS</td>
<td>62</td>
</tr>
</tbody>
</table>

Other common nutritional, toxic, infectious, autoimmune, and hereditary optic neuropathies, such as B12, copper, heavy metals, LHON, MS, etc., were ruled out. Hemogram and metabolic panel were normal.

Conclusion:
Our patients had bilateral optic neuropathy associated with low serum Zn levels. After cimetidine withdrawal and Zn supplementation, some visual improvement occurred. It is unclear whether our patients had a pre-existing Zn deficiency worsened by the intake of cimetidine. This report provides evidence that the Zn chelating property of cimetidine can contribute to Zn deficiency optic neuropathy.

References:

Key Words: Zinc Deficiency, Optic Neuropathy, Cimetidine

Financial Disclosure: None
A Case of Bilateral Orbital Metastases from Prostate Carcinoma

Elena Geraymovych, Vivian Rismondo-Stankovich

1University of Maryland Medical Center, Baltimore, MD, United States, 2Greater Baltimore Medical Center, Baltimore, MD, United States

Introduction:
Orbital metastases are very uncommon and present in about 5% of all patients with systemic metastatic cancer. Prostate cancer is the most common cancer in men in the U.S. It accounts for approximately 10% of all orbital metastases. Radiographically, prostate cancer has a high tendency to metastasize to the bone, and in the orbit it is commonly located in the sphenoid bone as an osteoblastic lesion. It is very unlikely for metastatic prostate cancer to present with simultaneous bilateral orbital metastases. We present a case of bilateral orbital metastases in an unusually young patient with a known diagnosis of prostate cancer that resulted in bilateral decreased vision, proptosis, and EOM limitations.

Methods:
Case presentation and review of relevant literature.

Results:
A 55-year-old male presented with new onset of diplopia and blurred vision in both eyes. He was diagnosed with prostate cancer in 2008 and was undergoing chemotherapy treatment. On examination, his best-corrected vision OD 20/40 and 20/50 OS, no APD, limited abduction on both sides, bilateral proptosis, decreased color and stereo vision, and bilateral swollen optic nerves. CT demonstrated periosteal reaction and enhancing soft tissue component extending into the posterior lateral aspect of the left and right orbits. Left and right sphenoid lesions extended into the posterolateral aspect of the orbits and approached the orbital apex. Right sphenoid lesion was smaller than the left. The optic canals appeared patent. No biopsy of the lesions was done given known diagnosis and patient’s medically unstable condition.

Conclusion:
Bilateral orbital metastases from prostate cancer are extremely rare with only a few reports in the literature. Bilateral metastatic orbital involvement should be considered in a patient with new onset ophthalmic symptoms and metastatic prostate cancer.

References:

Key Words: Orbit, Prostate Cancer, Bilateral Metastases

Financial Disclosure: None
Introduction:
Traumatic optic neuropathy (TON) is an acute injury to the optic nerve often following blunt head trauma to the orbital rim. It is presumed to be a consequence of shearing forces transmitted to the orbital apex through the bones of the orbit. Initially, based on the National Acute Spinal Cord Injury Study 2 (NASCIS 2) corticosteroid treatment for TON became widely accepted even in the absence of strong evidence supporting its use. Later, the International Optic Nerve Trauma Study (IONTS) showed no difference in outcome between the treatment groups. There have been no subsequent clinical studies of corticosteroid treatment for TON. Additionally, since 2000, several animal studies have suggested that high-dose methylprednisolone may exacerbate apoptosis.

Methods:
Retrospective chart review of TON cases from the University of Maryland Ophthalmology Consult Service logbooks from 1999 to 2009.

Results:
A total of 117 patients were diagnosed with TON by the University of Maryland Ophthalmology Consult Service over the course of 10 years, with an average of 15 patients diagnosed per year. As expected, TON was most frequently caused by motor vehicle collisions and assaults and was more common in male patients. The percentage of TON patients receiving steroid treatment drastically decreased over the study period. A poor visual outcome (Snellen visual acuity < 20/800) was more common in patients who did not receive steroid treatment when compared to patients who had received steroids.

Conclusion:
Systemic corticosteroid treatment for TON has dramatically declined over the past 10 years, despite an absence of new clinical data. A retrospective analysis of our cases suggested a possible positive effect of corticosteroids on visual outcome. The consistent number of cases of TON identified at our center every year suggests that a prospective treatment trial might be feasible.

References:

Key Words: Traumatic Optic Neuropathy, Corticosteroids, Visual Acuity

Financial Disclosure: None
Poster 15

MRI Restricted Diffusion in Lymphomatous Optic Neuropathy

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Introduction:
Diffusion-weighted imaging (DWI) has been largely applied to the diagnosis of acute cerebral infarction, but restricted diffusion has been reported in brain parenchymal lymphoma1 and attributed to the dense cellularity of this tumor. Although restricted diffusion has been reported in ischemic and inflammatory optic neuropathy, it has not previously been reported in lymphomatous optic neuropathy. We describe such a case.

Methods:
A single case report and review of literature

Results:
A 44-year-old man with a recent diagnosis of non-Hodgkin B cell lymphoma experienced sudden painful loss of vision in the left eye. There was no light perception in the affected eye which had an ipsilateral afferent pupillary defect and a minimally swollen optic disc. Neurologic examination showed no other abnormalities. Magnetic resonance imaging (MRI) showed no evidence of brain parenchymal or meningeal lymphoma, but showed restricted diffusion in the intraorbital portion of the affected optic nerve, suggesting lymphomatous infiltration of the nerve. Cerebrospinal fluid flow cytometry and cytology were positive for a mature B cell neoplasm. Review of an earlier abdominal biopsy yielded a diagnosis of Burkitt lymphoma. Despite treatment with corticosteroid, standard chemotherapy, and orbital x-irradiation, visual function did not improve. Restricted diffusion persisted on MRI 4 months later.

Conclusion:
We report for the first time restricted diffusion in the affected optic nerve in a case of lymphomatous optic neuropathy. We presume that this phenomenon resulted from superimposed infarction or dense cellularity of the tumor. Restricted diffusion has been documented in 9 case reports2-10 involving either ischemic optic neuropathy (8 cases) or inflammatory optic neuropathy (1 case). Where follow-up DWI has been reported (2 cases), diffusion has reverted to normal. The persistence of restricted diffusion after 4 months in our patient is unexplained.

References:

Key Words: Lymphomatous Optic Neuropathy, Restricted Diffusion In Optic Nerve, Central Nervous System Lymphoma

Financial Disclosure: None
Bilateral Idiopathic Iris Atrophy

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Introduction:
We report 2 patients who developed bilateral symmetric, non-reactive, mydriatic pupils with 360 degrees of iris transillumination defects after respiratory illnesses and subsequent antibiotic use. Both patients had pupils that were non-reactive to light or accommodation with no response when challenged with 2% Pilocarpine, suggesting iris sphincter end-organ damage. Both patients were female, in their late 50’s to early 60’s, and had been treated with the oral fluoroquinolone, Moxifloxacin, for pneumonia several months prior to developing symptoms. Neither patient had any prior history of herpetic eye disease, previous trauma, heterochromia, glaucoma, or ocular inflammation.

Methods:
Extensive PubMed English language literature search.

Results:
No reported cases in the literature of antibiotic use leading to iris end-organ damage, or post-infectious causes.

Conclusion:
We conclude that either a new presentation of a post-infectious syndrome or a side-effect of antibiotic use resulted in bilateral symmetric iris sphincter damage in two patients.

References:

Key Words: Drug, Toxicity, Iris, Sphincter, Damage

Financial Disclosure: None
Poster 17

**Time-Domain and Spectral Domain Optical Coherence Tomography of Retinal Nerve Layer Thickness in MS Patients and Healthy Controls**

Alex Lange¹, Reza Sadjadi¹, Jameelah Saeedeh¹, Janette Lindley¹, Fiona Costello², Anthony Traboulsee¹

¹University of British Columbia, Vancouver, BC, Canada, ²University of Calgary, Calgary, AB, Canada

**Introduction:**
Optical Coherence Tomography (OCT) is a non-invasive technology for measuring Retinal Nerve Fiber Layer Thickness (RNFL). RNFL was shown to correlate to clinical findings and is a possible surrogate marker for brain atrophy in MS patients. Since recently, a new generation called Spectral Domain optical coherence tomography (SD OCT) is available and is hoped to be superior to older generation technology (TD OCT). However, the new generation OCT needs to be evaluated before it can be reliably used as standard of care. The aim of this study was to compare the newer generation OCT to the older generation in MS patients and healthy controls, to compare RNFL in MS eyes with optic neuritis, without optic neuritis and healthy controls, and to find correlation between RFNL and disease duration and refraction.

**Methods:**
In a cross-sectional study, eyes of 28 MS patients and 35 healthy controls underwent both TD- and SD-OCT measurements. RFNL measurements were correlated between the two machines, between MS eyes with and without optic neuritis and healthy controls, and disease duration and refraction.

**Results:**
A strong correlation (Pearson r=0.921, p<0.001), but a statistically significant difference of 2 μm (p<0.001) was shown between the two machines. RNFL was significantly different between MS eyes with history of optic neuritis (mean RFNL (SD) 72.21 μm (15.83 μm), MS eyes without history of optic neuritis 93.03 μm (14.25 μm) and healthy controls 99.07 μm (7.23 μm) (P<0.001) measured by SD OCT.

**Conclusion:**
SD- and TD-OCT values are highly correlating between the two machines, but show differences in absolute values. The results between different machines are not interchangeable which should be taken into account comparing values between different studies.

**References:** None

**Key Words:** Spectral Domain Optical Coherence Tomography, OCT, Multiple Sclerosis, Spectralis, Retinal Nerve Fiber Layer

**Financial Disclosure:** None
Spectral Domain Optical Coherence Tomography in Patients with Neuromyelitis Optica

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Introduction:
Neuromyelitis optica (NMO) is a type of CNS demyelinating disease seen more commonly in Asian population. NMO is distinguished from other types of demyelinating diseases by its unique lesion distribution in optic nerve, optic chiasm and spinal cord. NMO is often associated with systemic autoimmunity and the presence of NMO antibodies. The main aim of this study is to investigate factors contributing to Retinal Nerve Fiber Layer (RNFL) loss measured by Spectral Domain Optical Coherence Tomography (SD-OCT).

Methods:
A prospective cross-sectional study was performed in our NMO population (n=27). All patients underwent neurological examination including EDSS and had SD-OCT measurements of their RNFL and macular volume at the time of assessment.

Results:
RNFL in NMO eyes without history of optic neuritis (96.78 μm, SD 11.45) was significantly different from RNFL in eyes with history of optic neuritis (mean 64.81 μm, SD 19.52). Moreover, there was a significant difference between RNFL values in NMO eyes compared to MS eyes and healthy controls from other studies. NMO antibodies, and brain and spinal MRI findings did not significantly contribute to RNFL loss in either groups.

Conclusion:
OCT measures may be more reliable than NMO antibodies and other imaging findings as an outcome measure in NMO.

References:
1. Paratt, Prineas, Neuromyelitis optica: a demyelinating disease characterized by acute destruction and regeneration of perivascular astrocytes. Multiple Sclerosis, 16(10) 1156-1172, 2010

Key Words: OCT, NMO, RNFL, MS, Spectral Domain Optical Coherence Tomography

Financial Disclosure: None
Poster 19

Blink Rates, Ocular Surface Disease, Corneal Sensitivity, and Corneal Nerve Density in Progressive Supranuclear Palsy Patients and Parkinson's Patients

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Introduction:
Progressive supranuclear palsy (PSP) and Parkinson's disease have both motor and non-motor features. PSP and Parkinson's patients are frequently diagnosed with dry eye syndrome, but these patients rarely complain of dry eye symptoms. The aim of this study was to determine the relationships between blink rates, asymptomatic dry eye, and corneal sensitivity in PSP and Parkinson's patients, and compare these findings to those of normal control eyes.

Methods:
In this cross-sectional clinical study, 14 eyes of 7 patients with PSP, and 8 eyes of 4 patients with Parkinson's disease were examined and compared to 8 normal eyes of 4 age-matched control subjects. Blink rate, Cochet-Bonnet esthesiometry, meibomian gland disease, tear meniscus height, tear film breakup time, fluorescein staining, rose Bengal staining, lissamine green staining, and corneal subbasal nerve density (measured by using confocal microscopy in vivo) were determined. Comparisons between variables were made by using generalized estimating equation models to account for possible correlation between fellow eyes of the same subject.

Results:
Overall, dry eye test abnormalities were significantly more common in both PSP and Parkinson's compared to controls. PSP and Parkinson's groups had lower blink rates (p< 0.001) and decreased corneal sensitivity compared to controls (p<0.001). Both PSP and Parkinson's groups had normal corneal subbasal nerve density compared to controls (p<0.58 and p= 0.06 respectively). Blink rates and corneal sensitivity were lower in the PSP group compared to the Parkinson’s group (p<0.001, p=0.05 respectively). There was no correlation between corneal sensitivity and subbasal nerve density (r= 0.21, p=0.93).

Conclusion:
PSP and Parkinson' patients manifest signs of dry yet remain asymptomatic. The lack of symptoms might be explained by decreased corneal sensitivity which is not related to loss of corneal nerves.

References: None

Key Words: Progressive Supranuclear Palsy, Blink Rates, Corneal Sensitivity, Corneal Nerve Density, Dry Eye

Financial Disclosure: None
Physician Witnessed Ophthalmic Artery Occlusion: Poor Visual Outcome Despite Optimum Intravenous And Intra-arterial Thrombolysis

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Introduction:
A 78-year-old woman who presented for routine visit developed acute painless loss of vision in OD while in the exam room. She was diagnosed with presumed central retinal artery occlusion (CRAO) and immediately sent to the hospital for treatment with recombinant tissue plasminogen activator (rtPA).

Methods:
Case Report

Results:
Patient's initial visual acuity was 20/40 and suddenly changed to counting fingers. There was no associated headache or eye pain and she was noted to have right relative afferent pupillary defect (RAPD). Dilated fundus exam was normal, no cherry red spot and no embolus noted within the retinal arterial vessels.

Shortly after digital massage and paracentesis the patient was transferred to a tertiary care center emergency room with the presumed diagnosis of right CRAO.

Intravenous rtPA was administered within 2 hours of onset of symptoms, followed by intra-arterial rtPA within 4.5 hours of vision loss. The angiogram revealed filling defect at the origin of the right ophthalmic artery and in the distal right ophthalmic artery with poor filling of the distal right ophthalmic artery branches. There was no improvement of her visual acuity despite post thrombolysis angiographic evidence of normal filling of the ophthalmic artery and improved filling of its distal branches. The final visual acuity was light perception.

Conclusion:
Although controversy remains in regards to the treatment of choice in CRAO, there is consensus about the impact of ischemia duration on the visual outcome.1-5 We present a case of witnessed sudden painless vision loss with resultant light perception despite treatment with paracentesis followed by intravenous and intra-arterial rtPA within optimum time. The present case suggests that, in addition to duration of ischemia, the site of occlusion within the ophthalmic artery territory can be an even stronger determinant for visual outcome with more proximal occlusion compromising direct and collateral flow concomitantly.

References:

Key Words: None

Financial Disclosure: None
Poster 21

The Usefulness of Spectral Domain Optic Coherence Tomography in Diagnosing Foveal Hypoplasia in Suspected Ocular Albinism

Alaa Al-Ali, Raed Behbehani

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Introduction:
To demonstrate the value of spectral domain optical coherence tomography (SD-OCT) in diagnosing foveal hypoplasia in a subject with poor vision, nystagmus and clinical features oculocutaneous albinism.

Methods:
Case report and literature review.

Results:
A 24-year-old man presented with a history of nystagmus, poor vision since childhood, amblyopia, and strabismus surgery. Family history was positive for nystagmus. His best-corrected visual acuity was 20/80 OD and 20/70 OS. Refraction was +5.50 +1.75 x 110 OD and +4.50 +2.00 x 80 OS. Pupils were briskly reactive OU with no afferent pupillary defect. He had horizontal jerk nystagmus that was equal in amplitude and similar in all gaze direction. Anterior segment examination showed iris transillumination defects and fundus examination showed retinal hypopigmentation and ill-defined foveal reflexes. Three dimensional SD- OCT showed absence of foveal depression, preservation of the inner and outer retinal layers (normally absent in the fovea), but diminished retinal nerve fiber layer. These features confirmed the diagnosis of foveal hypoplasia.

Conclusion:
OCT has emerged a non-invasive way to image retinal diseases with very high resolution. SD-OCT offers the advantage of faster image acquisition and higher sensitivity over conventional time-domain OCT. There are few reports describing the OCT features of foveal hypoplasia in the literature. [1-4] Foveal hypoplasia can be isolated, or associated with albinism and aniridia. The presence of nystagmus and poor fixation ability in patients with albinism occasionally makes it difficult to diagnose foveal hypoplasia in these patients using fundoscopy. Imaging with SD-OCT offsets some of the problems associated with time-domain OCT such as long scanning time and allows more precise identification of the foveal structure in patients with nystagmus associated with loss of vision caused by foveal hypoplasia.

References:

Key Words: Spectral domain optical coherence tomography, Foveal Hypoplasia, Ocular Albinism

Financial Disclosure: None
Choroidal and Optic Nerve Involvement in Erdheim-Chester Disease

Elvira Naftaliev, Igal Leibovitch, Michaela Goldstein, Anat Kesler

Tel Aviv Medical Center, Tel Aviv, Israel

Introduction:
Erdheim-Chester disease is a rare, systemic histiocytosis that involves multiple organ systems

Methods:
A 39-year old woman presented with diffuse skeletal pain, headaches, and bilateral optic disc swelling. A computerized tomography revealed bilateral enhancement of the optic nerves and lacrimal glands, with extensive soft tissue infiltration around the optic nerves. A Lumbar puncture revealed high pressure with normal content. A biopsy from the tibia and lacrimal gland confirmed the diagnosis of Erdheim-Chester disease (ECD), a rare type of multi-organ histiocytosis. Treatment with oral acetazolamide, prednisone and immunosuppressive drugs resulted in improvement in systemic symptoms and optic disc swelling.

Results:
Two years later, while still on immunosuppressive drugs and with no ocular symptoms, optic nerve swelling was noted on a routine follow-up visit. Retinal examination revealed a new finding of bilateral large yellow-white choroidal lesion occupying the entire area between the disc and the macula with overlying subretinal fluid, the choroidal mass probably represents infiltrative lesions of ECD.

Conclusion:
Although ECD is known to involve different parts of the orbit and the periocular area, intraocular involvement is extremely rare, and to the best of our knowledge was reported in the literature only once.

References: None

Key Words: Histiocytosis, Optic Nerve, Choroid

Financial Disclosure: None
Poster 23

Orbital Rosai-Dorfman Disease: Case Report and Diagnostic Challenges

Ashref AlMoosa¹, Raed Behbehani¹, Abdulmohsen Hussain², Abdullah Ali³

¹Al-Bahar Eye Center, Kuwait, Kuwait, ²ENT dept, Zain and Sabah Hospital, Kuwait, Kuwait, ³Hussain Maki Al-Jomaa Center for special surgeries, Kuwait, Kuwait

Introduction:
Rosai Dorfman disease is characterized by histiocytic proliferation and massive cervical lymphadenopathy. About 40% of patients have extra-nodal involvement. Histopathological and immunohistochemical confirmation are essential in establishing the diagnosis. We report a case of orbital Rosai-Dorfman in a six year old child.

Methods:
We herein report a challenging case of extra-nodal Rosai Dorfman disease in a six year old child, initially misdiagnosed as orbital inflammatory disease after an initial orbital biopsy.

Results:
Our patient presented with a picture resembling orbital inflammatory disease which led to diagnostic confusion, even after radiographic imaging. Treatment with steroids for a prolonged period of time was not effective. Due to absence of follow-up for a considerable amount of time the inflammatory lesion had extended into adjacent tissues and intra-cranially making complete surgical excision impossible.

Conclusion:
The role of repeated biopsy if necessary in difficult clinical situations cannot be overemphasized.

References: None

Key Words: Orbit, Rosai-Dorfman, MRI

Financial Disclosure: None
Feasibility and Quality of Non-Mydriatic Fundus Photographs in Children

Daniela Toffoli, Beau B. Bruce, Cédric Lamirel, Amanda Henderson, Nancy J. Newman, Valérie Biousse

Emory University School of Medicine, Atlanta, Georgia, United States

Introduction:
Funduscopic examination is difficult in young children and almost never attempted by non-ophthalmologists. The FOTO-ED Study has shown that non-mydriatic fundus photographs might replace funduscopic examination in an adult Emergency Department (ED). Our objective was to determine if high-quality non-mydriatic fundus photographs can be obtained in children.

Methods:
Non-mydriatic fundus photographs were obtained by a fellow and medical student on children in a pediatric ophthalmology clinic. Photographers received <30 minutes of training. Photographs were taken of both eyes in dim light. Ease of fundus photography was recorded on a 10 point Likert scale. Photographs were reviewed by a neuro-ophthalmologist and quality was graded from 1 to 5 (1: inadequate for any diagnostic purpose, 2: unable to exclude all emergent findings, 3: only able to exclude emergent findings, 4: not ideal but still able to exclude subtle findings, 5: ideal quality). The primary outcome measure was image quality by age.

Results:
We included 878 photographs of 212 children (median age 6 years [1-18 years]). We obtained photographs of at least one eye in 190/212 (89.6%) and both eyes in 181/212 (85.3%). Median rating for ease of photography was 7/10 (10=very easy). Photographs of some clinical value were obtained in 67% of children <3 years and 95% ≥3 years. High-quality photographs (grade 4 or 5) were obtained in both eyes in 7% of children <3 years; 57% of children ≥3 to <7 years, 85% of children ≥7 to <9 years, and 65% of children ≥9 years. The youngest patient with high-quality photographs in both eyes was 22 months.

Conclusion:
Non-mydriatic fundus photographs of similar quality to adult patients can easily be obtained in children over the age of 3, and in some children as young as 22 months. These results suggest the application of the FOTO-ED study to pediatric EDs should be explored further.

References:

Key Words: Funduscopic Examination in Children, Non-Mydriatic Fundus Photography

Financial Disclosure: None
Poster 25

A Comparison of Saccade Metrics with other Clinical Neurophysiological Tests

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Introduction:
Of all motor systems, the oculomotor system is the best understood and can be most accurately measured. Oculomotor studies should therefore lend themselves to the understanding of the pathophysiology of human movement disorders and, ideally, to the diagnosis of these conditions. Sadly, despite the fact that many studies have looked at the way(s) in which saccade metrics (e.g. latency, gain, peak velocity) differ from normal in these diseases, oculomotor recording has not become a widespread clinical tool.

A major reason for this is that within- and between-subject variance in normal saccade metrics is large so any disease-related departure from "normal" cannot be accurately detected unless the result is very abnormal. Accordingly, most studies looking at diseases have had to use groups of patients to demonstrate significant disease-related effects. Many authors have suggested that oculomotor recording might be useful for clinical diagnosis, but in practice this has never eventuated.

The aim of this study was to search for possible correlation(s) between saccade metrics and various standard clinical neurophysiological tests to try to control for between-subject variance.

Methods:
Thirty normal volunteers (aged 43.6 ± 11.6 years) underwent visual evoked responses (VER), nerve conduction studies (NCS) and recording of their reflexive saccadic eye movements. In addition, hand grip strength measurement and body composition analysis (BCA) were performed.

Results:
No significant correlation could be found between age, VER NCS, or hand grip strength and saccade metrics. Interestingly, however, saccade latency and peak velocity were significantly correlated with reactance as determined by the BCA.

Conclusion:
Correlation with standard clinical neurophysiological parameters does not provide a means of reducing inter-subject variance of reflexive saccadic eye movement. The potential significance of reactance and possible other strategies for reducing variance will be discussed.

References: None

Key Words: Saccades, Visual Evoked Responses, Nerve Conduction Studies, Saccadic Metrics

Financial Disclosure: None
Poster 26

Normal Exophthalmometry Measurements in a United States Pediatric Population

Johanna Dijkstal, Erick Bothun, Andrew Harrison, Michael Lee

University of Minnesota, Minneapolis, United States

Introduction:
Exophthalmometry in children requires reference data for all ages. We sought to determine the normal distribution of exophthalmometric measurements in a U.S. pediatric population as a reference for clinical practice.

Methods:
This was a cross-sectional study conducted with a Hertel Exophthalmometer to measure normal subjects between 1-17 years of age, randomly selected from patients presenting to our institution. Normal volunteers also participated at a booth at the annual state fair. Subjects were excluded for a prior history of orbital tumor, craniofacial anomaly, thyroid disease, orbital trauma, or inability to tolerate the measurement.

Results:
There were a total of 673 subjects (52% female) with a mean age of 9.6 years. The data is presented in Table 1. Mean exophthalmometry for African American subjects was significantly greater than all other ethnicities (p<0.01). Table 2 shows an increase in exophthalmometry with increasing age. There was no difference in exophthalmometry between male and female subjects. Asymmetric measurements were seen in 100 (14.9%) subjects, with a 2 mm maximal difference in 2 subjects.

Table 1:

<table>
<thead>
<tr>
<th>Race</th>
<th>N</th>
<th>Mean Exophthalmometry (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>All Subjects</td>
<td>673</td>
<td>15.1 (11.5 - 18.9)</td>
</tr>
<tr>
<td>Caucasian</td>
<td>595</td>
<td>15.0 (11.4 - 18.8)</td>
</tr>
<tr>
<td>Asian</td>
<td>27</td>
<td>14.2 (10.8 - 18.2)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>31</td>
<td>15.4 (11.9 - 18.9)</td>
</tr>
<tr>
<td>African American</td>
<td>16</td>
<td>16.8 (13.0 - 20.7)</td>
</tr>
</tbody>
</table>

Table 2:

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>N</th>
<th>Mean Exophthalmometry (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-4</td>
<td>53</td>
<td>13.4 (10.1 - 16.6)</td>
</tr>
<tr>
<td>5-8</td>
<td>166</td>
<td>14.5 (11.3 - 17.6)</td>
</tr>
<tr>
<td>9-12</td>
<td>164</td>
<td>15.3 (11.9 - 18.7)</td>
</tr>
<tr>
<td>13-17</td>
<td>181</td>
<td>16.2 (12.8 - 19.7)</td>
</tr>
</tbody>
</table>

Conclusion:
Exophthalmometry varies with age among the pediatric population. We present reference data for each age group in a U.S. cohort. Similar to adults, African Americans tended to have significantly higher Hertel measurements.

References: None

Key Words: Exophthalmometry, Exophthalmos, Pediatric, Children, Hertel

Financial Disclosure: None
Poster 27

Comparison of Magnocellular Ganglion Cell Function Between Collegiate Athletes and Nonathletes

Khizer Khaderi

Doheny Eye Institute, Los Angeles, CA, United States

Introduction:
To determine whether the magnocellular pathway is heightened in athletes compared to the general population.

Methods:
The frequency doubling technology (FDT) instrument, designed to stimulate M cell neurons, which are involved in motion detection was used to test participants. Using this instrument, while wearing trial frames with neutral density filters, the thresholds of M cell neuron activity were examined. While participants concentrated on a fixation point, sinusoidal, flickering bar gratings were presented in the periphery, superiorly and inferiorly. Participants identified and responded to the stimulus by pressing a hand-held button.

Results:
Results of FDT testing of participants wearing 0.3 log unit neutral density filters, was statistically significant by t-test analysis ($P<0.05$). The difference between the mean scores of 13.71 for athletes (N=14) and 3.71 for nonathletes (N=17) was 10.00 (LSD 8.67). No statistical significance was appreciated for the participants on FDT testing while wearing 0.6 log unit neutral density filters ($P=0.16$). The difference in mean scores for athletes (16.57) and nonathletes (7.24) was 9.33 (LSD 13.44).

Conclusion:
Nonathletes performed better than athletes at the FDT 0.3 filter level, an unexpected result. Since nonathletes had superior performance with the FDT at the 0.3 filter level, other factors that could contribute to this result were investigated. Through ANOVA and ANCOVA testing, other factors that directly influenced the hypothesis were found to be statistically significant. In regard to M-cell regulation, further research is needed to determine the full-effect of heightened M-cell function.

References: None

Key Words: Magnocellular Cell Neuron, Frequency Doubling Technology, Sporting Activity, Video Games, Ocular Dominance

Financial Disclosure: Author has patents related to research presented and is a founding member in a company related to the presented research. Financial relationships that relate to the topic of this presentation:

•Khizer Khaderi MD, MPH, co-Founder of Vizzario Inc. and Vizzario Labs
•Registered Patents
  •US Pat. 11867477 - Filed Oct 4, 2007 - iSport, LLC
  •US Pat. 11867486 - Filed Oct 4, 2007 - iSport, LLC
Poster 28

Anti-Suppression Binocular Treatment of Amblyopia

Behzad Mansouri, Benjamin Thompson, Robert Hess

1Section of Neurology, Department of Internal Medicine, University of Manitoba, Winnipeg, Manitoba, Canada, 2Department of Optometry and Vision Science, University of Auckland, Auckland, New Zealand, 3McGill Vision Research, Department of Ophthalmology, McGill University, Montreal, Quebec, Canada

Introduction:
Amblyopia is the most common cause of monocular impairment worldwide. The present treatments for amblyopia are monocular aiming to improve the vision in the amblyopic eye (AME) through either patching of the fellow fixing eye (FFE) or visual training of the AME. This approach is problematic, not least of which because it seldom results in establishment of binocular function. Previously we have shown that amblyopes possess binocular cortical mechanisms for both threshold1 and suprathreshold stimuli2. We have also outlined3 a novel procedure for measuring the extent, to which the FFE suppresses the AME, rendering what is a structurally binocular system, functionally monocular. Our results show that prolonged periods of viewing (under the artificial viewing conditions in which stimuli of different contrast are viewed by each eye) during which information from the two eyes is combined leads to a strengthening of binocular vision in amblyopes.

Methods:
We used a random-dot-kinematogram paradigm with a signal-to- noise measure of performance for images of different contrasts presented separately to each eye. We measured the degree of suppression by finding the contrast ratio that leads to equal performances of the two eyes (the balance point)3. Patients were repeatedly tested at this balance point. At the end of each block of testing; visual acuity, stereovision and binocular vision were tested.

Results:
Exposure to binocular combination significantly improved binocular vision (p=0.003), stereopsis (p=0.01) and acuity in the AME (p<0.0001).

Conclusion:
Concomitant improvement in monocular acuity of the AME occurs with a reduction in suppression and strengthening of binocular fusion. Furthermore, in a majority of patients tested, stereoscopic function was established for the first time. This provides the basis for a new treatment of amblyopia, one that is purely binocular and aimed at reducing suppression as a first step. Furthermore it is effective for adults well past the critical period.

References:
1. Baker et al, 2007 IOVS 48, 5332-5338
2. Baker et al, 2008 Vision Res. 48, 1625-1640
3. Mansouri et al 2008 Vision Res. 48, 2775-2784

Key Words: Amblyopia, Suppression, Binocular, Treatment, Perceptual Training

Financial Disclosure: Provisional patent: Binocular Vision Assessment or Therapy (registration number 37796/23-10-2007).
Optic Nerve Head Drusen (ONHD) in Black Patients

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Introduction:
Several studies have suggested racial differences in the prevalence of ONHD. 2% of autopsied American patients (of unknown race) had ONHD [1], whereas 0.2% of the Chinese general population [2] had ONHD on fundus photography. A study from Miami (where the percentage of non-white patients is high) found that only 2/98 patients (2%) with exposed ONHD were black [3]. The goal of our study was to determine the prevalence of black patients among ONHD patients presenting to a large academic center that sees a similar proportion of white and black patients.

Methods:
Retrospective chart review of consecutive patients with ONHD on examination or B-scan ultrasonography, seen between 1989 and 2010. Race was assessed based on patient appearance or self-report. All patients had a standardized neuro-ophthalmic assessment, including perimetry and fundus photography. Wherever possible, relatives were examined.

Results:
Of 196 patients with ONHD reviewed, 10 were black (5.1%) (7 women; age 8-61 yrs, mean 25 yrs, median 15 yrs). None were related and none of their examined family members had ONHD on fundus examination. 6/10 patients had bilateral ONHD. Most (7/10) had no visual symptoms. Visual acuity was normal in most eyes with ONHD (range -0.1 to 0.4, mean 0.08, median 0.0; logMAR). 15/16 eyes with ONHD had small cupless optic discs, 13/16 had elevated ONH, and 5/16 had exposed drusen. Perimetry was normal in 4/16 eyes, showed an enlarged blind spot in 5/16, a constricted field in 5/16, a nasal defect in 2/16, a central defect in 1/16, and generalized depression in 1/16 eyes.

Conclusion:
ONHD are rare in black patients, possibly due to the presence of a larger cup-to-disc ratio [4] and a lack of predisposing genetic factors. The disproportion is not attributable to the patient population seen at our institution, where approximately 50% of patients examined are black.

References:

Key Words: Optic Nerve, Drusen, Black

Financial Disclosure: None
Binocular Summation of High- And Low-Contrast Visual Acuities in Multiple Sclerosis Patients Compared with Normal Humans

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Introduction:
High- and low-contrast visual acuities (VA) are higher with binocular than with monocular viewing in normal humans due to binocular summation [1,2]. Low-contrast vision is often abnormal in multiple sclerosis (MS) patients, even when high-contrast VA is normal [3]. However, it is unclear if, and to what extent, binocular viewing might alter high- and low-contrast VA in MS patients compared to normal humans. We aimed to study high- and low-contrast VA with binocular versus monocular viewing in MS patients compared to normal humans.

Methods:
We studied 26 patients with clinically-definite MS (24 male, 2 female; age 38-69 yrs; with or without prior optic neuritis) and 15 normal humans (12 male, 3 female; age 30-71 yrs). All had measurements of monocular and binocular high-contrast (100%, ETDRS) and low-contrast VA (25%, 10%, 5%, 2.5% and 1.25%, Sloan charts), and stereopsis (Randot). Binocular ratio (BR) was calculated as the ratio of binocular VA to best monocular VA at each contrast level.

Results:
Binocular VA was better than best monocular VA at all contrast levels in the normal humans (p<0.05). Binocular VA was better than best monocular VA at 100%, 10%, 2.5%, and 1.25% contrast levels in the MS patients (p<0.05). The difference between the monocular VA of the two eyes was greater in the MS patients than the normal humans at all contrast levels (p<0.05). In several MS patients, however, VA improved with binocular viewing despite poor VA in one eye or poor stereopsis. BR was higher in the normal humans than the MS patients at all contrast levels except 100% (p<0.05).

Conclusion:
VA is better with binocular than with monocular viewing in normal humans and MS patients. The improvement in low-contrast VA with binocular viewing is less marked in MS patients, possibly due to a larger discrepancy in low-contrast VA between the eyes [4].

References:

Key Words: Multiple Sclerosis, Optic Neuropathy, Contrast Vision, Binocular Summation

Financial Disclosure: None
Risk of Stroke after Acute Ischemic Retinal Events in China

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Introduction:
Risk of developing stroke after acute ischemic retinal events (AIRE) varies according to different diseases course and studies. To investigate risk of developing stroke in Chinese AIRE patients and the correlated risk factors.

Methods:
AIRE patients were prospectively collected. While clinical data was recorded, internal carotid artery(ICA) and cardiac ultrasound, blood test for hypercoagulation and vasculitis were tested. Patients were followed up for visual outcome and new cerebral or retinal stroke. Data was statistically analyzed to see the correlated high risk factors of stroke after AIRE.

Results:
In 102 of 128 AIRE cases (79.7%) which had 1-year follow-up, cerebral stroke was reported in 13 cases (12.7%, 95%CI, 6.2%-19.2%). The cumulative risk of cerebral stroke was 2.0% (95%CI, 0-4.7%) within 7 days, 3.9% (95%CI, 0.2%-7.6%) within 2 weeks, 7.8% (95%CI, 2.5%-13.1%) within 1 month, 9.8% (95%CI, 4.1%-15.5%) within 3 months and 10.8% (95%CI, 4.7%-16.9%) within 6 months since AIRE. In 35 cases of retinal transient ischemic attack (R-TIA), 17 cases (48.6%, 95%CI, 32.1%-65.1%) had retinal stroke and 5 cases (14.3% (95%CI, 2.7%-25.9%) had cerebral stroke during the follow-up. Severe carotid artery stenosis, past medical history of ischemic cardiac attack and cerebral stroke were found to be significantly correlated with higher risk of stroke after AIRE.

Conclusion:
While totally 12.7% of AIRE patients had cerebral stroke within 1 year, 9.8% happened with 3 months. Retinal stroke occurred to nearly a half of R-TIA patient. Severe carotid artery stenosis, history of ischemic cardiac attack and cerebral stroke were found to be the high risk factors for stroke in AIRE group.

References:

Key Words: Stroke, Ischemic, Retinal, Cerebral, Outcome

Financial Disclosure: None
Kjellin’s Syndrome: Three new cases

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Introduction:
Kjellin’s syndrome is a hereditary neuro-ophthalmological syndrome characterized by progressive and symmetric spastic paraparesis associated with amyotrophy, memory impairment, severe cognitive disturbances, spatiotemporal disorientation, praxic and gnostic disorders, and thinning of the corpus callosum at MRI. Confirmation is provided by genomic analysis (SPG11 or SPG15 genes for autosomal recessive forms).

Methods:
We report three new patients, a brother (02/11/67) and his sister (5/27/68) as well as an unrelated man (02/07/85) with spastic paraplegia of unknown genetic causes. An abnormal eye fundus prompted us to look for SPG11 mutation. Other complementary examinations (Photo, OCT, ERG, EOG, VEP, Colour vision,) were performed.

Results:
SPG11 mutations have been identified in all 3 cases. Eye fundus aspect is variable and depends on the evolution of the retinopathy. In the initial stages some bilateral and symmetric, macular or paramacular flecks characterize the disease. Some of these flecks may even encroach on the foveola. They will then increase in number and be associated with peripheral reticulations such as in pattern dystrophy. Photographs with filters or autofluorescence accentuate their features. OCT provides additional information on the position and depth of these flecks. Visual field is difficult to achieve because of cognitive impairment. A mild blue-yellow dyschromatopsia is found. ERG and EOG are not helpful.

Conclusion:
Eye fundus in spastic paraplegia patient is a good pathognomonic feature of spastic paraplegia SPG11.

References:
1. Kjellin Syndrome: Long-term Neuro-ophthalmologic Follow-up and Novel Mutations in the SPG11 Gene B. Puech; A. Lacour; G. Stevanin; B. Sautiere; D. Devos; Ch. Depienne; E. Denis; E. Mundwiller; D. Ferriby; P. Vermersch; S. Defoort-Dhellemmes. Ophthalmology article in press

Key Words: Kjellin’s Syndrome, SPG11, SPG15, Hereditary Spastic Paraplegia, Corpus Callosum

Financial Disclosure: None
A Rapid Screening Tool (King-Devick Test) for Sports-Related Concussion

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Introduction:
Sports-related concussion has received increasing attention as a cause of short- and long-term neurologic symptoms. A test of rapid number naming (King-Devick [K-D] test) requires seconds to complete and captures impairment of eye movements and other correlates of suboptimal brain function. In a recent study, the K-D test was shown to be a reliable method for identifying athletes with head trauma. We investigated the K-D test as a potential rapid visual screening tool for concussion in collegiate athletes.

Methods:
Athletes from the Penn varsity football, sprint football, and women’s and men’s soccer teams underwent K-D testing prior to the start of the 2010-11 playing season (baseline). The sprint football team had two baseline tests to evaluate for learning effects. For athletes who sustained head trauma during a game or practice, the K-D test was administered on the sidelines. K-D test scores (time in seconds) were correlated with Immediate Post-Concussion and Cognitive Testing (ImpACT).

Results:
Among 189 athletes tested at baseline, 103 were varsity football players, while 36 played sprint football. The women’s and men’s soccer teams each had 25 study participants. Minimal improvement of K-D scores was noted between the two baseline tests (40.8±6.9 vs. 36.7±6.3 seconds, p<0.0001, signed-rank test), reflecting learning effects. For the 4 athletes who developed concussions, K-D testing on the sidelines immediately after impact showed changes from baseline that were uniformly in direction of worsening (range 0.5 to 9.5 second increases, p=0.06 vs. baseline). These K-D scores correlated significantly with the ImpACT composite score for visual memory (r=0.99, p=0.004).

Conclusion:
The K-D test is a strong candidate rapid sideline visual screening tool for concussion, showing consistent worsening of scores following head trauma. Ongoing follow-up in this study with additional concussion events will further examine the effectiveness of the K-D test in collegiate athletes.

References:

Key Words: Vision, King-Devick Test, Eye Movements, Concussion, Sports

Financial Disclosure: Dr. Devick is an employee of King-Devick test, LLC.
How Similar are Ethambutol-Induced Optic Neuropathy and Leber's Hereditary Optic Neuropathy?

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Introduction:
It is known that toxic optic neuropathy and hereditary optic neuropathy have similar clinical characteristics. By comparing ethambutol-induced optic neuropathy (EON) and Leber's hereditary optic neuropathy (LHON), the representative diseases of each entity, respectively, we studied their similarities and differences.

Methods:
In this study we retrospectively examined medical records of patients diagnosed with the two diseases at the Neuro-Ophthalmology Department for 10 years, from January 2000 to September 2010.

Results:
The number of EON and LHON patients was 32 and 23 persons, respectively. LHON had more male patients at 83% of the total (EON 56%), and the mean age of occurrence (variance 9 - 48 yrs) was lower at 23.0 years (EON 58.3 yrs, variance 18 - 86 yrs). EON occurred in both eyes simultaneously (with the exception of 2 cases), but LHON occurred successively in each eye with an average of 40 days between occurrence in one eye and the other. There was no difference in visual acuity at first diagnosis, being hand motion to normal, but upon follow-up examinations, improvement in visual acuity was better in EON at 47%, compared to 17% in LHON. The most common visual field disorders, central or cecocentral scotoma, comprised 67% of EON and 87% of LHON. Five EON patients were tested for the LHON gene, but all appeared negative.

Conclusion:
The two diseases are similar in that they occur in both eyes and that disorder in visual acuity and visual field progress similarly, but they show many differences in detailed clinical characteristics such as gender of patients, successive occurrence in both eyes, age of first occurrence, recovery rate and pattern of visual field disorders. Therefore they should be considered as different diseases with distinct boundaries of its own.

References: None

Key Words: Ethambutol-Induced Optic Neuropathy, Leber's Hereditary Optic Neuropathy, Visual Loss, Visual Field Defect, Visual Recovery

Financial Disclosure: None
Enlarged Arterial Reflexes as a Possible Clinical Sign of Segmental Optic Nerve Hypoplasia

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Introduction:
Since the late 1970ies several case series of congenital segmental optic nerve hypoplasia were published, representing a developmental disorder of the nasal, the superonasal or the superior portion of the optic disc. The four characteristic clinical signs are eccentric emergence of retinal vessels, adjacent parapapillary scleral halo, pallor of the affected optic disc portion, and thinning of the corresponding retinal nerve fiber layer.
Our data suggest enlarged reflexes of the proximal retinal arteries as a possible additional clinical finding in segmental optic nerve hypoplasia.

Methods:
Description of three patients with segmental optic nerve hypoplasia and enlargement of the proximal arterial reflex within the area of the hypoplastic retinal nerve fiber layer.

Results:
Case 1: Superonasal optic nerve hypoplasia in the right eye explained the inferotemporal visual field defect. In addition to the known clinical signs of the disorder, the patient also exhibited a slightly enlarged arterial reflex along the proximal segment of the superonasal arterial branch.
Case 2: Incidental detection of an inferotemporal visual field defect in the left eye led to the diagnosis of superonasal optic nerve hypoplasia. The retinal arterial bifurcation within the area of retinal nerve fibre layer thinning showed an enlarged reflex.
Case 3: The phenomenon of arterial reflex enlargement was most prominent in this patient with bilateral superonasal optic nerve hypoplasia.

Conclusion:
In 1978 Frisen and Holmegaard already mentioned the appearance of broad reflexes along the major retinal vessels in optic nerve hypoplasia. They interpreted it as vascular indentations of the posterior vitreous or the internal limiting membrane due to retinal nerve fiber thinning. A developmental anomaly of the vessel adjacent to the hypoplastic optic nerve segment may be considered as well. Independent of the underlying cause, enlarged arterial reflexes might serve as an additional clinical sign of segmental optic nerve hypoplasia.

References:

Key Words: Segmental Optic Nerve Hypoplasia, Arterial Reflex

Financial Disclosure: None
The Effects of Anisometropic Amblyopia on Visually-Guided Reaching

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Introduction:
The effects of impaired spatiotemporal vision in amblyopia on visuomotor skills have rarely been explored in detail. The goal of this study was to examine the influences of amblyopia on visually-guided reaching.

Methods:
Eighteen patients with anisometropic amblyopia and 18 control subjects were recruited. Participants executed reach-to-touch movements toward targets presented randomly 5° or 10° to the left or right of central fixation in 3 viewing conditions: binocular, monocular amblyopic eye, and monocular fellow eye viewing (left and right monocular viewing for control subjects). Visual feedback of the target was removed on 50% of the trials at the initiation of reaching.

Results:
Reaching accuracy was comparable between patients and control subjects during all three viewing conditions. Patients’ reaching responses were slightly less precise during amblyopic eye viewing, but their precision was normal during binocular or fellow eye viewing. Reaching reaction time was not affected by amblyopia. The duration of the acceleration phase was longer in patients in comparison to control subjects under all viewing conditions, whereas the duration of the deceleration phase was unaffected. Peak acceleration and peak velocity were also reduced in patients.

Conclusion:
Amblyopia affects both the programming and execution of visually-guided reaching. The increased duration of the acceleration phase, as well as the reduced peak acceleration and peak velocity might reflect a strategy or adaptation of feedforward / feedback control of the visuomotor system to compensate for degraded spatiotemporal vision in amblyopia, allowing patients to optimize their reaching performance.

References: None

Key Words: Amblyopia, Binocular Vision, Reaching

Financial Disclosure: None
Clinical Neuro-Ophthalmic Findings in Familial Dysautonomia

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Introduction:
Familial dysautonomia (FD) or Riley Day Syndrome is a recessive hereditary sensory and autonomic neuropathy caused by mutations in the IKBKAP gene. Despite knowing the defective gene, the cause of the observed neuropathy remains unclear. While atrophic keratopathy is a well known complication, the neuro-ophthalmic features of FD have not been well described. We evaluated the neuro-ophthalmic features of patients with FD to better understand the visual phenotype and the cause of the vision loss in these patients.

Methods:
Sixteen patients with FD (n=32 eyes), ages 11-61 years, underwent thorough neuro-ophthalmic evaluation. Optical coherence tomography (OCT) was also done to analyse the retinal nerve fiber layer in 12 patients.

Results:
Visual acuity was affected in most cases (30 eyes ranging from 20/40-20/400). Mild corneal opacities were common but significant in only two eyes. All cases had red-green color vision impairment, and central and centrocentral defects on automated visual fields, even in those with normal visual acuity. Pupillary responses in all eyes were sluggish. Temporal optic nerve pallor was consistent and associated with retinal nerve fiber layer loss in the papillo-macular region. This was confirmed by optical coherence tomography. Relatively non-specific ocular motility abnormalities were also seen.

Conclusion:
The optic neuropathy in FD is characterized by predominant loss of the papillo-macular bundle. As this is similar to other hereditary optic neuropathies with abnormal mitochondrial DNA, a similar abnormality may occur in patients with FD.

References:

Key Words: Riley-Day Syndrome, Familial Dysautonomia, Optic Neuropathy, Optical Coherence Tomography, Central Scotoma

Financial Disclosure: None
Optic Nerve Biopsy in the Management of Progressive Optic Neuropathy

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Introduction:
Standard work-up for optic neuropathy can be challenging, at times yielding non-specific laboratory and imaging findings. In the setting of diagnostic uncertainty, optic nerve biopsy can be considered when empiric treatment risks outweigh benefits or progressive visual decline ensues despite therapy. We reviewed our long-term experience with optic nerve biopsy at a tertiary care institution, evaluating diagnostic and therapeutic utilities in a diverse set of clinical circumstances.

Methods:
Retrospective chart review. Optic nerve biopsies at the University of Pennsylvania from 1990–2010 were identified from hospital pathology databases and a survey of treating physicians. Medical records were reviewed.

Results:
Sixteen patients underwent optic nerve biopsies with a mean age of 48.6 +/- 15.4 (SD) years. At the time of biopsy, visual acuity was NLP in 7 (44%) of eyes, LP to CF in 6 (37%), and 20/400 or better in 3 (19%). Of the specimens sampled, 7 included full-thickness nerve, 4 contained only the dural sheath, and 5 were limited to adjacent masses adherent to the optic nerve. Diagnoses encountered included sarcoidosis (n=2), cavernous hemangioma (n=2), glioma (n=2), meningioma (n=2), and miscellaneous (n=8). In all but one case, the biopsy contributed to a clinical diagnosis and specific management. In 6 patients (37%), the fellow eye had experienced sequential but less profound vision loss prior to biopsy, prompting a tissue diagnosis. None of these patients had experienced further vision loss in the fellow eye at last follow-up.

Conclusion:
Optic nerve biopsy should be considered in the setting of progressive optic neuropathy when there is profound visual loss and when standard testing does not ascertain an etiology. Biopsy can be beneficial in substantiating the diagnosis and possibly halting vision loss in the affected or fellow eye. To our knowledge, this represents the first analysis of a large series of optic nerve biopsies.

References:

Key Words: Optic Nerve, Biopsy, Optic Neuropathy

Financial Disclosure: None
**Poster 39**

**Frequency of Involvement of the Different Extra-ocular Muscles in Myasthenia Gravis**

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**Introduction:**
Each extra-ocular muscle may be involved in myasthenia gravis masquerading any type of ocular motor pathology. The frequency of involvement of each muscle is not well established in the medical literature. This study was designed to determine if there is any tendency for a specific muscle or muscles combination to be predominantly affected.

**Methods:**
Retrospective chart review of myasthenic patients with extra-ocular muscles involvement. Included were 30 patients that had a clinical diagnosis of myasthenia gravis confirmed by at least one additional positive test: Tensilon test, acetylcholine receptor antibodies, thymoma on chest CT or suggestive EMG. Only patients in whom the neuro-ophthalmological examination documented involvement of a specific extra-ocular muscle (or muscles) were included.

**Results:**
Frequency of involvement of each muscle in this cohort was as follows: inferior oblique 63%, lateral rectus 30%, superior rectus 23%, inferior rectus 20%, medial rectus 13%, and the superior oblique was involved in 6% of the patients.

**Conclusion:**
The inferior oblique was the most frequently affected muscle. Diplopia caused by paresis this muscle is rarely encountered (other than as a part of oculomotor nerve palsy); hence, when a patient presents with vertical diplopia that results from an isolated inferior oblique palsy, myasthenic etiology should be highly suspected.

**References:**
None

**Key Words:** Myasthenia Gravis, Diplopia, Extraocular muscles

**Financial Disclosure:** None
Poster 40

Quetiapine Induced Idiopathic Intracranial Hypertension

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Introduction:
Quetiapine fumarate (Seroquel) is indicated for the treatment of acute manic disorder associated with bipolar disorder, and schizophrenia. It is an antagonist at multiple neurotransmitter receptors including serotonin, dopamine, histamine, and adrenergic. One side effect of Quetiapine is weight gain. Excessive weight gain is associated with idiopathic intracranial hypertension (IIH).

Methods:
A 15 year old girl presented with 8 weeks of progressive horizontal diplopia without headache. 5 months prior she had gained 40 pounds over 3 weeks after beginning Quetiapine XR 400mg/day. Other medications included Modafinil (Provigil), Fluoxetine (Prozac), and Lisdexamfetamine (Vyvanse). Her vision acuity was 20/20. There were mild bilateral abduction deficits and both optic nerves were swollen.

Results:
MRI / MRA of the brain were unremarkable. On lumbar puncture the opening pressure was 33cmH2O with normal protein, glucose, and cell count. She was begun on Acetazolamidine 500mg 3x per day. The Quetiapine, Modafinil and Lisdexamfetamine were discontinued. Ten weeks later she had lost 14 pounds, no diplopia, and resolved optic disc swelling.

Conclusion:
Weight gain is a know side effect of Quetiapine occurring in 7% to 35% of patients within the first 12 weeks of treatment. Most studies concentrate on the long term implications while IIH associated with excessive weight gain is not discussed. We report a 15 year old with IIH thought secondary the weight gain from Quetiapine.

References: None

Key Words: Quetiapine, Idiopathic Intracranial Hypertension, Weight Gain, Papilledema, Sixth Nerve Palsy

Financial Disclosure: None
Poster 41

Idiopathic Intracranial Hypertension in a Hutterite Family

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Introduction:
Idiopathic Intracranial Hypertension (IIH) is a condition characterized by elevated intracranial pressure whereby no etiology can be identified. There have been a few studies that have reported familial IIH (FIIH); however, the mode of inheritance responsible for FIIH remains unknown. Autosomal dominant, autosomal recessive and multifactoral causes have all been postulated. We report a case series of apparent FIIH in a single family of Hutterite women in order to elucidate further information about the pathogenesis and pattern of inheritance of this condition.

Methods:
Retrospective chart review. Nine medical records from a single family of Hutterite women with a high frequency of IIH were reviewed for data including demographics, age of onset, body mass index, past medical history, associated medical conditions, co-morbid conditions, symptoms of IIH, documented increased intracranial pressure, medications, visual outcome, and response to treatment. Available brain imaging results were also reviewed.

Results:
Five of seven sisters had convincing evidence of IIH. Of the two of seven sisters who did not have definite IIH, one had headaches and mildly constricted visual fields without papilledema, and the other had headaches only. Two young daughters of one sister had headaches only. None of the women had markedly elevated body mass index. One of the sisters with IIH developed venous sinus thrombosis years after diagnosis. Age of onset was similar in all five sisters with IIH, and visual outcome was highly variable. All were poorly responsive to treatment, and remain symptomatic.

Conclusion:
The frequency of IIH in this single family of first-degree relative Hutterite women further supports the idea that a genetic link may be present in this disorder, especially considering the high degree of consanguinity and intermarriage that exists within the Hutterite community. Further studies are needed to help identify a genetic link that may contribute to the pathogenesis of FIIH.

References:

Key Words: Idiopathic Intracranial Hypertension, Familial Idiopathic Intracranial Hypertension, Hutterite, Anabaptist, Pseudotumor Cerebri

Financial Disclosure: None
Post 42

Laboratory Profile of Idiopathic Intracranial Hypertension Patients: Does it Have Any Prognostic Value?

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Introduction:

Idiopathic intracranial hypertension (IIH) is a disorder of unknown cause affecting predominantly obese women of childbearing age. Previous studies reported association with hormonal imbalance, vasculitic and thrombophilic markers. However, as the pathophysiological influence of these conditions is not clear also their prognostic value had not been established.

Our objective was to evaluate laboratory findings of patients with IIH and to define any correlation between abnormal findings and disease course and outcome.

Methods:

All records of consecutive IIH patients who were followed for at least one year were reviewed. Demographics, medical history as well as ophthalmic manifestation, laboratory findings, management and outcome were collected. Main outcome measurements were visual functions at end of follow up, number of recurrences, number of drugs or surgery needed.

Results:

We included 83 patients with a mean follow up of 62 months, 74 women and 9 men. The mean age was 31± 13 years and mean BMI of 31±7 kg/m2. Seventy-three patients (88%) had at least one abnormal laboratory result. The most frequent abnormal results were inflammatory markers (25%), abnormal liver function (20%) anaemia (14%) and abnormal hormonal levels (9%).

Increased cortisol level correlated with a larger number of recurrences. In addition, it was found that patients with an elevated Lactate dehydrogenase (LDH) level were prone to a worse visual outcome. Patients exhibiting an elevated C-reactive protein level or a thrombophilic disturbance were less likely to experience a decline in the visual functions. The two groups did not differ regarding BMI.

Conclusion:

As in previous reports anaemia, endocrine abnormality and inflammatory markers were found. Liver functions abnormality was more frequent than in other studies. According to these results laboratory tests upon admission are not only of etiologic work up value but have also prognostic significance. Our study is limited being retrospective. A multicenter prospective study is needed.

References: None

Key Words: Idiopathic Intracranial Hypertension, Pseudotumor Cerebri

Financial Disclosure: None
Poster 43

Light-Adapted ERG b-Wave Amplitude in Neuromyelitis Optica

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Introduction:
The purpose of the study is to describe light-adapted electroretinograms (ERGs) responses in a series of patients with neuromyelitis optica (NMO).

Methods:
After comprehensive ophthalmological evaluation including best-corrected visual acuity measurement (BCVA), ERGs were recorded in 11 patients (7 anti-aquaporin-4 antibody-positive). 5 patients experienced bilateral and 6 unilateral neuritis, consequently 16 eyes NMO optical neuritis (group NO) and 6 unaffected fellow-eyes (FE). As control group (CTRL), eyes from 21 normally-sighted subjects, in the same age range, were examined.

ERG protocol was based on ISCEV standard, extended with a light-adapted luminance–response series to derive descriptors for the interrelation between b-wave amplitude and stimuli energy, from an non-normalized Gaussian function (Hamilton et al., 2007): h: saturation amplitude; x0: luminance for saturation, and w: non-dimensional slope-related parameter. Optical coherence tomography (OCT) was performed for retinal nerve fiber layer thickness (RNFL) measurement.

Results:
BCVA ranged from no-light-perception to 20/25 in NO and was 20/20 or better in CTRL and FE. Mean±SD light-adapted b-wave saturation amplitude (μV), parameter h, was NO: 172.36±49.32; and FE: 148.15±65.88, showing no statistically significant difference to CTRL: 182.80±26.71 (P=0.4190; ANOVA), while parameter x0 (cd.s/m²) was increased in NO: 0.91±0.31 and in FE: 0.80±0.24 in comparison to CTRL: 0.54±0.07 (P<0.05; Tukey-Kramer). Overall RNFL ranged from 33 to 79 μm in NO but was within the normative range for FE and CTRL.

Conclusion:
We found abnormalities for the interrelation between light-adapted b-wave amplitude and stimuli luminance in NO; and FE with normal BCVA and RNFL. Light-adapted ERG might add valuable information for diagnosis and follow-up in NMO.

References:

Key Words: Neuromyelitis Optica, Retina, Electroretinography

Financial Disclosure: None
Optical Coherence Tomography (OCT) Evaluation of Choroidal Folds in Patients with Idiopathic Intracranial Hypertension (IIH)

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Introduction:
Choroidal folds are parallel grooves or striae in the choroid of the posterior pole and may be a marker of progression in patients with IIH. The purpose of this study is to determine the ability of OCT to detect choroidal folds.

Methods:
Patients with a diagnosis of IIH were recruited from neuro-ophthalmology clinics at the Ottawa Hospital. Three dimensional colour and red-free fundus photographs of the disc-macula were obtained using a retinal camera (Topcon TRC-50DX). OCT topographical T-Scans of the central 28 degrees along with horizontal and vertical B-Scans through the macula were obtained using a spectral domain OCT (OPKO Spectral OCT/SLO).

Fundus photos and OCT images were viewed by staff neuro-ophthalmologists and clinical electrophysiologists and choroidal folds were graded on a 4 point scale: 1. No choroidal folds, 2. Choroidal folds on OCT only. 3. Choroidal folds on fundus photos only, 4. Choroidal folds on both fundus photos and OCT.

Examiners were blinded to both patient's names and clinical history.

Results:
Patient recruitment for this study began in February 2010. We have collected data from 40 eyes from 20 patients. 31 eyes were graded as category 1 and 7 eyes in category 4. Interestingly 2 eyes were graded in category 3.

Conclusion:
Patients with choroidal folds identifiable on clinical examination also had folds visible on OCT (category 4). This study demonstrates that OCT is a viable method for identifying choroidal folds in patients with IIH, although it may not be able to detect folds in all patients (category 3). This is a preliminary report for this study as patient recruitment is ongoing. We also intend to use multispectral retinal imaging technology (Annidis Health Systems) to determine whether one imaging modality may be better than another in detecting choroidal folds. At this time our study represents the largest cohort of IIH patients that have been examined by OCT.

References:

Key Words: Idiopathic Intracranial Hypertension, Optical Coherence Tomography, Choroidal Folds

Financial Disclosure: None
Poster 45

Experience with Normative Values for the Clinical Practice of Electroretinography

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Introduction:
It is generally recommended that every electroretinography (ERG) laboratory obtain their own set of normative values. In practice this is rarely done, and there are almost no publications reporting experience with locally obtained normal values for ERG. Individual ERG laboratories do not use the same equipment or configurations. Some institutions have older equipment, and others are very technologically advanced. It is important to know how an institution’s recordings compare to published normal values in order to define a practical range, to ensure high quality recordings, and to reliably interpret results locally and between institutions.

Methods:
Recordings were performed according to the International Society for the Clinical Electrophysiology of Vision protocol. Twenty normal adult subjects were recruited (40 eyes). Amplitudes and latencies of waveforms were measured by the same experienced Neurophysiology Technologist. Data was compared to results from a published study which used similar methods and the same number of subjects.

Results:
Our locally obtained normative values showed dramatic differences, compared to other published results. We found a wider range of normal responses between the 5th and 95th percentiles, markedly higher amplitudes, and variable implicit times. We experienced challenges measuring the intensity of the flash stimulus, calculating electrode impedance and reliably calibrating our older ERG equipment.

Conclusion:
At our institution, we have been using other published sets of normal values for the past 25 years to interpret our ERGs, and the differences we found between those results and our own demonstrate the importance of obtaining a set of normal values unique to each ERG lab for quality assurance. The validity and reliability of ERG results obtained on older equipment may be questionable, and results interpreted on the basis of other published normal values could lead to misdiagnosis.

References:

Key Words: Electroretinography, Normative Values, ERG

Financial Disclosure: None
Introduction:
Retinal nerve fibre layer (RNFL) thickness has been shown to be able to predict the visual prognosis in compressive optic neuropathies and quantify axonal loss in multiple sclerosis (MS). We attempt to study the relationship between RNFL thickness and visual acuity and visual field after optic neuritis (ON) to see if it is useful in predicting the cause of ON.

Methods:
This is a retrospective study. Patients with optic neuritis were identified using our neuro-ophthalmology database. Visual acuity, Humphrey visual field (HVF) and RNFL thickness were recorded at least 6 weeks after the acute episode of ON. The data were analysed using scatter plots.

Results:
Fourteen eyes were studied. Patients with ON secondary to MS had thinner average RNFL compared to those secondary to clinically isolated syndromes for the same mean deviation on the HVF, as well as visual acuity. We had one eye each with chronic relapsing inflammatory optic neuritis (CRION) and ON secondary to sjogrens syndrome. Both these eyes had poorer visual acuity as well as more severe field loss on HVF. Average RNFL thickness was thinnest in the eye with CRION.

Conclusion:
Average RNFL thickness done at least 6 weeks after the acute episode of ON may be useful in delineating the cause of optic neuritis. Prospective studies will be needed to further evaluate this hypothesis.

References:

Key Words: Retinal Nerve Fibre Layer, Optic Neuritis, Visual Field, Clinically Isolated Syndrome, Multiple Sclerosis

Financial Disclosure: None
Poster 47

Spasmus Nutans-Like Nystagmus, a Clinicopathological Case Presentation

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Introduction:
Spasmus nutans is a rare idiopathic disorder in the pediatric population comprising the clinical triad of nystagmus, head nodding and torticollis. Initially considered a benign self-limiting disorder, there have now been multiple reports associating the condition with parasellar tumours. This case aims to reiterate the importance of a careful neuro-ophthalmic examination and neuroimaging in atypical cases of spasmus nutans.

Methods:
A clinicopathological case presentation comprising video recording of the spasmus nutans-like nystagmus, neuroimaging, and pathological slides of the causative lesion.

Results:
A 3 year old boy admitted for pneumonia was incidentally noted to have a right ‘wobbly’ eye which had been present for the past 3 months. Examination revealed 6/60 vision in the right eye and a right relative afferent papillary defect. There was bilateral optic atrophy on fundoscopy. Magnetic resonance imaging revealed a cystic parasellar mass. The patient underwent transcranial excision and histology revealed a low-grade astrocytoma.

Conclusion:
A substantial proportion of patients presenting with spasmus nutans-like nystagmus have intracranial lesions. Atypical features in the history or examination should assist in the need for neuroimaging.

References:

Key Words: Spasmus Nutans, Nystagmus, Glioma, Head Nodding, Torticollis

Financial Disclosure: None
Intravascular Lymphoma Causing Sellar Mass Effect and Pituitary Apoplexy

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Introduction:
Intravascular lymphoma is a rare subtype of large cell non-Hodgkin's lymphoma characterized by proliferation of lymphoma cells within the lumina of small vessels. A literature review revealed no previous cases of intravascular large B cell lymphoma involving the pituitary gland presenting with neuro-ophthalmic findings.

Methods:
We present a case of a 68 year-old female with headache, vomiting, a right third nerve palsy and Horner's syndrome, found on MRI to have a sellar mass extending into the right cavernous sinus. The clinical presentation, laboratory results, and neuroimaging were initially consistent with apoplectic pituitary macroadenoma. The patient developed worsening headache, bilateral complete ophthalmoplegia and ptosis. Repeat neuroimaging revealed aggressive growth of the suprasellar mass into the clivus and cavernous sinuses bilaterally. Systemic exam was negative for malignancy.

Results:
Preliminary pathology from a transsphenoidal pituitary biopsy revealed areas of necrosis, consistent with pituitary apoplexy. The key pathological finding in this case – the presence of endovascular malignant lymphoid cells – was almost overlooked on the pituitary biopsy; however, the detection of these cells within a nasal polyp, incidentally biopsied on the approach to the pituitary gland, allowed the diagnosis of intravascular large B-cell lymphoma to be made. The patient had complete MRI resolution of tumor after three cycles of CHOP-R chemotherapy. At follow-up, she had a residual right third nerve palsy, but her other neuro-ophthalmic findings had completely resolved.

Conclusion:
We present a rare case of intravascular large B cell lymphoma within the pituitary gland, causing sellar mass effect and pituitary apoplexy. We suggest considering this rare pathology in the differential diagnosis of an atypically-behaving sellar mass. Treatment with CHOP-R is an effective treatment for this malignancy.

References:

Key Words: Pituitary Tumor, Intravascular Lymphoma, Diplopia, Third Nerve Palsy, Horner's Syndrome

Financial Disclosure: None
**Poster 49**

**Concordance of Neuro-imaging in Patients Referred to a University-Based Neuro-ophthalmology Service**

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**Introduction:**
Neuro-imaging studies are frequently ordered to investigate neuro-ophthalmic symptoms. These studies are expensive and time-consuming, and in some patients may be unnecessary or sub-optimal. Neuro-ophthalmologists may also re-interpret these studies based upon history and examination. The frequency of sub-optimal neuro-imaging studies and of re-interpretation in neuro-ophthalmology clinics is under-studied.

**Methods:**
We performed a prospective study of consecutive new patients referred to a university-based neuro-ophthalmology clinic for evaluation. All participants had received a neuro-imaging study in the last 12 months specifically related to their presenting symptoms. Participants then underwent a complete neuro-ophthalmic evaluation followed by a review of all prior neuro-imaging. Questions regarding quality of prior imaging and concordance of interpretation were answered by the attending physician.

**Results:**
The majority of study participants (83.5%) underwent MRI prior to referral. The rate of sub-optimal neuro-imaging studies was 38.8%, with incorrect study type being the most common cause (e.g. CT of head in evaluation of possible optic neuritis). After complete examination and review of prior neuro-imaging, our interpretation was in accord with the majority (77.4%) of the official radiology interpretations. When discordance occurred (15 events), the scan was most commonly re-interpreted as being abnormal.

**Conclusion:**
We found a high rate of sub-optimal neuro-imaging studies in patients referred to a neuro-ophthalmology clinic. We will present our final data, including specific rates and type of discordance. We will also include data regarding study type, location of pathology and change in diagnosis when discordance occurred and frequency of repeat neuro-imaging studies. These findings may have implications for resource allocation and utilization.

**References:** None

**Key Words:** Neuro-imaging

**Financial Disclosure:** None
Introduction:
Neuromyelitis optica (NMO) and Multiple sclerosis (MS) can both present with acute optic neuritis (ON), while differing considerably in their prognosis and management. Differences in the clinical course, serological testing and brain and spinal cord imaging of these diseases have been well documented. The purpose of this study was to look systematically for any potential differences in the imaging appearance of optic nerve in NMO and MS related ON.

Methods:
Magnetic Resonance Imaging of brain and orbits obtained within 6 weeks of acute optic neuritis episode in patients with securely diagnosed NMO (n=5) and MS (n=11) were retrospectively analysed by a neuroradiologist masked to the final diagnosis. Standardized scoring system was used to assess and analyze the extent and nature of the optic pathway involvement.

Results:
Of the 5 NMO patients, 4 had unilateral ON and 1 had bilateral disease. The most affected segment of the nerve was the orbital apex region in 4 patients, and chiasmal in 1. All 11 MS patients had unilateral disease, affecting the anterior retrobulbar portion of the nerve in 3 patients and the orbital apex in 8. No significant differences were observed in the presence, degree, or the type of signal alteration and contrast enhancement of the affected nerve segments between NMO and MS groups. A trend towards a higher chiasmatic involvement was noted in NMO group, with chiasmatic enhancement exclusively seen in NMO related ON (2 patients, p=0.057).

Conclusion:
In our retrospective review and limited sample size, the data suggests a higher propensity of NMO related ON to affect more posterior parts of the optic nerve including chiasm, and to have a simultaneous bilateral disease. Further study with larger sample size is needed.

References: None

Key Words: Magnetic Resonance Imaging, Optic Neuritis, Neuromyelitis Optica, Multiple Sclerosis

Financial Disclosure: None
Poster 51

Effects of Parietal Lobe Lesions on the Generation of Antisaccades

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Introduction:
Antisaccades are directed away from visual targets. They require suppression of reflexive saccades toward a visual target, as well as dispatch of volitional saccades in the opposite direction, to an instructed location with no visual target. Impaired antisaccade generation has been attributed to frontal lobe damage. However the role of the parietal lobe is not established. We studied antisaccade generation after unilateral focal parietal lobe lesions.

Methods:
Thirteen patients (age 45± 7 years; 7 women) with focal lesions (9 tumors; 5 vascular lesions) were instructed to make horizontal saccades away from a target light (antisaccades) flashed for 100 ms randomly to the right or left 10° from central gaze. The task was to look 10° in the opposite direction to the target flash. After 1 second a target appeared at the instructed position of the eye after the antisaccade (20 degrees from the target flash position. The percentage of antisaccades relative to wrongly directed reflexive saccades toward the target flash (prosaccades) and their accuracy and latencies were computed. Saccades were recorded by magnetic search coil technique. Ten healthy age-matched subjects served as controls.

Results:
The patient group made antisaccades in only 49.7 ± 32.7% of contraversive trials (visual target flash ipsilateral to lesions) and 49.6 ± 38.4% of ipsiversive trials. In the other trials they made errors; i.e. saccades toward the target flash. In contrast control subjects made 82.8 ± 13.7% correctly directed antisaccades. Eight patients showed subnormal antisaccade generation. Their imaged lesions were traced onto axial templates of brain anatomy, revealing overlap of lesions in white matter deep to Brodmann cortical areas 19, 39 and 37

Conclusion:
Generation of voluntary saccades is impaired by parietal lobe lesions. Impaired generation of antisaccades does not specify frontal lobe disease. Antisaccade tasks provide a means of measuring executive saccade function independent of visual guidance.

References:

Key Words: Saccades, Antisaccades, Parietal Lobe, Reflexive Saccades

Financial Disclosure: None
Balint’s Syndrome Presenting in the Posterior Reversible Encephalopathy Syndrome

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Introduction:
Balint’s syndrome of simultagnosia, optic ataxia, and misdirection of visually guided saccades usually occurs with bilateral parieto-occipital lesions. Posterior reversible encephalopathy syndrome (PRES) associated with systemic lupus erythematosus (SLE) has seldom been reported. Balint’s syndrome had not been reported in PRES. We describe Balint’s syndrome as an initial presentation of PRES in a patient with SLE.

Methods:
Case study of a 37 year old woman with abdominal pain, nausea and vomiting, alopecia, malar rash and arthritis who had abnormal anti double stranded DNA and anti cardiolipin antibodies. SLE complicated by nephritis was diagnosed and treated with Intravenous methyl prednisolone. She soon developed impaired vision initially attributed to cerebral SLE.

Results:
Visual acuity was difficult to record because of erroneous saccades toward optotypes, but was at least 20/400 in each eye. Although she made a full range of volitional saccades to directional commands she could not make visual guided saccades to small or large objects in any region of the visual fields where she could identify them by sight. Her hand movements were dysmetric when she tried to grasp a viewed object. She was unable to recognize more than one item at a time. Humphrey visual field showed a inferior right homonymous quadrantanopia. Blood pressure was 190/150. MRI showed: extensive FLAIR hyperintensity within both parietal lobes and the occipital lobes but normal diffusion weighted images and contrast venography. Optic ataxia resolved six days after initiation of treatment of hypertension and SLE. Simultanagnosia and impaired visually guided saccades had resolved after three months. MRI FLAIR at 6 months showed increased signal intensity in the occipital lobes suggesting residual gliosis.

Conclusion:
The relative roles of acute arterial hypertension and cerebral SLE in the pathogenesis of PRES are not known. Either of both may have been responsible. Balint’s syndrome can be an initial presentation of PRES.

References:

Key Words: Balint's Syndrome, Posterior Reversible Encephalopathy Syndrome (PRES), Systemic lupus Erythematosus, Saccades, Optic Ataxia

Financial Disclosure: None
Hemianopic Defects in Patients with Glaucoma

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Introduction:
A step across the vertical midline is an early sign of a hemianopic defect that suggests lesions of the visual pathways at or anywhere posterior to the chiasm. It is mandatory for a physician who finds a vertical step to rule out a neurological etiology before anything else. In the old literature a vertical step was also suggested to be present in approximately 20 - 30% of glaucomatous visual field defects on manual or automated perimetry. We have recently encountered glaucoma patients who had a persistent hemianopic defect on automated static perimetry. None of them had a CNS lesion by thorough neuro-imaging studies including MRI. It is suggested that some differences in vulnerability between the crossed and uncrossed optic nerve fibers may manifest themselves in glaucoma.

Methods:
Cases studied with MRI, perimetry and OCT

Results:
1) A 75-year-old woman with pseudo-exfoliation glaucoma OD had a sustained superonasal vertical step OD during a two-and-a-half-year follow-up period. Her OCT showed the corresponding thinning of the NFL OD. Her MRI did not show any causative lesion along the visual pathways. 2) A 77-year-old man with POAG OU had a superonasal quadrantanopia OS for more than 3 years since his first visit. His OCT showed corresponding NFL drop-out interotemporally along the optic disc OS. There were no positive findings on repeat MRI.

Conclusion:
Superonasal visual field is known to be particularly vulnerable to glaucoma damage. In our patients, the visual field defects approached the vertical meridian from the nasal hemifield and remained there respecting the vertical midline, which may suggest the greater resistance of the temporal hemifield. Optic nerve fibers subserving the nasal hemifield which do not cross the optic chiasm are phylogenetically younger structures. It is thus suggested that phylogenetic backgrounds may contribute to vulnerability of optic nerve fibers to glaucoma, as was suggested by Damgaard-Jensen (1977).

References:
1. Gilpin, Stewart, Shields, Miller, Hemianopic offsets in the visual field of patients with glaucoma, Graefe’s Arch Clin Exp Ophthalmo. 228,450-453,1990
3. Pereira, Kim, Zimmerman, Alward, Hayreh, Kwon, Rate and pattern of visual field decline in primary open-angle glaucoma, 109, 2232-2240, 2002

Key Words: Hemianopia, Vertical Step, Glaucoma, Automated Static Perimetry

Financial Disclosure: None
Clinical Features of the Patients with Oculomotor Nerve Palsy after Surgical Clipping of Posterior Communicating Artery Aneurysms

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Introduction:
Isolated oculomotor nerve palsy (ONP) induced by an unruptured posterior communicating artery (PCoA) aneurysm is a well-known clinical entity necessitating urgent surgical or endovascular treatment. The authors performed surgical clipping via a superciliary keyhole approach instead of a conventional pterional approach, and then performed preoperative and postoperative neuro-ophthalmological examinations.

Methods:
Nine patients presenting with complete (n = 6) or incomplete (n = 3) ONP were operated on using a superciliary approach. Preoperative and postoperative neuro-ophthalmological examinations including measurements of deviation angles, limitation of ocular motility, degree of lid ptosis and assessment of the field of binocular single vision were performed.

Results:
Preoperative mean deviation angle was 29 PD of exotropia (15-50 PD), average value of limitation of abduction of affected eye was -2 mm (-1~-4 mm) and all patients had diplopia in all gaze. Surgical clipping of the aneurysm was successfully performed in all patients. All 3 (100%) patients with incomplete ONP recovered completely within 1-2 months after the surgery, whereas 5 (83%) of the 6 patients with complete ONP recovered completely within 1-6 months after the surgery. Eight patients had no diplopia within 30º of binocular VF at postoperative 3 months.

Conclusion:
Surgical clipping of an unruptured PCoA aneurysm inducing ONP via a superciliary approach can provide favorable outcomes due to the complete obliteration of the aneurysm.

References: None

Key Words: Oculomotor Nerve Palsy, Unruptured Aneurysm, Minimally Invasive Surgical Clipping Of Aneurysm, Binocular Single Vision, Prognosis

Financial Disclosure: None
Additional Retrobulbar Triamcinolone Acetonide Injection for Retrobulbar Optic Neuritis Patients: Can this Procedure Accelerate Early Recovery of Visual Acuity?

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Introduction:
To evaluate the effect of additional retrobulbar triamcinolone acetonide (TA) injection on early recovery of visual acuity in retrobulbar optic neuritis

Methods:
We conducted a prospective, randomized clinical study including 31 patients of retrobulbar optic neuritis between March 2005 and July 2007. Patients were randomly divided into two groups: Group 1 (n=15) with retrobulbar TA (40mg) injection on the first day of ONTT protocol and group 2 (n=16) with conventional ONTT protocol. The following parameters were measured and analyzed: patient's sex, age, pupillary reactions, color vision, visual field and best-corrected visual acuity before treatment and after 1 day, 1 week, 2 weeks, 1 month and 3 months of follow-up.

Results:
Mean visual acuity before treatment was 1.00±0.89 logMAR units in the group 1 and 0.98±0.75 logMAR units in the group 2. One day after injection, visual acuity was better in the group 1 (0.50±0.42 logMAR units) than in the group 2 (0.73±0.61 logMAR units), however, there was no statistically significant difference between the two groups (p=0.07). There was no significant difference in visual acuity, recovery of RAPD, color vision and visual field at 3 months of follow-up. No serious side effect related to retrobulbar TA injection was observed.

Conclusion:
Additional retrobulbar TA injection may help the patients of optic neuritis who have special need for prompt visual recovery. However, further studies will be required to ascertain whether this procedure can help early recovery of visual acuity on retrobulbar optic neuritis.

References: None

Key Words: Retrobulbar Optic Neuritis, Additional Triamcinolone Acetonide Retrobulbar Injection, ONTT Protocol, Visual Acuity Change

Financial Disclosure: None
Poster 56

Features of Ocular Motor Abnormality in Early Stage of Brain Stem Infarction

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Introduction:
To investigate the characters of ocular motor abnormality in the early stage of brain stem infarction, and evaluate its value in the early diagnosis.

Methods:
A total of 24 patients (19 males, 5 females) with initial diplopia were chosen from 287 patients with brain stem infarction. The mean age was 64 (range, 42-81). their clinical features, accompanied symptom and related examination were retrospectively analyzed.

Results:
The risk factors included hypertension in 20 cases (83.3%), coronary heart disease in 9 cases (37.5%), diabetes mellitus in 7 cases (29.12%), atrial fibrillation in 3 cases (12.5%). About 87.5% patients complained with dizziness, and 16.7% with nausea, 16.7% with limb ataxia, 8.3% with vertigo, 4.2% with limb weak. The infarcts of 9 patients (37.5%) were located in midbrain, 14 patients (58.3%) in pons and 1 patient (4.2%) in medulla. Nuclear oculomotor nerve palsies were seen in the midbrain infarction, in which medial rectus weakness (8 cases) was dominant compared with other ocular muscles. The oculomotor disturbances of pons infarction were various, which included internuclear ophthalmoplegia in 3 cases, the abduct nerve palsy in 3 cases, the oculomotor nerve palsy in 2 cases, the para-Horner syndrome in 1 case, and nystagmus in 10 cases.

Conclusion:
In brain stem infarction, the main characters of ocular motor abnormality include nuclear ophthalmoplegia, internuclear ophthalmoplegia and nystagmus, which play an important role in the early diagnosis.

References: None

Key Words: Brain Stem Infarction, Ophthalmoplegia, Neuro-Ophthalmic Signs, Diagnosis

Financial Disclosure: None
**Clinical Manifestation of Intracranial Aneurysms in Neuro-Ophthalmology**

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**Introduction:**
To explore the principals of relationship between ophthalmic changes and intracranial aneurysms by investigating ophthalmic symptoms result from intracranial aneurysms.

**Methods:**
We analyzed the relationship between different ophthalmic changes and different intracranial aneurysms, the effect of different operational interventions on the ophthalmic symptoms' improvement.

**Results:**
Among 23 patients with ophthalmic changes, 12 cases (52.17%) harbored posterior communicating aneurysms, petrosal segment internal carotid artery and paraclinoid internal carotid artery aneurysms were both 2 cases (8.70%), ophthalmic internal carotid artery, anterior cerebral artery, middle cerebral artery, posterior inferior cerebellar artery, intracavernous primary trigeminal artery, carotid bifurcation aneurysms were all 1 case (4.35%). The main ophthalmic symptoms of posterior communicating aneurysms were various degrees of oculomotor nerve palsy. Paraclinoid aneurysm accompanied by visual acuity decreasing, primary trigeminal artery aneurysm by abducens nerve palsy and petrosal segment internal carotid artery aneurysm by paroxysmal diplopia respectively. No improvement appeared in patients who underwent emboilization. All patients who underwent neck clipping had no or limited improvements.

**Conclusion:**
There exist some rules between ophthalmic changes and intracranial aneurysms with different locations and properties. With ophthalmic abnormal changes we can obtain earlier diagnosis of intracranial aneurysms, thus improve the prognosis of intracranial aneurysms remarkably.

**References:** None

**Key Words:** Intracranial Aneurysms, Ophthalmic Symptoms, Cranial Nerve Palsy

**Financial Disclosure:** None
Bálint’s Syndrome and Visual Allochiria in a Patient with Reversible Cerebral Vasoconstriction Syndrome

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Introduction:
We describe a patient with Bálint’s syndrome and allochiria from Reversible Cerebral Vasoconstriction Syndrome (RCVS).

Methods:
Case study.

Results:
A 67-year-old female presented with “confusion” and left arm weakness. Medical history included hypertension, hyperlipidemia, and ocular migraines. She was hospitalized the week previous after experiencing thunderclap headache (TCH). During that admission, MRI showed enhancement within the leptomeninges; neurologic exam and multiple CSF/serologic studies were normal and she was diagnosed with Posterior Reversible Encephalopathy Syndrome.

On exam she had right gaze preference, simultagnosia, and left-field deficit. Left-field visual stimuli were occasionally reported on the right. Voluntary saccades and arm reaching were abnormal. Strength was 3/5 throughout the left arm.

MRI showed acute infarction involving the right parietal-occipital region. Repeat MRI 9 days later showed multiple interval infarctions including the left parietal-occipital junction. MRA and conventional angiography showed multi-focal concentric narrowing of proximal intracranial arteries bilaterally.

RCVS was diagnosed and verapamil started. At 1 month, she demonstrated improvement in neglect, saccades, arm reaching, and strength. MRA showed near-complete resolution of MCA and ACA stenoses.

Conclusion:
This patient has Bálint's syndrome: simultagnosia, optic ataxia, and ocular apraxia. She also demonstrates allochiria, a phenomenon of right parietal dysfunction in which a stimulus in the left hemi-space is reported to be present to the right.

Bálint’s syndrome is typically caused by bilateral parietal-occipital pathology. Our patient’s initial imaging revealed infarction limited to the right hemisphere. Bálint’s syndrome likely occurred from infarction to the right parietal-occipital region with concurrent left parietal-occipital ischemia causing functional impairment without radiologic abnormality. Perfusion imaging may have been confirmatory of left-hemispheric ischemia.

RCVS is characterized by reversible vasoconstriction involving intracranial arteries. Patients typically present with TCH, normal CSF, and various neurologic deficits; rapid improvement of vasospasm is common. This case introduces RCVS as a novel cerebrovascular cause of Bálint’s syndrome.

References:

Key Words: Bálint's Syndrome, Visual Allochiria, Cortical Visual Deficits, Cerebrovascular, Reversible Cerebral Vasoconstriction Syndrome

Financial Disclosure: None
**Poster 59**

**OCT Confirmation of Myelinated Nerve Fiber Layer in Autosomal Recessive Spastic Ataxia of Charlevoix-SAguenay**

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**Introduction:**
Autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) is a neurodegenerative process characterized by ataxia, dysarthria, spasticity, distal muscle wasting, finger and foot deformities, nystagmus and myelination of retinal nerve fibers as well as atrophy of the cerebellar vermis. We report a case of ARSACS examined with optical coherence tomography.

**Methods:**
Neurologic and ophthalmological examinations were performed. Optical coherence tomography (OCT) of the disc and macula was performed. Genetic testing was done.

**Results:**
OCT showed the presence of a highly reflective myelin layer inferior to the internal limiting membrane and emanating from the optic disks.

Sequence analysis of the spastic ataxia of Charlevoix-Saguenay (SACS) gene revealed two different mutations: A missense mutation c3932T>A (p.Met1311Lys) was found in addition to an in-frame deletion in exon 9: c.1065110656del. (p.Met3551 Leu3552del) which is a deletion of two highly conserved amino acids

**Conclusion:**
This is a genetically confirmed case of ARSACS examined with optical coherence tomography confirming the presence of myelinated nerve fibers.

**References:**

**Key Words:** Myelinated Nerve Fibers, Spastic Ataxia

**Financial Disclosure:** None
Ocular Complications of Pediatric Bone Marrow Transplantation

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Introduction:
Late Complications of BMT affect various organs including the eyes. In previous studies on ocular complications of BMT in the pediatric population, all children underwent ablation of the bone marrow by cytoreductive chemotherapy and total body irradiation (TBI) prior to the transplant. The overall rate of complications for the anterior segment were 35.5% (1) and 75.8% (2) and for the posterior segment 6.9% (1) and 13.5% (2). The objective of our study was to assess the rate of ocular complications in children who had undergone allogeneic BMT without TBI.

Methods:
Prospective study of children undergoing BMT without TBI. The outcome measures were visual acuity, slit-lamp biomicroscopy, fundus examination and Shirmer test or BUT (tear break-up time) measurements. Children were examined prior to BMT and periodically thereafter.

Results:
Thirty-seven children participated in the study, 22 boys and 15 girls. Twelve children were excluded because of the absence of ophthalmologic follow-up. Indications for BMT were: Aplastic anemia (3 cases), myelodysplastic syndrome (4), various types of leukemia (13), Fanconi anemia (3), thalassemia (1), combined immune deficiency (1), Wiscott Aldrich (1). Mean age was 8.6 ± 6.3 years. Average follow up was 21.0 ± 16.8 month. On the first follow up examination (9.0±5.9 month), 4 out of 25 (16%) had ocular complications. One patient had cataract that required surgery, and one patient had severe graft versus host disease (GVHD) that required insertion of punctual plagues and intensive lubrication. None of our patients had posterior segment complications.

Conclusion:
Ocular complications of BMT in our study were not as common as previously reported (1,2). Ocular symptoms were generally mild and the majority of patients retained excellent visual functions. As opposed to the other studies, our patients did not receive TBI before BMT; this may explain lower rate of ocular complications and milder complications.

References:

Key Words: Bone Marrow Transplant, Ocular Complications, Graft Versus Host Disease (GVHD)

Financial Disclosure: None
**Poster 61**

**Downbeat Nystagmus and Convergence Spasm in a Patient with GAD Autoimmunity Syndrome**

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**Introduction:**
Glutamic acid decarboxylase (GAD) enzymatically catalyses decarboxylation of glutamate to gamma-aminobutyric acid (GABA), the major inhibitory CNS neurotransmitter. High levels of autoantibodies to GAD are associated with numerous neurologic disorders including stiffman syndrome, limbic encephalitis, epilepsy, and cerebellar ataxia. Many patients have comorbid type I diabetes. We treated and memorialized on film a patient with cerebellar ataxia, downbeat nystagmus, slow saccades and intermittent convergence spasms with proved GAD autoimmunity syndrome.

**Methods:**
Case report and literature review.

**Results:**
A 48 y/o man developed slowly progressive gait ataxia associated with asymmetric downbeat nystagmus. Over a three year period additional findings included slowed refixational saccades and intermittent severe brief convergence spasms and he developed type I diabetes. Diagnostic profiling included normal MRI scanning and serologic investigations for vitamin and mineral deficiency, paraneoplastic profile, hereditary cerebellar degeneration.

Endomesial antibodies were present but subsequent multiple small bowel biopsies demonstrated no evidence of either celiac or Whipple’s disease. GAD – 65 autoantibodies however were 70 times normal and CSF analysis revealed 10 oligoclonal bands. Treatment with immunoglobulin lessed the nystagmus and ataxia but convergence spasm persists.

**Conclusion:**
Downbeat nystagmus occurs consequent to many conditions: structural, metabolic and immunologic including, the GAD antibody syndrome. The additional findings of slow saccades and convergence spasm in our patient are rather unique and in some ways resemble eye movement findings observed in Whipple’s disease.

A serologic investigation for the presence of GAD antibodies should be included in the standard evaluation of individuals with downbeat nystagmus. The presence of type I diabetes should increase the suspicion for this autoimmune syndrome. As in our case, functional improvement in other GAD associated neurologic disorders with either immunoglobulin infusion or plasmapheresis has been seen.

**References:**

**Key Words:** Downbeat Nystagmus, GAD, Convergence Spasm, Autoimmunity Syndrome

**Financial Disclosure:** None
Introduction:
The diagnosis of giant cell arteritis (GCA) is based on clinical signs and symptoms and confirmed by serological tests (elevated ESR, CRP, Platelets) and positive temporal artery biopsy. GCA may affect multiple arterial beds. We treated a patient with biopsy proven GCA in whom sural nerve biopsy also showed evidence of arteritis.

Methods:
Observational case report and literature review.

Results:
An 83yr-old man presented with 3-month history of unremitting temporal headaches and blurred vision. On examination he exhibited temporal scalp tenderness. Temporal artery biopsy confirmed the diagnosis of giant cell arteritis. Steroid therapy was initiated, however due to progressive bilateral lower extremity weakness and sensory loss plus persistently elevated inflammatory markers, a sural nerve biopsy was performed. Histological exam showed focal perivascular lymphocytic infiltrates and focal epineural blood vessel scarring consistent with arteritis.

Conclusion:
Peripheral nerve involvement may occur temporally associated with other signs and symptoms in GCA. Arteritis may injure peripheral nerves diffusely or individually, example peroneal, tibial, sural nerves. Brachial plexus is particularly vulnerable and may be the presenting sign of temporal arteritis. As is the case in other large vessel arteritides, peripheral nerve biopsy may lend supportive evidence in suspected cases of giant cell arteritis.

References:

Key Words: Peripheral Nerve, Giant Cell Arteritis, Temporal Arteritis, Sural Nerve

Financial Disclosure: None
False Negative Apraclonidine Test in a Patient with Horner Syndrome due to Carotid Endarterectomy

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Introduction:
Horner syndrome is characterized by miosis and ptosis associated with ipsilateral injury to the sympathetic nervous system. The current gold standard of pharmacologic testing is performed using cocaine, which is costly and difficult to obtain for administration in topical form. More recently, topical apraclonidine has been used and found to result in a reversal of anisocoria in Horner syndrome in most cases. We report a case of Horner syndrome which was tested with apraclonidine on two separate days and found to have false negative results based on positive cocaine testing on a separate day.

Methods:
Our patient was found to have ptosis and miosis 35 hours following carotid endarterectomy. One drop of 0.5% apraclonidine was instilled in both eyes and the pupils were assessed after 30 minutes. Apraclonidine testing was repeated 14 days after the presumed insult. Cocaine testing was also performed by instilling two drops of 10% cocaine in both eyes and the pupils were assessed after 45 minutes.

Results:
On two separate days of apraclonidine testing, no pupillary changes occurred on either day. With cocaine testing, the anisocoria increased, indicating a positive test result.

Conclusion:
The current criterion for positive apraclonidine testing is a reversal of baseline anisocoria. In our case, a patient with symptoms consistent with Horner syndrome tested negative based on apraclonidine testing despite a positive test with cocaine. Our results suggest that apraclonidine may not be an equivalent method of testing compared to cocaine in cases with high suspicion for Horner syndrome. Further large studies need to be conducted to compare the sensitivity and specificity of these pharmacologic tests.

References:

Key Words: Horner Syndrome, Apraclonidine, Cocaine, False Negative, Pharmacologic Testing

Financial Disclosure: None
Is there a Critical Dosage and a Minimum Interval in Amiodarone-Related Optic Neuropathy?

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Introduction:
While amiodarone-related optic neuropathy has become a well recognized entity its exact cause is still unclear. Usually, the interval between initiation of orally administered amiodarone treatment and onset of visual loss is several months. No critical dosage and no critical time interval have been defined so far.

Methods:
A 64-year-old patient is reported who developed bilateral, simultaneous, insidious onset of visual loss 48 hours after initiation of intravenous amiodarone treatment to restore normal sinus rhythm in atrial fibrillation. In addition, a literature review regarding doses at diagnosis and intervals between starting amiodarone therapy and onset of optic neuropathy was conducted.

Results:
The patient had received a total of 2 g intravenous amiodarone. Visual acuity was 20/25 in the right eye and 20/60 in the left eye. Fundus examination revealed bilateral optic disc edema. Visual field examination showed a right concentric defect and a left superior altudinal plus central scotoma. Visual acuity slightly recovered in the left eye (20/40) while visual fields did not change over time. According to the literature, the typical patient with amiodarone-related optic neuropathy develops the disease during oral ingestion (400 mg daily) 6-7.7 months after treatment initiation.1,2

Conclusion:
The incidence of optic neuropathy in patients receiving amiodarone has been estimated at 1.79%.3 However, in the vast majority of cases neuropathy developed in a scenario of month-long oral ingestion (mean, 6-7.7 months), with a mean maintenance dosage of about 400 mg/d. To our knowledge, only one patient has been reported with optic neuropathy after receiving intravenous amiodarone. Our case underlines the potential harm of amiodarone to the optic nerve. In intravenous administration the higher, potentially toxic dosage and a hypotension due to vasodilatation and depression of myocardial contractility may contribute to the pathogenesis.

References:

Key Words: Amiodarone, Toxic Optic Neuropathy, Vision Loss, Visual Field Loss

Financial Disclosure: None
Introduction:
A raised erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are highly sensitive for a diagnosis of GCA. It has been reported that thrombocytosis is a more useful marker than an elevated ESR or CRP as measured by the positive predictive values (PPV) and negative predictive values (NPV)\(^1\). We show in a larger study the importance of the context of the PPV and report and validate a higher likelihood ratio (LR) associated with thrombocytosis as compared to elevated ESR and CRP.

Methods:
Retrospective 5 year chart review. Cases identified sequentially, underwent temporal artery biopsy (TAB) in a single tertiary referral unit. Inflammatory markers prior to biopsy were analysed. A histological diagnosis was recorded in addition to a clinical diagnosis based on the final diagnosis of the leading clinician.

Results:
Of 270 biopsies, 66 were ‘positive’ that is, diagnostic of giant cell arteritis. 191 were ‘negative’. The sensitivity for a retrospective diagnosis of GCA was highest for a raised CRP followed by ESR and lastly a raised platelet count (94%, 78% and 76% respectively). Conversely the specificity was highest for thrombocytosis, intermediate for a raised ESR and lowest for a raised CRP (67%, 40% and 20% respectively). The positive predictive values (PPV) behaved differently to the sensitivity and specificity values in that they varied according to the diagnostic criterion used. Likelihood ratios were not influenced by diagnostic criteria and showed the importance of thrombocytosis which achieved higher values (2.2 – 2.3) than either a raised ESR (1.26-1.3) or CRP (1.2).

Conclusion:
Measuring the platelet count is an important part of the assessment of patients suspected of having GCA. The higher likelihood ratio associated with thrombocytosis as compared to a raised ESR or CRP shows the greater value of this finding in supporting a diagnosis of GCA.

References:

Key Words: Giant Cell Arteritis, Thrombocytosis, Temporal Arteritis, Erythrocyte Sedimentation Rate, C-Reactive Protein

Financial Disclosure: None
Ultrasound as the Primary Diagnostic Tool of Orbital Vascular Lesions

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Introduction:
Accurate diagnosis of orbital vascular lesions is critical in management. Current diagnostic imaging techniques include magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), computed tomography (CT), CT angiography, nuclear scintigraphy, color Doppler imaging, and A- and B-mode orbital echography.1-5 Recent authors advocate dynamic MRI/MRA as superior.6,7 However, MRI and CT present issues of radiation exposure, potential reactions to contrast material, extended testing times, and expense. They do not permit real-time assessment of lesion characteristics with alteration of patient’s posture or activity. We demonstrate the utility of orbital ultrasound in evaluation and management of suspected orbital vascular malformations.

Methods:
15 patients with presumed orbital vascular malformations were examined with Standardized Echography by an experienced echographer, and data were compared with other available imaging modalities to determine appropriate management. Patients were monitored for improvement, stability, or worsening of symptoms and signs.

Results:
Ultrasonography findings include low reflectivity at blood-filled regions, compressibility, and both irregular and well-circumscribed borders.Valsalva maneuver during ultrasound examination showed lesion enlargement in three patients. Based on ultrasound results, three patients were diagnosed with lymphangiomas, three with varices, one with distensible venous malformations, one with arteriovenous malformation, and six with nonspecific vascular malformations. Based on echographic data, five underwent surgery whereas nine were observed. CT, MRI, or both were performed in all patients. Radiographic diagnosis concurred with ultrasound in eight (57%) cases and provided no conflicting diagnostic information. Confirmation of final diagnosis was achieved via histopathology in seven patients and additional imaging, clinical course, or both in the remainder. During follow-up for 0 – 14 (mean 3.3) years, all patients demonstrated improvement or remained stable. None were found to have a different type of lesion, and no complications developed from misdiagnosis.

Conclusion:
Orbital ultrasound provides reliable imaging parameters and can be used as the primary imaging modality when evaluating suspected orbital vascular malformations.

References:

Key Words: Ultrasound, Orbit, Vascular malformation, Diagnosis

Financial Disclosure: None
Poster 67

Adenoid Cystic Carcinoma with Simultaneous Compressive Optic Neuropathy and Choroidal Metastasis

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Introduction:
Adenoid cystic carcinoma has only been reported very rarely to have choroidal metastases. Herein, we describe the first case reported in the literature in which adenoid cystic carcinoma metastasizes to the choroid, in addition to extending intracranially to involve the optic nerve and causing optic neuropathy.

Methods:
Case report and review of the literature.

Results:
A 26 year old woman with metastatic adenoid cystic carcinoma arising from the left maxillary sinus underwent resection, radiation and Erbitux. One year later, she experienced the sudden onset of visual hallucination of “sunlight” in the central portion of her left eye, associated with ipsilateral headache. Examination revealed visual acuity of 20/20 OD and 20/400 OS, dyschromatopsia and a left afferent papillary defect. Visual fields showed a superonasal depression OD and only an inferonasal island OS. Dilated fundus examination revealed normal optic nerves OU, but there a raised choroidal mass in the inferotemporal macula of the right eye.

MRI revealed an enhancing soft tissue mass in the sphenoid sinus with intracranial extension. The left prechiasmatic optic nerve was displaced and encased with associated enlargement and edema.

Ultrasound of the right eye revealed high internal reflectivity of the choroidal mass.

Intravenous steroids were given and the next day she began taxotere, but by 1 week later, the left eye had declined to no light perception. Fractionated focal radiation therapy to the tumor was started to prevent spread to the chiasm.

Conclusion:
Adenoid cystic carcinoma is an uncommon form of malignant neoplasm arising most commonly from the salivary glands of the head and neck. Intracranial extension with infiltrative optic neuropathy is rare while choroidal metastases are extremely rare. To our knowledge, the presence of intracranial optic neuropathy along with choroidal metastasis has not yet been reported.

References:

Key Words: Optic Neuropathy, Choroidal Metastases, Adenoid Cystic Carcinoma

Financial Disclosure: None
A National Survey of Practice Patterns: Unilateral Vs Bilateral Temporal Artery Biopsy

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Introduction:
Temporal artery biopsy (TAB) remains the gold standard in establishing the diagnosis of giant cell arteritis. Several studies have assessed the similarities in bilateral specimens obtained from affected patients. (1-3) Discordance in the right and left side has varied from 1% (1) to 13% (3). This difference has been used by advocates in recommending both unilateral and bilateral TAB, with no clear consensus. This study assesses current clinical practice patterns among oculoplastic surgeons and neuro-ophthalmologists.

Methods:
A survey was sent via e-mail to the members and affiliates of the American Society of Ophthalmic Plastic and Reconstructive Surgery (ASOPRS) and the North American Neuro-Ophthalmology Society (NANOS) using the Survey Monkey website.

Results:
Primary sub-specialty of the 262 respondents are as follows: oculoplastic surgery (n=129), neuro-ophthalmology (n=120), both oculoplastic surgery and neuro-ophthalmology (n=14), and other (n=4). Among the respondents, some (n=192) perform TAB whereas others refer for biopsy (n=72). Overall, 83% (n=210) advocated initial unilateral TAB, 8% (n=19) advocated bilateral biopsy, and the remainder (9%, n=24) recommended either unilateral or bilateral TAB depending on clinical suspicion. Of those that advocated initial unilateral TAB, opinions were split as to whether they would (n=56, 27%) or would not (n=98, 47%) advocate biopsy on the fellow side. The remaining respondents reported the clinical situation would have to be considered before deciding whether or not to biopsy the second side. One difference in practice patterns was found: oculoplastic surgeons were less likely to advocate biopsy of the second side (n= 25, 30%) than neuro-ophthalmologists (n=31, 58%, p<0.05, Fisher’s exact test).

Conclusion:
TAB practices vary greatly with regards to laterality. Most (83%) advocate initial unilateral biopsy but opinions vary on when it is appropriate to biopsy the second side. This lack of consensus between and within each sub-specialty underscores the need for a systematic assessment of our varying practice patterns.

References:

Key Words: Giant Cell Arteritis, Temporal Artery Biopsy

Financial Disclosure: None
Poster 69

A National Survey of Practice Patterns: Effect of Prior Treatment with Corticosteroids on Diagnostic Capability of Temporal Artery Biopsies in Giant Cell Arteritis

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Introduction:
Temporal artery biopsy (TAB) remains the gold standard in establishing the diagnosis of giant cell arteritis (GCA). To ensure vision preservation, immunosuppressive therapy is often initiated prior to obtaining the TAB. The effect of treatment with corticosteroids on the pathological diagnosis of GCA has been assessed by several investigators with conflicting results. (1, 2) This study was designed to assess the current opinion within the ophthalmologic community of the effect of steroids on the diagnostic accuracy of TAB.

Methods:
A survey was sent via e-mail using Survey Monkey to the members and affiliates of the American Society of Ophthalmic Plastic and Reconstructive Surgery (ASOPRS) and the North American Neuro-Ophthalmology Society (NANOS).

Results:
Primary sub-specialty of the 262 respondents were as follows: oculoplastic surgery (n=129), neuro-ophthalmology (n=120), both oculoplastic and neuro-ophthalmology (n=14), and other (n=4). Opinions varied on steroid treatment prior to TAB and were as follows: results are unreliable after 7 days (n=46, 21%), 10 days (n=33, 15%), 14 days (n=47, 21%) and greater than 14 days (n=95, 43%) after initiation of systemic steroids. More oculoplastic surgeons (n=33, 31%) felt a biopsy was unreliable after 7 days of steroid therapy than neuro-ophthalmologists (n=13, 12%, p<0.01, Wilcox rank sum test). In line with this trend, 54% of neuro-ophthalmologists reported a biopsy was still reliable after 14 days of steroid treatment, whereas only 31% of oculo-plastic surgeons felt the biopsy was reliable in the same time frame (p<0.01, Wilcox rank sum test).

Conclusion:
Opinion varied greatly regarding the accuracy of TAB in the diagnosis of GCA following initiation of systemic corticosteroid treatment. Although a slight majority of neuro-ophthalmologists (54%) believe TAB to remain accurate after 14 days of steroid therapy, no trend was seen among oculoplastic surgeons. This lack of consensus underscores the need for a systematic assessment of the effect of steroid treatment on histological features indicative of GCA.

References:

Key Words: Giant Cell Arteritis, Treatment, Temporal Artery Biopsy, Pathology

Financial Disclosure: None
Primary Divergence Insufficiency Mainly Occurs in the Caucasian Population with a Loss of Orbital Elasticity

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Introduction:
Divergence Insufficiency (DI) is an acquired comitant esotropia which manifests as horizontal diplopia at distance, but not near. Varying subgroups have been described including young versus older patients, and idiopathic (Primary) versus those with underlying neurologic disease (Secondary). Secondary DI includes increased intracranial pressure, cerebellar degeneration, brainstem stroke, etc

Methods:
After IRB approval, a retrospective chart review was conducted of patients diagnosed with DI from 2005 to 2010 at our clinic. All patients were evaluated by one neuro-ophthalmologist. The data collected included age, race, sex, esotropia in prism diopters (PD) at distance, length of follow-up and change in PD, treatment, neuroimaging, and PMH.

Results:
Data from 20 patients was collected. Mean age was 78, with range of 53-92, with all but the youngest older than 67. All of our patients were Caucasian. There was a female predominance of 15:5. Initial esotropia ranged from 2-25 PD with a mean of 7. Twelve of the twenty patients had follow-up with a mean of 15 months and a range from 1 month to 3 years. Overall change in prism diopters ranged from -9 to +4, with two patients improving, seven patients remaining stable, and three patients worsening. Two of the patients required no treatment, fourteen received Fresnel prisms, four received ground-in prisms, and none required surgery. Neuroimaging revealed eight patients with MRI, six patients with CT, and six patients (30%) with no neuroimaging. MRI revealed: 5/8 with ischemic changes, 6/8 with cortical atrophy. CT revealed: 2/6 showed ischemic changes, 4/6 showed cortical atrophy. Hypertension was found in 14/20 (70%), hyperlipidemia in 7/20 (35%), and diabetes in only 1/20 (5%).

Conclusion:
Divergence Insufficiency patients presenting to our clinic were older Caucasians. This may be related to a loss of orbital elasticity which would correlate with the loss of skin elasticity seen in aging Caucasians compared to African-Americans.

References:

Key Words: Divergence Insufficiency, Strabismus, Esotropia

Financial Disclosure: None
Poster 71

Pseudotumor Cerebri as a Complication of Renal Dialysis

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Introduction:
Pseudotumor Cerebri (PTC) has been described in individual patients with chronic renal failure and dialysis.\textsuperscript{1-6} We report a series of 4 patients with PTC associated with chronic renal failure and dialysis.

Methods:
Case studies of four patients with brain imaging, fundus photography, perimetry, and cerebrospinal Fluid (CSF) analysis.

Results:
Patient 1, a 45 year old woman peritoneal dialysis presented with headaches, confusion, and decreased vision. She had right disc swelling and an atrophic left disc. CSF opening pressure (OP) was 31 cm H\textsubscript{2}O. She was placed on acetazolamide. Patient 2, a 26 year old woman on peritoneal dialysis developed decreased vision in both eyes. She had bilateral disc edema. MRV demonstrated narrowing of the left distal transverse sinus. CSF OP was 31 cm H\textsubscript{2}O. Acetazolamide was started. Patient 3, a 46 year old female presented 5 years post renal transplant and on dialysis with decreased vision. MRI/MRV was normal. CSF OP was 42 cm H\textsubscript{2}O. She was started on acetazolamide. Patient 4, a 44 year old woman on peritoneal dialysis developed transient vision loss post dialysis for the past year. On examination, she had light perception vision and bilateral disc edema. MRI was normal, and CSF OP was 30 cm H\textsubscript{2}O. Acetazolamide improved her symptoms.

Conclusion:
Patients with visual disturbances during and after hemodialysis should be suspected as having dialysis associated PTC. During dialysis there is a relative increase in brain or CSF osmolarity due to the delayed removal of urea. Recurrent or chronic dialysis disequilibrium with osmotic influx of water can increase CSF pressure and result in permanent visual loss from papilledema.

References:

Key Words: Pseudotumor Cerebri, Idiopathic Intracranial Hypertension, Renal Failure, Papilledema, Dialysis disequilibrium

Financial Disclosure: None
Poster 72

Partial Blink Rate in Parkinson Disease and Progressive Supranuclear Palsy

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Introduction:
Parkinson disease (PD) and progressive supranuclear palsy (PSP) are neurodegenerative diseases that share some motor features. Ocular motility and blink rate dysfunction of these diseases have been characterized, but partial blinks (PB) have not been characterized in the literature.

Methods:
Videos of 22 patients with PD and 6 with PSP were reviewed and the rate of partial blinks recorded.

Results:
Among the PD cohort, 18/22 patients demonstrated PB with a mean rate of 6.4 partial blinks/minute; the 4 PD patients without PB were all female. Among the PSP cohort, 5/6 patients had PB with a mean rate of 4.5 partial blinks/minute.

Conclusion:
Partial blinks are seen in both PD and PSP, represent a newly characterized motor feature, and may be a common marker of extrapyramidal diseases.

References: None

Key Words: Parkinson Disease, Progressive Supranuclear Palsy, Partial Blink, Extrapyramidal

Financial Disclosure: None
Monovision Correction for Small Angle Binocular Diplopia

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Introduction:
The management of diplopia is based on eliminating the doubled image or restoring binocularity surgically or optically. Monovision correction may prove to be a treatment option for presbyopic patients with diplopia as it blurs one of the doubled images and facilitates suppression. This is the only prospective study in literature assessing the efficacy of monovision correction in the treatment of acquired small angle binocular diplopia in adult patients.

Methods:
Twenty patients with symptomatic uncorrected diplopia were consecutively enrolled in a prospective treatment trial. All had stable deviations of 10 prism diopters or less for over three months. Each received monovision spectacles and/or contact lenses with distance correction in the dominant eye. Half of the patients received +3.00 diopter (D) add to their distance refraction in the fellow eye and the others received +2.50 D add. Subjects were re-examined after six to eight weeks. The validated and standardized Diplopia Questionnaire and Amblyopia and Strabismus Questionnaire (ASQE) were used to quantify the efficacy of monovision correction for diplopia by measuring the functional impact on vision-specific quality of life.

Results:
Based on the Diplopia Questionnaire, both groups had a significantly less diplopia with monovision (scores: 42.9 in +2.50D group, 34.0 in +3.00D group) versus baseline (scores: 62.2 in +2.50D group, 67.2 in +3.00D group) (P<0.005). Patients noted improvements in daily functioning with monovision correction, especially in double vision and social contact functional subscales of the ASQE. This improvement was offset by a trend towards an increase in disorientation and a decrease in distance estimation.

Conclusion:
Fifty percent of the subjects felt that monovision helped decrease or eliminate their symptoms of diplopia and they reported improved daily functioning and quality of life with monovision. Monovision correction may be a promising new treatment method in the management of small-angle diplopia, especially in patients who have failed conventional therapy.

References: None

Key Words: Diplopia, Monovision, Therapy, Strabismus, Quality of Life

Financial Disclosure: None
Introduction:
Idiopathic Intracranial Hypertension (IIH) is characterized by increased cerebrospinal fluid (CSF) pressure in the absence of common causes of intracranial hypertension. Due to its chronic nature and significant risk for visual loss, IIH patients should be seen by a physician on a regular basis. A recent review article stated (without source) that “newly diagnosed patients…may need to be reviewed every 2–4 weeks… (but) stable patients may be followed up every 3–6 months.”\(^{(1)}\) As to date, there has been no research done on how physicians are currently scheduling the frequency of these follow up appointments. In addition, management strategies for IIH, such as weight loss, medical therapy (acetazolamide) and surgery (ONSF), have been well documented\(^{(2,3)}\) but information on how physicians are currently dosing medication and/or recommending surgical intervention for the disease is sparse.

Methods:
A 35 question survey was designed to examine the criteria physicians are currently using to schedule follow-up appointments, dose medication, and recommend surgical procedures. The mailing list was generated through the AMA and the survey was sent to 1,172 fourth year ophthalmology or neurology residents. The main objective was to highlight areas where the treatment of IIH is significantly variable. Any observed variations would then become the subject of future research to determine the impact on patient outcomes.

Results:
The survey has been sent and analysis will be conducted in the next month after responses are collected.

Conclusion:
It is expected that survey responses will be gathered over the next month and analyzed for statistically relevant variations. Further data and conclusions will be available in November or December of this year. Regardless, this project will result in findings that will expand current understanding for how IIH is being treated and should thus enhance outcomes for patients.

References:
2. Digre, Idiopathic Intracranial Hypertension, Current Treatment Options in Neurology, 1, 74-81, 1999

Key Words: Idiopathic Intracranial Hypertension, Acetazolamide, Follow-Up Frequency

Financial Disclosure: None
Common and Distinctive Features of Inner Retinal Physiology and Encoding of Visual Information among Varied Forms of Retinal Degeneration

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Introduction:
Successful treatment of retinal degenerative diseases requires thorough understanding of how they disrupt the inner retina’s encoding of visual information. We previously demonstrated two cardinal electrophysiologic features shared by ganglion cells of several animal models of hereditary retinal degeneration: spontaneous hyperactivity and diminishing responsiveness to light. Here we show that both of these changes are common to various forms of retinal and neurodegenerative diseases, but demonstrate quantitative differences in stimulus-response relationships and information-carrying capacity.

Methods:
Multielectrode recordings of ganglion cells were made in the in vitro retina of mice with deficient retinoid recycling (Rpe65), phototransduction (Pde6-ß), ciliary transport (Cep290), or cell growth and axonal guidance (Tsc-2). Multiple spatiotemporal parameters were compared quantitatively among the strains to assess: dynamic properties of developmental waves; time course of emerging hyperactivity and diminishing light responses; frequency distribution and bursting dynamics of spontaneous activity; amplitude, threshold, transiency and duration of light-evoked responses; and Shannon mutual information.

Results:
Among these neurodegenerative disease models, normal early developmental “waves,” later sustained spontaneous hyperactivity, and diminishing light responsiveness were common features. However, distinctive patterns included: constant but supranormal firing rates in rd1 and rd10, periodic sudden elevations of spontaneous activity in Rpe-65 and Tsc-2, extremely slow waves in Cep-290, initially augmented light-evoked responses in rd10, preferential degradation of ON responses with narrowed dynamic range, and reduced information-carrying capacity in rd1.

Conclusion:
Large-scale changes in spontaneous and light-driven activity are common to many forms of retinal degenerative disease. However, detailed spatiotemporal features may 1) identify specific disorders, 2) suggest mechanisms underlying changes in inner retinal circuits, and 3) represent unique distortions of the neural code for the retina’s communication with the brain. We predict that these specific activity patterns will cause distinctive patterns of downstream visual system reorganization, and indicate optimal time windows for effective treatment of these diseases.

References: None

Key Words: Retinal Degeneration, Multielectrode, Ganglion Cell, Electrophysiology, Neural Encoding

Financial Disclosure: None
Retinitis Pigmentosa Sine Pigmento as an Unexplained Visual Field Loss

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Introduction:
We have evaluated a 33-year-old woman referred as being suspicious for glaucoma with an abnormal field in a preoperative exam for LASIK. We report a retinitis pigmentosa sine with asymptomatic unexplained field loss.

Methods:
A patient without subjective symptom was referred for constricted visual field. Complete ophthalmic examinations, color fundus photo, corneal thickness, refractive error, color vision, fluorescein angiography, optical coherence tomography, electroretinogram and visual field (C30-2) have been performed.

Results:
Examination revealed normal central vision and intraocular pressures. Color vision and retinal nerve fiber layer thickness were normal. Central corneal thickness was 501um and 519um in each eye. Perimacular thickness was decreased in both eyes. Humphrey Visual fields confirmed severe constriction in the central 30 degree for both eyes. Close fundus examination revealed the finding of a subtle greyish lesion along the macula and fluorescein angiography revealed window defect in accordance with retinal atrophic lesion in both eyes. The subtle greyish lesion showed complete correlation with constricted visual field. Peripheral retina was normal. Visual field was normal outside the central 30 degree. Electroretinogram revealed normal a wave and decreased b wave in rod response and maximal combined response.

Conclusion:
The diagnosis of retinitis pigmentosa sine pigmento from purely ophthalmoscopic finding is rather difficult. It could appear constricted field within central 30 degree without symptom. There were none of the pigmentary changes usually associated with retinitis pigmentosa. Close fundus exam and visual field outside 30 degree are necessary to find subtle defect in retinal epithelium.

References:

Key Words: Atypical Retinitis Pigmentosa, Constricted Central Visual Field, Suspicious Glaucomatous Visual Field, Electroretinogram, Normal Peripheral Visual Field

Financial Disclosure: None
Parinaud’s Syndrome Presenting with Asthenopia

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Introduction:
Parinaud’s syndrome, also known as dorsal midbrain syndrome is a group of abnormalities of eye movement and pupil dysfunction. It is caused by lesions of the upper brain stem. We have evaluated a 29-year-old man with asthenopia for 6 months. We report a Parinaud syndrome with asthenopia.

Methods:
We evaluated a patient with asthenopia for 6 months. Ophthalmic examinations, refractive error, alternate cover test, and ocular motility have been performed.

Results:
Best corrected visual acuity was 20/20 at distance and near in each eye. Fundus examination revealed incyclotorsion in left eye. He had 20° exophoria at near and 10° exophoria at distance in primary position. Intermittent left eye drifting was observed during convergence. Ocular motility showed unrecognized limitation in upgaze. He had no stereopsis and near reflex was normal. His pupils showed decreased light reflex. 0.125% pilocarpine test was negative. We performed brain MRI and 3*3.5*3cm mass invading around pineal gland and midbrain was found.

Conclusion:
The diagnosis of Parinaud’s syndrome without neurologic symptom is rather difficult. It could appear with asthenopia in young men. There was no sign of neurologic condition associated with parinaud’s syndrome. Ocular motility test necessary to find unrecognized limitation in certain gaze and confine the lesion.

References:

Key Words: Parinaud's Syndrome, Doesal Midbrain Syndrome, Astenopia

Financial Disclosure: None
Introduction:
Natural history of CCF was studied in 20 cases

Methods:
After a detailed neuro-ophthalmic work-up, the cases had undergone neuro-imaging and digital subtraction angiography to characterize the type and site of fistula. The cases with direct CCF were subjected to endovascular embolization and were followed up 3-monthly for a period of one year.

Results:
95% of our cases had direct CCF. Proptosis was seen to be the first presentation followed by ptosis, limitation of extraocular movements and loss of vision. Seventy-one percent of our patients with direct CCFs had successful balloon embolization. In the literature, successful closure of direct CCSFs has ranged from 58 to 100%. Two out of 19 cases had Superior ophthalmic vein thrombosis following embolization.

Conclusion:
The study highlights that increased experience in embolization techniques reduced the morbidity and mortality in CCF cases.

References: None

Key Words: Carotico-Cavernous Fistula, Proptosis, Ophthalmoplegia

Financial Disclosure: None
Poster 79

Magnetic Resonance Imaging as a Diagnostic Tool in Internuclear Ophthalmoplegia

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Introduction:
Lesions affecting the medial longitudinal fasciculus (MLF) cause internuclear ophthalmoplegia (INO). Magnetic resonance imaging (MRI) is the ideal modality for visualizing detailed structures of the brainstem. The purpose of this study was to determine the accuracy of radiologists in detecting MLF lesions in patients with clinically diagnosed INO based on MRI radiology reports.

Methods:
In this retrospective observational study, MRI reports of patients with an ICD-9 diagnosis of 378.86, INO, were reviewed from 2000-2010 from an outpatient neurology clinic and an inpatient service of a university hospital. A total of 22 subjects had 34 available MRI radiology reports. Reports were classified into “Definite,” “Possible,” and “Negative.” Reports which described lesions to the anatomic locality of the MLF were characterized as “Definite.” “Possible” reports described lesions in a broader defined localization. “Negative” meant lesions to the brainstem were not described in the report.

Results:
Of the 34 MRI reports, 9/34 (26.47%) were Definitive, 12/34 (35.29%) were Possible, and 13/34 (38.24%) were Negative. Of the 7 reports that provided ophthalmoplegia or an INO syndrome as the indication, 4/7 (57.14%) were Definite, 2/7 (28.57%) were Possible, and 1/7 (14.29%) were Negative. Of the remainder 27 reports, 5/27 (18.52%) were Definite, 10/27 (37.04%) were Possible, and 12/27 (44.44%) were Negative. Of the 12 reports read by general radiologists, 3/12 (25%) were Definite, 3/12 (25%) were Possible, and 6/12 (50%) were Negative. Of the 22 reports read by neuroradiologists, 6/22 (27.27%) were Definite, 9/22 (40.91%) were Possible, and 7/22 (31.82%) were Negative.

Conclusion:
In subjects with an INO, radiologists as a group less frequently reported MLF lesions on MRI unless ophthalmoplegia or an INO syndrome were provided as the indication for the studies. However, clinicians seldomly specified INO or ophthalmoplegia in the MRI requests. Neuroradiologists were more likely than general radiologists to report brainstem findings on MRI in subjects with an INO.

References: None

Key Words: Internuclear Ophthalmoplegia, Magnetic Resonance Imaging, Ocular Motility, Medical Imaging

Financial Disclosure: None
Intracranial Hypertension Caused by an Ependymoma of the Lumbar Thecal Sac with Normal Cerebrospinal Fluid Composition

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Introduction:
Tumors in the spinal canal are a rare but well-known cause of intracranial hypertension. Possible pathophysiologic mechanisms include disruption of CSF absorption at the arachnoid granulations due to high protein or degradation products in the CSF or blockage of cerebrospinal fluid (CSF) outflow from tumors.

Methods:
Case report

Results:
A 37-year-old woman presented with an 8-week-history of gradually progressive back pain, urinary hesitation, lower extremity weakness, headaches, blurred vision, intracranial noises and diplopia. Examination revealed normal visual function, bilateral optic disc edema, bilateral abduction limitation and paraparesis. Enhanced MRI of the brain and spine were unremarkable except for the presence of an intradural mass filling the thecal sac at the L3-L5 level. A CT-guided C1-C2 puncture revealed an elevated opening pressure of 31 cm of water with normal CSF protein and constituents. Acetazolamide treatment was discontinued 3 weeks later at the time of resection of the tumor, diagnosed as a myxopapillary ependymoma. One month post-operatively, her symptoms resolved and the examination normalized.

Conclusion:
The ependymoma in the lumbar thecal sac was the most likely cause of our patient's intracranial hypertension and clinical presentation. This case illustrates that tumors in the spinal canal may cause intracranial hypertension by mechanisms other than decreased CSF absorption at the arachnoid granulations due to high concentration of CSF protein or blood products. We propose that the most likely mechanism of intracranial hypertension in our case was either a mechanical obstruction of CSF flow into the spinal cul-de-sac or decreased absorption of CSF in the arachnoid villi adjacent to lumbar spinal nerve roots by the tumor itself; or a combination of both.

References:

Key Words: Intracranial Hypertension, Papilledema, Spinal Neoplasm

Financial Disclosure: None
Poster 81

The Use of Serum Glial Fibrillary Acidic Protein (GFAP) in the Diagnosis of Acute Isolated Optic Neuritis

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Introduction:
Cerebrospinal (CSF) Glial Fibrillary Acidic Protein (GFAP) has been shown to be a marker for CNS damage (1). Elevation of CSF GFAP has been shown in both multiple sclerosis (2) and Neuromyelitis Optica NMO (3). There are no studies to date which examine the role of serum GFAP in the context of inflammation within the central nervous system. This study suggests a potential role for the use of GFAP in diagnosing the aetiology of acute isolated optic neuritis, at first presentation.

Methods:
Serum samples of forty patients presenting either acutely (< 2 weeks) or late (<2 years) with optic neuritis were analysed for levels of GFAP by Enzyme-linked immunosorbent assay. These patients were subsequently diagnosed with Multiple Sclerosis (MSON), Recurrent Isolated Optic Neuritis (RION), Chronic Relapsing Inflammatory Optic Neuropathy (CRION) or Aquaporin IV positive optic neuritis (NMO). All patients presented with either their first or a subsequent episode of optic neuritis.

Results:
1. The serum GFAP values in patients presenting acutely with NMO related optic neuritis were significantly higher than in patients presenting acutely with MS related optic neuritis (P=0.016).
2. The serum GFAP values in patients presenting acutely with NMO related optic neuritis were significantly higher than in patients presenting late with NMO related optic neuritis (P=0.024).
3. There was no significant difference in the serum GFAP values between patients presenting acutely with CRION related optic neuritis and patients presenting acutely with MS related optic neuritis (P=0.985)

Conclusion:
This is the first known study looking at the role of serum GFAP in the diagnosis of neurological disease. We show that serum GFAP may be used to identify the aetiology of acute isolated optic neuritis.

References:

Key Words: Optic Neuritis, Glial Fibrillary Acidic Protein (GFAP), Neuromyelitis Optica (NMO), Multiple Sclerosis (MS)

Financial Disclosure: None
Poster 82

Does Lacrimal Gland Size on MRI Scans Differentiate Sarcoid From Demyelinating Optic Neuritis

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Introduction:
Sarcoid optic neuritis may be difficult to establish, especially in those without prior diagnosis or systemic manifestations. Since sarcoidosis frequently involves the lacrimal gland, which is spared in typical demyelinating optic neuritis, we examined whether lacrimal gland size on pretreatment MRI scan might differentiate sarcoid versus demyelinating optic neuritis.

Methods:
Eligible subjects were selected from a pool of patients from two neuro-ophthalmologists. Inclusion criteria were: age >18 years, diagnosis of optic neuritis, pre-treatment contrasted MRI scan with orbit/optic nerve views performed at one center on a 1.5 or 3 Tesla unit. Lacrimal glands were evaluated with axial and coronal views, fat-suppressed, Gadolinium-enhanced T1-weighted MRI images. One ophthalmologist, blinded to the patients' final diagnosis, measured the size of the glands on a PACS workstation. In each patient, only the scan with the largest lacrimal gland area was measured (area mm² = number of pixels x 0.0625 (mm²)). Patients with demyelinating optic neuritis were used as controls.

Results:
Six patients who were newly diagnosed with sarcoid optic neuritis met our inclusion criteria, had pretreatment MRI scans and strong clinical evidence supporting their diagnosis. Six age-and gender-matched patients were used for the control group. Average age in each group: 39 years. Average lacrimal gland size: 5.53 mm² (SD 1.49) in sarcoid group and 4.46 mm² (SD 1.13) in control group. P-value was 0.19.

Conclusion:
In our series, an enlarged lacrimal gland size on MRI scans suggested a final diagnosis of sarcoid as opposed to typical demyelinating optic neuritis. A larger sample size in a prospective study will be needed to determine if the observed difference is statistically significant.

References: None

Key Words: Sarcoid, Optic Neuritis, Lacrimal Gland

Financial Disclosure: None
Poster 83

Novel Treatment for Radiation Optic Neuropathy with IV Bevacizumab

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Introduction:
Radiation optic neuropathy is a devastating form of vision loss that is usually treated with steroids and hyperbaric oxygen. Generally the visual outcome is poor. There is evidence that Bevacizumab (Avastin) can reduce radiation necrosis by decreasing capillary leakage. We have experience where administration of IV Bevacizumab reversed the effects of radiation necrosis involving the optic nerves, chiasm and right temporal lobe in a 24 year old female patient. This is the first case report of this kind involving the visual pathways.

Methods:
Retrospective chart review of 3 patients with radiation necrosis of the visual pathway treated with hyperbaria compared to our 4th patient treated with IV Bevacizumab. The patient’s courses, MRI changes, and outcomes are compared and analysed. Literature review performed.

Results:
A 24-year-old female who underwent low-grade external beam radiation for treatment of a pilocytic astrocytoma and 5 years later developed bilateral radiation optic neuropathy with early radiation necrosis in the right temporal lobe. Our patient was treated with IV Bevacizumab (7.5 mg/kg/3-week schedule) and dexamethasone and showed marked recovery of her vision with this drug cocktail over a 3 month period. At 12 months post therapy she is stable with no recurrence of disease. Compared to the 3 other patients her outcome was markedly more favorable with actual reversal of visual loss, visual field loss and MRI changes. The therapy with IV Bevacizumab was also much less burdensome for the patient.

Conclusion:
We present a case of a patient with radiation optic neuropathy who responded dramatically to treatment with IV Bevacizumab, a VEGF inhibitor that decreases capillary leakage, thereby decreasing inflammation and swelling in tissues. IV Bevacizumab, commony used in Oncology, has not been used in cases of CNS radiation necrosis cases of the visual system primarily because of its novelty, but also because of medical insurance issues. We believe this to be the 1st case report of this type. This important clinical case will be presented and the presumed mechanism of action of Bevacizumab will be discussed. Bevacizumab should be considered as a first line of treatment in patients with acute vision loss from radiation induced optic neuropathy.

References:

Key Words: Radiation Necrosis, Pilocytic Astrocytoma, Glioma, Avastin, Novel Treatment

Financial Disclosure: None
Unilateral Proptosis Caused by Ocular Myasthenia Gravis

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Introduction:
Proptosis is a little-recognized finding in ocular myasthenia gravis, except when attributed to coexistent thyroid ophthalmopathy. Administration of edrophonium can temporarily reverse myasthenic deficits for diagnostic purposes.

Methods:
We describe the case of a previously healthy 76-year-old man, who presented with nonfluctuating horizontal binocular diplopia, partial right ptosis, and right proptosis, which had worsened steadily over the previous nine months. He denied any history of bulbar dysfunction, or proximal muscle weakness, and had no afferent neuro-ophthalmic complaints. His visual acuity was 20/25 OU. There were 2 mm of proptosis OD on Hertel exophthalmometry, and 5 mm of ptosis OD. Ocular motility examination revealed near-complete ophthalmoplegia OD and subtle dysmotility OS. Anteror segment, pupillary, and funduscopic examinations were unremarkable. Facial sensation and strength were intact. General neurological examination was unremarkable.

Contrast-enhanced MRI/MRA/MRV was notable only for right proptosis, with no direct or indirect evidence for dural AV fistula or carotid-cavernous fistula. Extraocular muscles were normal. Thyroid studies, VDRL, ACE, and thiamine levels were normal.

Results:
Hertel exophthalmometry was performed, and the measurements photographed, before and after the administration of edrophonium. Before edrophonium, exophthalmometry with a base of 106 mm was 19 mm OD and 16 mm OS. After edrophonium, exophthalmometry with a base of 106 mm was 17 mm OD and 16 mm OS. The patient’s ptosis improved by 3 mm OD, and ocular motility OD was also noted to improve with edrophonium. Acetylcholine receptor antibodies were absent. Single-fiber EMG of the orbicularis oculi revealed jitter or block in 13 of 31 fiber pairs, consistent with a diagnosis of ocular myasthenia gravis. The patient was started on Mestinon, and follow-up is pending.

Conclusion:
Ocular myasthenia gravis may cause proptosis, presumably through a reduction in extraocular muscle tone. Cholinesterase inhibitors may partially reverse this proptosis.

References:

Key Words: Myasthenia Gravis, Proptosis, Edrophonium

Financial Disclosure: None
Poster 85

Pupillary Tract Syndrome

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Introduction:
Afferent pupillary defect is a sine qua non of asymmetric optic nerve function. It may also occur with advanced retinal disease. Post chiasmal afferent pupillary defects have been recognized as a manifestation of optic tract involvement. When associated with visual field defects, a post chiasmal afferent still may be produced by involvement of the pupillary tract or midbrain. These have been felt to be rare.

Methods:
Retrospective chart review of patients seen at the University of Virginia neuro-ophthalmology service between 1985 and 2008. Patients identified as having presumed pupillary tract were identified and characterized.

Results:
Forty charts with suspected pupillary tract defects were examined with 22 patients exhibiting an APD with no homonymous hemianopsia on the same side and 2 patients with either an unusable visual field or no visual field test. Of the 16 patients with pupillary tract defects, seven had mass lesions consisting of the following: a parietal mass, a left parenchymal mass, a thalamic mass, a midbrain tumor, two pineal gland tumors, and one probable glioma in the pretectal area. The remaining patients were found to have either a stroke, aneurysm, or hemorrhage causing their lesion including a midbrain infarct, thalamic infarct, thalamic hemorrhage, basilar tip aneurysm, and arteriovenous malformation. The ischemic strokes consisted of the following lesions: two instances of midbrain infarct, one instances of thalamic infarcts, and an occipital infarct. Two patients also had basilar tip aneurysms, one patient had an AVM, and one patient presented after a posterior fossa craniotomy. Furthermore, one patient presented after a thalamic hemorrhage.

Conclusion:
Pupillary tract and midbrain afferent pupillary defects are not rare. They may be an unrecognized cause of small afferent pupillary defects that are otherwise discounted. In patients with midbrain pathology, careful assessment for subtle asymmetric pupillary response with filter stress may confirm pupillary pathway abnormalities. Similarly subtle APDs may be due to unrecognized midbrain pathology.

References: None

Key Words: Pupil, Afferent Pupillary Defect, Midbrain Pathology, Visual Fields, OCT

Financial Disclosure: None
Poster 86

The Prognostic Factors of Recovery of Visual Field Defect and Visual Loss after Operation for Pituitary Adenoma

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Introduction:
Pituitary adenoma is one of the most common brain tumor in adults. The symptoms of pituitary adenoma is mainly visual associated symptoms such as visual field defect and decreased visual acuity. However the results of the surgery for pituitary adenoma were different. So, we investigated the prognostic factors of visual recovery after the surgery.

Methods:
The patients for pituitary adenoma who undertaken surgery from 2006 to 2008 were enrolled in this study. Ophthalmic evaluation was comprised of best corrected visual acuity, visual field examination using Goldmann perimeter. The size of tumor before the surgery was determined in each case. The histology of tumor was also investigated. The time of onset of visual symptom to surgery was asked in each person

Results:
One hundred sixty-nine eyes of eight four patients were enrolled in this study. The mean visual acuity before the surgery was 0.310 logMAR. However the mean visual acuity six months after the surgery was 0.180 logMAR (P<0.05). The most common pattern of visual field defect was bitemporal hemianopsia and there was improvement of visual field defect after the surgery. The small size of the tumor, better visual acuity before the surgery and larger visual field defect had relationship of good prognostic factors in this study.

Conclusion:
There were significant improvement of visual field defect and visual acuity. The prognostic factors in this study were the size of the tumors

References: None

Key Words: Pituitary Adenoma, Visual Field, Visual Acuity

Financial Disclosure: None
Use of Macular Optical Coherence Tomography (OCT) to Detect Homonymous Hemianopia

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Introduction:
Optic tract syndrome is classically described with optic atrophy in a “bowtie” pattern. OCT of the retinal nerve fiber layer (RNFL) has been reported for optic tract syndrome, but most cases do not show a bow-tie pattern. We investigated whether OCT of the macula may be a more useful method for detecting optic tract syndrome.

Methods:
15 subjects were identified with homonymous hemianopia and who also had macular and RNFL spectral domain OCT scans that were part of a routine protocol in our neuro-ophthalmology clinic. Based on a careful review of neuroimaging studies, patients were categorized as having a likely lesion of the optic tract, a possible lesion of the optic tract, or a definite retro-geniculate lesion. Macula OCT scans were evaluated subjectively and quantitatively by comparing the ratio of the temporal retinal thickness in the intact and the hemianopic retina and also for the nasal retina.

Results:
11/15 patients had possible or likely optic tract lesions based on imaging. None of the patient had a clear pattern of “bow tie” atrophy on either fundus photography or OCT of the RNFL. 6 of the 11 patients had a subjective appearance of homonymous retinal thinning based on the macular OCT and correlated with a ratio of 1.1 or greater comparing the “intact” retina with hemianopic retina.

Conclusion:
OCT of the macula is an effective tool for evaluating retinal ganglion cell and axon loss from optic tract lesions. Further evaluation is being conducted to investigate the appearance of macular OCT in retrogeniculate lesions.

References:

Key Words: Macula OCT, Hemianopia

Financial Disclosure: None
**Poster 88**

**Ultra-High Resolution Optical Coherence Tomography for Imaging Retinal Nerve Fiber Layer in Multiple Sclerosis**

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**Introduction:**

**Purpose:** To determine the feasibility of imaging the retinal nerve fiber layer (RNFL) in multiple sclerosis (MS) patients with an ultra-high resolution optical coherence tomography (UHR-OCT) with improved image resolution.

**Methods:**

A slit-lamp based UHR-OCT was custom developed with advanced optical delivery system for retinal imaging. The system has a 3μm depth resolution and a computer-controlled fixation target. The scan speed was set to 24K A-scan per second and the field of the scan was set to about 20 degrees. A commercially available retinal spectral domain optical coherence tomography (SD-OCT, Cirrus, Carl Zeiss Meditec) was also used. Three clinically diagnosed MS patients (3 females, age 44 ± 5 yrs) and 3 normal subjects (1 female and 2 males, age 35 ± 12) were recruited and imaged. The thickness at the temporal side was recorded from the RNFL thickness scan report in SD-OCT. Semi-manual method was used to obtain the RNFL thickness at location of the temporal side half optic disk diameter in the UHR-OCT images. Both eyes were scanned at the same visit.

**Results:**

The RNFL thickness acquired with SD-OCT was 46.5 ± 8.0 μm in MS patients, which was significantly thinner than that with normal controls (57.8 ± 6.0 μm, P<0.05). Acquired with UHR-OCT, the RNFL was 51.7 ± 13.2 μm in MS patients which was significantly thinner that normal controls (63.8 ± 4.9 μm, P < 0.05). The correlation between two OCT devices in the measurement was 0.75.

**Conclusion:**

In this pilot study, we have demonstrated the feasibility using our UHR-OCT for imaging the RNFL, which appeared compatible to the commercial OCT. Further improvement will be the development of more robust software for segmental analysis and 3D rendering. The test of sensitivity and specificity of the prototype will be also needed with a large scale of sample size.

**References:** None

**Key Words:** Ultra-High Resolution Optic Coherence Tomography, Retinal Nerve Fiber Layer, Multiple Sclerosis

**Financial Disclosure:** None
High-Resolution Susceptibility-Weighted Imaging Findings in Patients with Swollen Optic Nerves

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Introduction:
Susceptibility-weighted imaging (SWI) is a new 3D-reconstructed gradient echo sequence that is highly sensitive for the paramagnetic effect of deoxyhemoglobin serving as an intrinsic contrast agent of cerebral veins. The most frequent cause of symmetrically swollen optic nerves is idiopathic intracranial hypertension, the cause of which is still unknown but thought to be related to the cerebral venous system.

The authors hypothesized that the highly sensitive SWI will reveal abnormalities suggestive of cerebral venous dysfunction in patients with IIH and pseudotumor cerebri.

Methods:
We performed a prospective study of 10 consecutive patients undergoing MRI of brain as part of the diagnostic evaluation for swollen optic nerves. All image data sets were reviewed by two experienced neuroimagers.

Results:
The authors describe the distinct SWI neuroimaging findings in patients with swollen optic nerves.

Conclusion:
SWI provides additional information in the complex diagnostic evaluation of patients with swollen optic nerves, and therefore should be included in the neuroimaging evaluation of patients with various causes of optic nerve swelling.

References: None

Key Words: Susceptibility-Weighted Sequence, Swollen Optic Nerve, Idiopathic Intracranial Hypertension, Cerebral Venous System, 3D Reformatted Gradient Echo Sequence

Financial Disclosure: None
A Case of Disseminated Choroid Plexus Papilloma with Parasellar Extension with Bilateral Papilledema in a Visually Asymptomatic Male

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Introduction:
Choroid plexus papillomas are rare tumors of neurectoderm origin more commonly seen in children arising in the lateral ventricle. In adults, the tumors account for 0.5% of intracranial neoplasm and most commonly arise within the third ventricle. Dissemination has rarely been reported (<25 cases) and among those cases, only a few noted parasellar involvement. Usually those cases with parasellar involvement report visual symptoms, such as diplopia or visual loss.

Methods:
Case report.

Results:
A 44 year old man with hypertension and bipolar disorder presented with 3 months of progressive bilateral hearing loss, vertigo and headaches. MRI of the brain revealed nodular enhancement along the course of cranial nerves five, seven and eight, within the cisterns, in the right parasellar region, and along the surface of the inferior cerebellar vermis and in the left side of the foramen magnum. MRI of the spine demonstrated extensive intradural nodular enhancement at multiple levels. Ophthalmologic exam revealed 20/20 vision OU, no APD, no EOM disturbance and a normal anterior segment exam. Dilated fundus exam revealed florid papilledema bilaterally, disc hemorrhages bilaterally and intraretinal hemorrhages along the left supero-temporal arcade. SITA Fast Humphrey visual field 24-2 was full OU. Orbital MRI demonstrated a parasellar enhancing mass which encased and superiorly displaced the right optic chiasm and right optic nerve and extended into both proximal optic canals. Surgical pathology revealed a choroid plexus papilloma, atypical, WHO grade II.

Conclusion:
Rarely, choroid plexus papilloma may have parasellar dissemination resulting in optic nerve pathology which does not produce visual symptoms.

References:

Key Words: Choroid Plexus Papilloma, Leptomeningeal Seeding, Dissemination, Parasellar, Papilledema

Financial Disclosure: None
Ocular Lateropulsion but not Radiographic Horizontal Gaze Deviation is a Predictor of Stroke in Patients with Acute Vestibular Syndrome

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Introduction:
Acute vestibular syndrome (AVS) usually results from vestibular neuritis or posterior fossa stroke. Horizontal conjugate gaze deviation (h-CGD) results from right-left bias in sensorimotor inputs to the oculomotor system, including vestibular lesions. It can be identified radiographically or clinically. Ocular lateropulsion (OL) is h-CGD in the upright position evident clinically after brief eyelid closure and has most often been described after lateral brainstem infarction. The diagnostic utility of different types of h-CGD for predicting central versus peripheral causes of AVS is unknown.

Methods:
We assessed h-CGD as part of an ongoing single-center study of AVS diagnosis. Radiographic h-CGD was defined as the angle between a line bisecting the lens in each eye and a line perpendicular to the naso-occipital axis by axial MRI obtained with the patient supine and eyes closed. We compared this with bedside OL (only in more recently enrolled patients, due to a change in the data collection protocol). Final lesion localization was based on MRI DWI or negative imaging plus clinical follow-up for those with vestibular neuritis.

Results:
Among 135 AVS patients (85 posterior fossa ischemic strokes, 34 vestibular neuritis, 16 other [15 central]), radiographic h-CGD was assessed in 117 and OL in 47. All patients were able to maintain the primary gaze position with eyes open. Radiographic h-CGD was present in 50% (n=58/117) and mean h-CGD was 23 degrees. Radiographic h-CGD was more frequent in peripheral than central lesions (68%, n=23/34 vs. 42%, n=35/83, $\chi^2$ p =0.01) while OL was found only in those with central pathology (52%, n=17/33 vs. 0%, n=0/14, Fisher’s exact p<0.001).

Conclusion:
Our study demonstrates that radiographic h-CGD occurs frequently in patients with peripheral as well as central causes of AVS, but that OL is seen only with central pathology, making its presence a strong sign of stroke in AVS.

References: None

Key Words: Differentiating Peripheral Versus Central Brainstem And Cerebellar Lesions, Ocular Lateropulsion, Radiographic Horizontal Gaze Deviation, Eye Examination, Acute Vestibular Syndrome

Financial Disclosure: None
Detection of Change in Papilledema Over Time: Comparison of Optical Coherence Tomography (OCT) with Digital Optic Nerve Photographs

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Introduction:
Detection of change in optic nerve appearance and function between clinical visits guide medical and surgical treatment decisions in patients with papilledema. However, there has been no study to date that has evaluated and compared methods of assessing papilledema over time. This study compared expert analysis of optic disc photographs with OCT.

Methods:
22 patients were studied on at least 2 clinic visits who had papilledema of at least Frisen Grade 2 and who had stereo fundus photography, spectral domain OCT (Cirrus) of the optic nerve, and visual field testing performed on each visit. Three neuro-ophthalmology experts independently classified the change between 2 visits as either no change, improvement, or worsening for optic nerve photographs (assessed monocular and stereo photos independently). A similar assessment was made from the OCT graphic report printout. A majority rule was used to reconcile any differences among experts for each assessment.

Results:
The greatest agreement was between stereo fundus photography and OCT; 88% of the assessments agreed and 12% were in disagreement (5% showed no change on analysis of stereo photographs, but OCT showed worsening; 7% showed improvement by stereo photo assessment but no change on OCT). Comparison of monocular digital photos with OCT showed agreement in 84% of assessments as did comparison of stereo with mono photo assessment.

Conclusion:
Although there was relative agreement between subjective assessments of change in disc appearance and OCT by experts, there were a significant number of cases where clinical disagreements occurred that might influence treatment decisions. We are currently analyzing OCT scans quantitatively and determining whether disagreements between modalities may influence treatment decisions.

References: None

Key Words: Papilledema, Optical Coherence Tomography (OCT), Digital Optic Nerve Photography

Financial Disclosure: None
What Terminates Saccades: Brainstem Omnipause Neurons or Cerebellar Fastigial Nucleus?

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Introduction:
During horizontal saccades, the agonist extraocular muscle contracts vigorously, driven by motoneurons receiving a pulse of innervation from ipsilateral pontine excitatory burst neurons (EBN). Simultaneously, the antagonist muscle relaxes because of a pause in motoneuron innervation due to inputs from contralateral medullary inhibitory burst neurons (IBN). Except during saccades, BN are tonically inhibited by pontine omnipause neurons (OPN). Before saccade onset, OPN cease discharge; just before saccade end, OPN recommence BN tonic inhibition. But, do OPN control saccade end? OPN pause duration does not correlate with saccade duration, and saccades are normometric after pontine lesions. Thus, it has been postulated that the cerebellar caudal fastigial nucleus (cFN) ends the saccade by firing ipsilateral IBN.

Methods:
We tested whether OPN were responsible for stopping saccades by studying gaze shifts between two targets aligned on the visual axis of one eye; such movements consist of a disjunctive saccade followed by a vergence movement. Prior studies have indicated that during the vergence movement, small, high-frequency conjugate oscillations occur, which are probably saccadic in origin, indicating that OPN are silent. We studied: (1) Healthy humans; (2) Late-onset Tay-Sachs disease (LOTS) patients, with premature saccade termination; (3) Cerebellar ataxia (SCASI) patients with pronounced saccadic hypermetria, indicating late termination of saccades.

Results:
Conjugate oscillations occurred in all groups at a time when the saccade was terminated and continued during the subsequent vergence.

Conclusion:
This evidence suggests that the cFN dictates saccade termination time, even when cerebellar disease causes saccades to become hypometric (as in LOTS) or hypermetric (as in SCASI). OPN appear to be more concerned with sustained inhibition of BN during fixation, and inducing their brisk discharge at the onset of saccades.

References: None

Key Words: Saccades, Omnipause Neurons, Burst Neurons, Fastigial Nucleus

Financial Disclosure: None
Convergence Insufficiency Responsive to Chronic Bilateral Subthalamic Nucleus Deep Brain Simulation in Parkinson’s Disease

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Introduction:
Chronic subthalamic nucleus (STN) deep brain stimulation (DBS) improves motor symptoms and decreases levodopa-related motor complications in Parkinson’s disease (PD). STN-DBS may cause ocular motor abnormalities in intraoperative and chronic states, including contraversive eye deviation and the ocular tilt reaction. In contrast, beneficial effects of STN-DBS on ocular motility have not been studied. Convergence insufficiency (CI) is a well-established cause of visual complaints in PD. While cases of levodopa-responsive CI are reported, many patients remain symptomatic despite levodopa and prism therapy.

Methods:
We report a patient with CI responsive to STN-DBS.

Results:
A 71 year-old man with a 12 year history of PD had long-standing binocular horizontal diplopia with reading due to CI, despite levodopa and prism therapy. Upon evaluation for vertical diplopia at distance following bilateral STN-DBS placement, he was no longer symptomatic of CI. Exam was performed in four states: (1) levodopa off/DBS on; (2) levodopa off/DBS off; (3) levodopa on/DBS on; (4) levodopa on/DBS off. In all, ocular motor range to a distant target was full and a vertical misalignment suggestive of alternating skew deviation on lateral gaze (ASD) was present. In states 1 and 3, convergence was nearly full with no diplopia, a near point of convergence (NPC) within 2 cm, and a 25 PD XT at near. In states 2 and 4, convergence was impaired with diplopia at near, a remote NPC, and a 40-45 PD XT at near.

Conclusion:
We report a patient with CI responsive to STN-DBS. The etiology of skew deviation is unclear, as ASD is typically associated with cerebellar disease. Skew deviation is reported in association with STN-DBS and it is feasible that is was an ocular motor complication of DBS in this patient. Elimination of symptomatic CI in this patient suggests that the effect of STN-DBS on CI deserves further study.

References: None

Key Words: Parkinson's Disease, Subthalamic Nucleus Deep Brain Stimulation, Convergence Insufficiency, Skew Deviation

Financial Disclosure: None
Introduction:
A 22-year old woman with a low-normal Vitamin B₁₂ level, mildly-elevated methylmalonic acid (MMA) level, and a normal homocysteine (HCys) level, presented with slowly progressive, painless visual loss over three years. OCT scans showed severe, bilateral, temporal thinning, and Visual Evoked Potentials (VEPs) indicated prolonged bilateral latencies.

Methods:
Case report; Neurology outpatient clinic.

Results:
Intramuscular Vitamin B₁₂ injections resulted in complete visual recovery.

Conclusion:
Painless, progressive bilateral optic neuropathy poses a clinical diagnostic challenge. Serum levels of B vitamins are frequently evaluated but judged to be “normal” if they fall within laboratory-specified reference ranges, without consideration of laboratory and biologic variability. Aside from the issues of laboratory reproducibility and intra-individual variation, some patients respond to Vitamin B₁₂ therapy even when pre-treatment levels of B₁₂, MMA and HCys are normal, whereas other patients who have significant neurologic or hematologic abnormalities together with low B₁₂ and high MMA and HCys levels do not respond to B₁₂ therapy.

This case demonstrates for the first time that OCT can be used to document structural optic nerve damage in optic neuropathy due to Vitamin B₁₂ deficiency. Whereas a patient’s electrophysiology and functional vision (including acuity, color vision, and contrast vision) results may improve to normal with treatment, OCT, which measures the retinal nerve fiber layer (RNFL), reveals permanent damage.

Although only .5% of Vitamin B₁₂ deficient patients have reported visual impairment, OCT imaging may also show that subclinical visual impairment is more common than previously thought, and OCT may be a valuable tool in helping diagnose B₁₂ deficient patients earlier. This report emphasizes the importance of setting a low threshold for Vitamin B₁₂ treatment in the appropriate clinical setting, despite a work-up that is negative using traditional criteria.

References:

Key Words: Low Threshold for Diagnosis of Vitamin B₁₂ Deficiency, Optic Neuropathy, Optical Coherence Tomography (OCT) as a Diagnostic Tool, Complete Visual Recovery, Reliability of Biomarkers in Detecting Vitamin B₁₂ Deficiency

Financial Disclosure: None
Use of the Optical Coherence Tomography in Predicting Post-Treatment Visual Outcome in Anterior Visual Pathway Meningiomas

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Introduction:
Optical coherence tomography (OCT) has been documented to reliably predict post-operative visual outcome by assessing the pre-operative peripapillary retinal nerve fiber layer (PRNFL) thickness in patients with compressive optic neuropathy secondary to pituitary adenomas. The aim of this study was to determine if OCT has the same prognostic value for anterior pathway meningiomas.

Methods:
This was a retrospective review of patients with anterior pathway meningiomas. The following pre-treatment data was collected: age, duration of symptoms, visual acuity, colour vision, visual field sensitivity as measured by static perimetry and average PRNFL thickness. The findings were compared with data from the last follow-up visit after treatment.

Results:
We defined thin PRNFL as at less than 95% percentile of the age-matched population. Twelve patients and 13 eyes were analyzed. Ten eyes had normal PRNFL thickness (mean: 94.39 microns +/- 18.26), whereas three eyes had thin PRNFL (mean 61.85 microns +/- 13.74). The mean duration of follow-up was 9.7 months. There was no significant difference in age, duration of symptoms or duration of follow-up between both groups (p = 0.6). After treatment, the normal PRNFL group experienced significant improvement in the visual acuity (p < 0.01), color vision (p < 0.02), mean deviation (p < 0.003) and pattern standard deviation (p < 0.01) but not foveal threshold (p = 0.06) compared with the group with thin PRNFL. Using multivariate analysis, we found no effect of age, duration of symptoms or follow-up duration on final visual outcome.

Conclusion:
Patients with compressive optic neuropathy due to anterior pathway meningiomas are more likely to improve post-treatment if they have a normal pre-treatment PRNFL.

References:

Key Words: Anterior Visual Pathway Meningioma, Retinal Nerve Fibre Layer Thickness, Optical Coherence Tomography, Visual Outcome, Prognosis

Financial Disclosure: None
**Poster 97**

**Fundoscopy on an Inpatient Neurology Service: An Observational Study**

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**Introduction:**
There is consensus among neurologists that fundoscopy is an essential part of the complete neurological examination. In theory, fundoscopy allows identification of disc swelling, vascular changes or retinal pathology that may have significant implications on patient management and outcome. The purpose of this observational study was to assess whether fundoscopy was being used effectively on a Neurology service.

**Methods:**
50 consecutive patients admitted to our General Neurology Service received an additional, separate fundus evaluation by the Ophthalmology Consult Service. Documentation of the fundus examinations from the initial Neurology admission note and Neurology progress notes were compared to the fundus findings documented by Ophthalmology.

**Results:**
A successful fundus examination was documented in only 52% of patients admitted to Neurology. The most common reasons cited for failure were "miotic pupil", "inability to visualize", or "uncooperative patient". In no cases were pupils dilated by Neurology (though in 10% of cases, patients were examined after they had been examined by Ophthalmology). When documented, the fundus findings reported by Neurology were generally limited to statements about the optic disc such as, "sharp disc margins" or "no papilledema". In less than 5% of cases was there any mention of other elements of the fundus examination. In contrast, the additional Ophthalmology evaluations yielded significant ocular findings in 60% of patients examined, including significantly increased cup-to-disc ratios, optic disc swelling, optic disc pallor, cotton wool spots, and attenuated retinal vessels.

**Conclusion:**
Our results suggest that thorough funduscopic examinations are not being performed by Neurology at our institution. When Ophthalmology is not involved, pupil dilation rarely occurs and findings seem to be limited to visualization of the disc margin. Unless these weaknesses are unique to our service or to our institution, this suggests that, without additional training, retinal pathology may be frequently missed by non-Ophthalmologists.

**References:**

**Key Words:** Fundoscopic Exam, Neurology, Inpatient

**Financial Disclosure:** None
Visual Outcome of Unaffected Eye in Idiopathic Intracranial Hypertension with Unilateral Papilledema

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Introduction:
Papilledema is the funduscopic hallmark of Idiopathic intracranial hypertension (IIH) and one of the diagnostic criteria [1]. Atypical cases of IIH in which papilledema is markedly asymmetric, unilateral, or even absent have been reported, and patients with IIH who developed visual loss in absence of papilledema have been documented [2-3]. In this study, we evaluated the long-term visual outcome of the eye without disc edema on presentation in “unilateral” cases of IIH.

Methods:
Retrospective chart review of all patients examined in neuro-ophthalmology division from 1989 to 2009 with a diagnosis of idiopathic intracranial hypertension.

Results:
Three IIH cases with unilateral optic disc edema at presentation were identified. Their opening pressures were 34, 37, and 35 cm water. Follow-up of these patients continued for a period of 3, 5, and 11 years respectively. Initial best corrected visual acuity of the affected eye was 20/20, 20/30, 20/25; while in the unaffected eye it was 20/20, 20/25, 20/20. Fundus examination showed optic disc edema of the affected eyes with sharp pink discs in the other eyes. Initial visual fields showed trace nasal step, enlarged blind spot with superior arcuate scotoma, and inferior nasal step of the affected eyes. In all three unaffected eyes, initial visual fields were full. On follow-up, one patient never developed disc edema in the other eye. The other two patients developed slight disc edema, one after 9 months and the other after 7 weeks of presentation. The final visual acuity of the three patients remained 20/20 in the initially unaffected eye. Final visual acuity in the affected eyes were 20/20, NLP, 20/20. Visual fields were full, NLP, and trace nasal step in the affected eyes. In the unaffected eyes, visual fields remained normal in two and the last developed superior and inferior arcuate scotoma.

Unilateral papilledema with IIH is an infrequent finding. In our small series, the clinical course and outcome in the initially unaffected eye was relatively benign.

References:

Key Words: Idiopathic Intracranial Hypertension, Pseudotumour Cerebri, Unilateral Papilledema, Visual Outcome

Financial Disclosure: None
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**Visuospatial Assessment by Virtual 3D Radial Optic Flow Illusion in Healthy Aging, Mild Alzheimer’s Disease (AD) and Pre-Deep-Brain Stimulation (DBS) Parkinson’s Disease (PD) Subjects**

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**Introduction:**
Optic flow (the pattern of motion at the eye) controls human walking. Perception of the focus of expansion (FOE) of a radially expanding pattern of moving dots (radial optic flow or ROF) is impaired in AD, but has not been investigated in PD. In the ROF illusion (ROFI), the FOE is perceived as shifted in the direction of a superimposed planar motion pattern (similar motion created by pursuit eye movements). The magnitude of the shift is dependent on the depth of the planar motion: decreased significantly for crossed disparity (in front) and nearly unchanged for uncrossed disparity (behind). Effects of aging and disease on the ROFI are unknown. We assessed perception of the ROFI in healthy older, AD, and pre-DBS-PD subjects.

**Methods:**
Healthy (n=26), mild AD (n=13), and non-demented, pre-DBS-PD (n=7) subjects with good stereoacuity participated. The ROFI was displayed in a 3D virtual environment with planar motion at 3 depths: same(0°), front(+1.2°), or behind(-1.5°).

**Results:**
AD was more likely than PD* and healthy* to not perceive an illusory shift in the FOE. A significant effect of depth was observed for healthy* and PD*, but not AD. Unlike healthy and published reports for young subjects, the magnitude of FOE shift for PD subjects was not significantly different between crossed and uncrossed disparities. (*p<0.05)

**Conclusion:**
This study indicates that AD and PD could potentially disrupt visually-driven neuronal networks involved in the control of human walking and ROFI processing in AD and PD warrants further study.

**References:**

**Key Words:** Alzheimer's Disease, Parkinson's Disease, Visuospatial Perception, Optic Flow, Human Walking

**Financial Disclosure:** None
An Inferior, Macular Sparing, Homonymous Quadrantanopia Associated with Segmental Optic Nerve Hypoplasia and Isolated, Unilateral, Anterior Medial Occipital Lobe Atrophy in an Asymptomatic Adult

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Introduction:
Bilateral hemioptic optic nerve hypoplasia has been associated with congenital hemiplegia, hemianopia and cortical atrophy. The presumptive cause of the hypoplasia in these cases is retrograde, trans-synaptic retinal ganglion axonal degeneration. Acquired retrogeniculate visual pathway lesions can also result in retinal nerve fiber layer loss due to trans-synaptic degeneration as noted in experimentally-induced lesions in primates and by observation and RNFL analysis in humans.

Methods:
We report the association of a macular sparing, right inferior quadrantanopia with bilateral superior segmental optic nerve hypoplasia and isolated left anterior medial occipital lobe atrophy in an asymptomatic adult.

Results:
A 39-year-old right handed woman was referred for neuro-ophthalmic evaluation after a screening visual field test revealed a right inferior quadrantanopia with macular sparing. Examination revealed 20/20 VA OU with normal color vision and a trace right APD. Bilateral, segmental superior optic nerve hypoplasia was noted and RNFL analysis revealed decreased thickness of the superior optic nerves with nasal (OD and some OS) and temporal (OS) NFL decrease. Brain MRI showed increased size of the left posterior horn of the lateral ventricle with decreased anterior medial occipital cortical thickness.

Conclusion:
Segmental bilateral optic nerve hypoplasia in our patient was associated with a homonymous, macular sparing, quadrantanopia and isolated, anterior medial occipital lobe atrophy. The lack of abnormal T2 signal in the occipital lobe argues for an isolated, prenatal insult to the left occipital lobe and supports trans-synaptic optic nerve degeneration as a mechanistic cause for segmental optic nerve hypoplasia. Until now, a homonymous quadrantanopia has not been previously described in association with segmental optic nerve hypoplasia and isolated, unilateral occipital lobe atrophy in an asymptomatic adult.

References: None

Key Words: Segmental Optic Nerve Hypoplasia, Homonymous Quadrantanopia, Macular Sparing, Anterior Medical Occipital Lobe Atrophy

Financial Disclosure: None
Acute Elevated Intracranial Pressure (ICP) Syndrome Associated with Unilateral Transverse/Sigmoid Dural Vein Thrombosis with Rapid Resolution Following Clot Lysis by tPA, Angiojet, and Balloon Angioplasty

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Introduction:
A 28-year-old woman presented with acute headache, diplopia, and intracranial noises for several days. MRI/V Brain revealed potential thrombosis of the right transverse/sigmoid sinus. Warfarin treatment was initiated. She was referred 3 weeks later for persistent diplopia and headache. On exam, VA was 20/20 OU, VF were normal (tangent screen), and bilateral abduction deficits (-3) and severe papilledema was present. RNFL OCT thickness was 553/590 microns OD/OS.

Methods:
LP opening pressure was 50 cmH20. Cerebral angiography revealed minimal flow within the right transverse/sigmoid sinus with right torcula pressure of 35 mmHg and lower right sigmoid sinus pressure of 9 mmHg. Lysis of the clot by tPA, angiojet, and balloon angioplasty was performed followed by 48 hour tPA drip. Repeat angiography the next day revealed good flow through the right transverse/sigmoid sinus with right lower sigmoid pressures of 13-14 mmHg and torcula pressures of 22-25 mmHg.

Results:
The patient’s headache syndrome improved within a few hours of the procedure. Two weeks post-lysis, LP opening pressure was 16 cmH20 and papilledema was mild-moderate (RNFL 247/301 microns OD/OS). Three weeks post-lysis, papilledema was nearly resolved (RNFL 147/177 microns OD/OS) and minimal residual bilateral abduction deficits were present. Follow-up VA and VF were normal. The patient is heterozygous for a prothrombin mutation without other risk factors and is maintained on Warfarin.

Conclusion:
Rapid resolution of this patient’s syndrome demonstrates that a severe elevated ICP syndrome can result from unilateral transverse/sigmoid sinus thrombosis and early intravenous lysis can provide rapid relief and prevent vision loss. Consideration for cerebral angiography with intradural lytic treatment should be made in cases of acute, unilateral thrombosis of the transverse sinus associated with elevated intracranial pressure syndromes.

References: None

Key Words: Unilateral Dural Vein Thrombosis, Papilledema, Headache, Clot Lysis, TPA

Financial Disclosure: None
Superior Segmental Optic Nerve Hypoplasia - A mfVEP and Frequency Domain OCT Study

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**Introduction:**
Superior segmental optic nerve hypoplasia (SSOH) is a developmental disorder characterised by a relative hypoplasia of the superior portion of the optic disc. Patients with SSOH usually have an inferior altitudinal or sector-like visual field defect. Maternal type 1 diabetes may be a risk factor for development of SSOH. Recently, several studies have looked at the optical coherence tomography (OCT) characteristics of SSOH.

We describe a patient with SSOH with the results of multifocal visual evoked potentials (mfVEP) and optical coherence tomography (OCT) testing.

**Methods:**
Observational case report with review of the visual field, mfVEP and SD-OCT testing.

**Results:**
A 54 year-old woman with diabetes mellitus (DM), hypertension and maternal DM was referred for neuro-ophthalmic evaluation for bilateral inferior visual field loss after a work-up failed to identify an etiology.

Examination demonstrated corrected visual acuities of 20/20 OU with normal color vision and pupillary responses. Ophthalmoscopy showed superior hypoplasia of the discs corresponding to bilateral inferior altitudional defects seen on automated perimetry. The multifocal ERG was normal. Testing with mfVEP revealed reduced amplitude with normal latencies. Stratus OCT and frequency domain OCT demonstrated superior RNFL thinning.

**Conclusion:**
A 54 year-old woman underwent an extensive work-up that included an MRI for bilateral visual field loss. Electrophysiology testing confirmed optic nerve origin. Careful evaluation of the optic discs revealed superior segmental optic nerve hypoplasia in a patient with maternal DM. Awareness of this condition can help avoid unnecessary ancillary testing.

**References:**

**Key Words:** Optic Disc Hypoplasia, OCT, mfVEP

**Financial Disclosure:** None
Introduction:
Kyrieleis’ phenomena, retinal peri-arterial plaques, is usually associated with ocular toxoplasmosis, but may be seen in syphilis, HIV infection, herpes, and tuberculosis. Kyrieleis plaques can mimic multiple emboli, particularly after the uveitis resolves or its history is unappreciated.

Methods:
Case report.

Results:
A fifty-seven year-old Dominican female presented to us with chronic irritation and blur in both eyes, with history of right eye infection one year ago, treated with ten days of oral penicillin and steroid injection. One year prior she had been treated for primary syphilis. General eye and neuro-ophthalmological exams were normal including slit lamp with tensions and gonioscopy. Pupils were unequal, 6mm OD with trace sectoral response to direct and accommodative targets in the 4:00 position, OS 2.5mm with brisk direct and consensual responses, no afferent defect. Dilated funduscoppy OD revealed clear media, multiple yellow peri-arterial plaques not limited to branch points, and an old chorioretinal scar in the mid-periphery. The left eye was normal. Imaging revealed normal MRI brain, MRA head and neck, carotid ultrasound, and trans-thoracic echocardiography. Trans-esophageal echocardiography revealed a small to moderate immobile plaque in the transverse and descending aorta. Laboratory evaluation revealed positive RPR at 1:2 titer with positive FTA-Abs. Toxoplasma IgG was elevated and IgM was negative.

Conclusion:
Retinal vascular plaques should always raise the suspicion of emboli, yet careful history taking and the presence of chorioretinal lesions indicative of prior inflammation should prompt consideration of Kyrieleis’ plaques of infectious origin, such as toxoplasmosis, syphilis, HIV, herpes, and tuberculosis. Kyrieleis’ plaques are in the arterial wall, multiple, immobile, and remain unchanged for long periods. Embolic plaques are within the arterial lumen, tend to be located at vessel bifurcations, and may be mobile.

References:
5. Kyrieleis, Uber atypische gerfaesstuberkulose der netzhaut. Arch Augenheilkd. 107, 182-190, 1933

Key Words: Retina, Vasculitis, Inflammatory, Masquerade

Financial Disclosure: None
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Mycophenolate Mofetil as First-Line Therapy for Ocular Myasthenia Gravis

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Introduction:
Ocular myasthenia gravis (OMG) may produce debilitating diplopia or ptosis leading to significant visual disability. Symptomatic treatment with pyridostigmine is frequently ineffective and immunosuppressive therapy with corticosteroids can have serious side-effects. Mycophenolate mofetil (MMF), a steroid-sparing agent, has been used in generalized MG. We present 15 cases of OMG treated with MMF as a first-line agent.

Methods:
We performed a retrospective analysis of 15 consecutive patients with isolated OMG or OMG and mild bulbar symptoms treated with MMF as first-line therapy. Diagnosis was confirmed by either single fiber electromyography (SFEMG) or elevated titers of acetylcholine receptor antibodies (AchR-Abs). Patients with prior or concurrent use of other immunomodulating therapies were excluded. All were treated with 1000-3000mg total daily. Response to therapy was assessed using the Myasthenia Gravis Foundation of America post-intervention status scale.

Results:
Mean age at symptom onset was 63 years (range 33-84 years). Eleven were male and 4 were female. Three were diagnosed by SFEMG and 12 by AchR-Abs. Twelve patients had isolated OMG, while 3 also had mild bulbar symptoms. None had thymoma. Average total follow up time was 11.5 months (range 4-36 months). All 15 patients improved (mean=3.4 months; range 1-8 months). Eight patients (53%) reached pharmacologic remission, 4 (27%) were minimal manifestations (MM)-1, and 3 (20%) were MM-3. No patients had complications related to MMF.

Conclusion:
Given the clear risk of corticosteroid therapy in our older patient population, we felt MMF should be considered as a first-line agent. We found no published series of the use of MMF as a first-line agent in OMG. MMF was effective as a first line therapy in patients with disabling OMG.

References:

Key Words: Myasthenia, Mycophenolate

Financial Disclosure: None
When The Kidneys Turn Off The Light. Uremic Optic Neuropathy

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Introduction:
Uremic optic neuropathy has been reported most commonly in the end-stage renal disease. We reported bilateral uremic optic neuropathy secondary to acute renal failure precipitated by bowel preparation with sodium phosphate solution (SPS).

Methods:
Case report

Results:
A 62-year-old male developed bilateral vision loss after colostomy closure surgery. Four days before the procedure, the patient was evaluated by colonoscopy previous bowel preparation with fleet’s phospho-soda; one day before surgery received additional douses. Twenty-four hours, after surgery, the patient began improved progressive vision in the left eye, but the right eye remains blind. The ophthalmic examination was normal and after 6 weeks the optic disc become pale. The patient was treated with steroids without improvement. The laboratory test were normal before bowel preparation but at the surgery day the creatinine, urea, electrolytes levels were elevated.

Conclusion:
Acute renal failure following sodium phosphate solution (SPS), resulting in acute nephrocalcinosis, Phosphate nephropathy can cause acute bilateral visual loss. The clinical case demonstrated that: acute renal failure may be responsible of optic neuropathy; some risk factor can precipitated it. Urgent treatment with steroids, hemodialysis or both is required. The prognosis is uncertain.

References:

Key Words: Uremic Optic Neuropathy, Acute Renal Failure, Nephrocalcinosis, Sodium Phosphate Solution, Ischemic Optic Neuropathy

Financial Disclosure: None
Introduction:
Acute Macular Neuroretinopathy (AMNR) is characterized by unilateral or bilateral visual loss, central ring scotomas and macular reddish-brown lesions, usually with normal electroretinogram (ERG) and fluorescein angiography. We described the sequence of optical coherence tomography (OCT) findings related with clinical manifestations in the acute and recovery phases.

Methods:
Two patients with unilateral and bilateral AMNR underwent completed neurophthalmic examination, visual fields, fluorescein angiography, neuroimaging and with spectral-domain and time domain optical coherence tomography.

Results:
The patients had acute visual loss, dyschromatopsia, and relative afferent pupillary defect in the unilateral case, ring scotoma and petaloid macular lesions. OCT demonstrated thickness of the macula and outer retinal layer findings. After the clinical manifestations improved the macular became thin and outer retinal layer change. Also, Spectral domain showed more specific findings that time domain.

Conclusion:
OCT can help in the diagnosis and follow up of patients with AMNR, especially with the more resolution devices.

References:

Key Words: Acute Macular Neuroretinopathy, Doughnut Maculopathy, Ring Scotoma, Acute Vision Loss, Optical Coherence Tomography

Financial Disclosure: None
Diagnostic Ability of Fourier-Domain (3 D OCT-1000®) OCT with a Normative Database to detect Band Atrophy of the Optic Nerve

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Introduction:
Patients with chiasmal compression often develop retinal nerve fiber layer (RNFL) loss in a pattern that is called band atrophy (BA) of the optic nerve. Previous studies demonstrated that OCT is able to identify RNFL and macular thickness reduction in eyes with BA but only one evaluated the sensitivity of the time-domain OCT to detect it based on a comparison with an internal normative database. Poor performance was observed when evaluating 30 degree-segments around the disc. The purpose of this study was to evaluate the diagnostic ability of a Fourier-Domain OCT in eyes with BA using the comparison with its internal normative database.

Methods:
Thirty-six eyes with BA and 36 controls were studied. All patients had been submitted to previous treatment of the suprasellar lesion. Fourteen eyes had complete and 22 partial temporal hemianopia. Subjects underwent imaging with the 3 D OCT-1000® (Topcon Corp., Japan). For each parameter, the software provides a classification: within normal limits (higher than 95% confidence level), borderline (below 95%) or outside normal limits (below the lower 99% confidence level of the normal population). Sensitivity and specificity were calculated for each parameter.

Results:
Tables 1 and 2 show the results for RNFL and macular thickness determinations, assuming borderline results as outside normal limits. The temporal average thickness parameter demonstrated the highest sensitivity for detection of abnormalities (94%) among the RNFL parameters and the nasal-inner thickness the highest sensitivity among the macular parameters (81%). Parameters corresponding to the 30-degree segments demonstrated lower sensitivity compared to other RNFL and macular thickness parameters but were significantly better than those obtained previously with the Stratus OCT.

Conclusion:
Macular and RNFL parameters of the 3D OCT-1000 showed high sensitivity to demonstrate axonal loss based on the normative database with significant improvement when compared with time-domain OCT, particularly in the 30-degree segment analysis.

References:

Key Words: Optical Coherence Tomography, Band Atrophy, Chiasmal Compression

Financial Disclosure: None
Saccadic Related Potentials Recorded in the Human Basal Ganglia and Thalamus During Deep Drain Stimulation are Volume Conducted Extraocular Muscle Potentials

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Introduction:
The presence of gamma (31–200 Hz) oscillations in local field potentials (LFP) of the basal ganglia and thalamus during limb movements prompted investigating LFPs during saccades. Recent studies show extraocular muscle spike potentials in scalp electroencephalograms (EEG), but depth electrode recordings were assumed to be secluded. Deep brain stimulation (DBS) provides opportunity to analyze and determine the origin of saccade-related LFPs in the basal ganglia and thalamus.

Methods:
We recorded activity during DBS in eleven patients: 6 in subthalamic nucleus (STN); 3 in globus pallidus interna (GPI); and 2 in thalamic ventralis intermedius nucleus (Vim). Patients performed visually-cued horizontal saccades while LFPs from quadripolar DBS electrodes, EEG, and electrooculograms were recorded. Averaged saccade onsets were determined from electrooculograms and aligned to gamma band-pass filtered LFPs. Wavelet spectrograms of the averaged LFPs, EEG and electrooculogram were constructed with time-frequency relationship and aligned to target light illumination and eye-movement onset; and event-related gamma synchronizations (ERS) were compared to baseline without eye motion.

Results:
STN, GPI and Vim LFPs and EEGs showed bilaterally symmetric gamma oscillations at and after saccade onset; but not during target light illumination without saccades. ERS of DBS LFPs divulged consistent quadripolar symmetry. Wavelet spectrograms of DBS LFPs, frontal EEGs and electrooculograms looked identical. ERS was also observed during vestibulo-ocular reflex smooth eye motion studied in one STN patient, suggesting far-field conduction potentials.

Conclusions:
ERS with bilateral as well as quadripolar symmetry recorded during saccades and vestibular smooth eye movements, similarity of ERS between the DBS LFPs and frontal EEGs, and absence of ERS during target light illumination endorse spike potentials rather than brain neural activity. ERS recorded from electrooculogram channels confirm this ‘gamma imposter’ to be a volume conducted extraocular muscle myogenic potential. Event related potentials should be interpreted in view of, and qualified by, the influence of saccades.

References:

Key Words:  Saccade, Local field potentials, Spike potentials, Gamma oscillations, Deep brain stimulation

Financial Disclosure:  None
Slack Extraocular Muscles Cause Diplopia on Eccentric Gaze in Enophthalmos

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Introduction:
The mechanism of diplopia from enophthalmos is not well characterized. We studied a patient who developed diplopia from enophthalmos caused by a bony orbital defect following craniotomy.

Methods:
Case report, with eye movements recorded at 60 Hz using two infrared video eye trackers.

Results:
A 55-year-old man presented with a one-year history of diplopia. He had undergone a left orbitozygomatic pterional craniotomy two years earlier for clipping of a basilar tip aneurysm. Diplopia was present mostly in eccentric gaze, but he also experienced transient diplopia upon returning to primary gaze. Visual acuity was 20/20 OU, pupils were normal, and there were 8 mm of enophthalmos OS. Ductions OD were full. Ductions OS showed slight limitation of elevation, adduction, and abduction. Binocular fixation fields revealed diplopia with upgaze beyond 10 degrees, or 15 degrees to either side. These findings were confirmed by eye movement recordings: ocular separation increased with gaze eccentricity. A CT scan revealed a large defect in the zygomatic and sphenoid bones, with a loop of the lateral rectus extending through the defect in the lateral orbital wall. The left medial rectus was 32 mm long, compared with a right medial rectus of 40 mm.

Conclusion:
If the orbital walls are not adequately reconstructed following transorbital craniotomy, there is a net increase in orbital volume. The resulting posterior displacement of the globe can create a relative slack or redundancy in the extraocular muscles, as visualized on CT. This has an effect similar to a recession of all 4 rectus muscles. Ocular rotation becomes limited in each direction, because the maximum force each muscle can generate is reduced. The same process may contribute to diplopia in Sunken Eyes, Sagging Brain Syndrome. Our case underscores the importance of orbital wall restoration after orbitotomy, and suggests a mechanism for how post-operative enophthalmos can produce diplopia.

References: None

Key Words: Diplopia, Enophthalmos, Orbitozygomatic Pterional Craniotomy, Eye Movement Recordings, Gaze Restriction

Financial Disclosure: None
Compressive Optic Neuropathy Caused by Post-Neurosurgical Cerebrospinal Fluid Leak

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Introduction:
Cerebrospinal fluid (CSF) leaks into the orbit are rare. They usually occur following trauma rather than post-surgically. We report a case of orbital CSF accumulation after surgery causing a compressive optic neuropathy.

Methods:
Case report

Results:
An 18 yo woman with choanal atresia undersent repeat endoscopic bone resection by general plastic surgery. She awoke with vision decreased to 20/50 OD and 20/100 OS with a bitemporal hemianopia. Emergent CT showed a postsurgical pathway through the sphenoid sinus, with bone posterior and superior to the sella, just posterior to the chiasm. Urgent exploration via pterional orbitozygomatic craniotomy revealed a disrupted chiasm. The bone fragment could not be safely removed. A lumbar drain was placed, and the patient was stable for several days. On POD#5 after drain removal, over several hours she developed progressive proptosis OS of 6 mm with a new left RAPD. CT scan revealed a collection within the left superior orbit consistent with CSF. After lumbar drain replacement with maximal output, 2mL orbital fluid was percutaneously aspirated with a retrobulbar needle. Beta-2 transferrin confirmed the fluid as CSF. Proptosis immediately improved. Over several days the patient improved, although the field defect persisted from the original chiasmal injury.

Conclusion:
Usually, CSF leaks present with rhinorrhea or otorrhea. The treatment is eliminating the anatomic communication. Following transcranial orbitotomy, small communications exist between the subarachnoid space and orbit, but leaks do not occur. Both a communication and elevated intracranial pressure (ICP) are required for leakage to occur. Thus, upon drain removal, an elevation in ICP forced CSF into the orbit. Management should be lowering ICP, successfully treating most cases. If there is an acute compressive optic neuropathy, percutaneous drainage of CSF may be necessary; however, unless the ICP is addressed, CSF will re-accumulate. Eliminating communications between the orbit and intracranial space is not necessary.

References: None

Key Words: Proptosis, Optic Neuropathy, Orbit, Increased Intracranial Pressure

Financial Disclosure: None
**Poster 111**

**Congenital Ocular Motor Apraxia without Joubert’s Syndrome: Overlooking the “Mini-Molar-Tooth” Sign and Subtle Hypoplasia of Inferior Vermis**

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**Introduction:**

Joubert’s syndrome and related disorders (JSRD) include several autosomal recessive “ciliopathies”, which are defined on axial MRI by the presence of the molar tooth sign (MTS), often with hypoplasia of cerebellar vermis. Clinically, JSRD are differentiated from isolated congenital ocular motor apraxia (COMA) by the presence of intellectual disability and hypotonia or respiratory abnormalities during infancy. We wish to draw attention to often-overlooked MR findings of a subtle MTS and occult inferior vermian hypoplasia in isolated COMA.

**Methods:**

Case reports and video-images.

**Results:**

Case 1: An 18-month-old girl had horizontal head thrusts starting at 6 months of age. She had no infantile hypotonia or respiratory difficulties. MRI at 6 months was interpreted as normal by a pediatric radiologist. She was delayed in talking and walking but otherwise had normal neurologic findings and was attentive and socially engaged.

Case 2: A 26-year-old businessman was diagnosed with COMA as an infant. Speech was delayed until age 3 and walking until age 3-4. He was treated for seizures until age 6. He had difficulty reading and slight dysarthria but normal intellect. At age 26, examination showed frequent and irregular conjugate bidirectional torsional intrusions and oscillations. Saccades were normal. MRI was interpreted as normal by a radiologist.

Review of MR scans in both cases showed a subtle MTS and occult hypoplasia of inferior vermis.

**Conclusion:**

1. MTS and inferior vermian hypoplasia are common but often unrecognized MR findings in COMA.
2. Isolated COMA may be a mild form of JSRD.
3. COMA may manifest as irregular torsional intrusions or oscillations of the eyes in adulthood.

**References:**


**Key Words:** COMA, Molar Tooth Sigh, Cerebellar Vermus, Joubert's Syndrome, MRI Brain

**Financial Disclosure:** None
Human Phantom Testing of Transorbital Therapy

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Introduction:
To overcome the challenge of access to the optic nerve, we developed a minimally-invasive, image guided system based upon tracking via deformation of a magnetic field. In this study we compare performance of image guided endoscopy to non-image guided endoscopy in a surgical model of orbital endoscopy.

Methods:
In previous work we developed a combined image-guided orbital endoscopic system for the delivery of therapy to the optic nerve. Human phantoms with one radio-opaque target and three distracter colored targets secured within the orbits were created. Twenty-eight surgeons then used the system in paired trials of image guided compared to non-image guided surgery.

Results:
Real time image guidance and endoscopic video information was demonstrated during these trials. Across all trials the time decreased, demonstrating a learning curve of using the orbital endoscope. There was a marginally significant increase in time using the image guidance, 97 seconds versus 62.5 seconds (p=0.056); but, particularly in the less experienced surgeons, the guidance system reduced the error rate.

Conclusion:
Image-guided flexible endoscopy may prove a useful tool for orbital biopsies, minimally invasive orbital explorations, optic nerve interventions and diagnosis of orbital disease.

References:

Key Words: Optic Nerve, Image Guidance, Minimally Invasive, Endoscope, Human Phantom

Financial Disclosure: /NCRR 1R21RR025806 Research to Prevent Blindness Physician Scientist Award; Patent 60/488,758 Ophthalmic Orbital Surgery Apparatus and Method and Image Guided Navigation System
Poster 113

Gyrus Rectus Syndrome after Craniopharyngioma Resection

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Introduction:
12 year old male presented to neuro-ophthalmology clinic for yearly follow up six years after resection of craniopharyngioma. After the craniopharyngioma resection six years ago the patient had developed a large scotoma in his left eye. The right eye was unaffected and remained normal on repeat visual fields until presentation at this visit. At the visit this year, his vision was 20/20 in the right eye and LP in the left eye. Dilated fundus examination was normal in the right eye and only significant for a pale optic nerve in the left eye. Visual fields showed a partial temporal scotoma in the right eye that was new compared to the previous year and a complete scotoma in the left eye. MRI of the brain showed no recurrence of the craniopharyngioma but further review showed herniation with compression of the right frontal lobe/gyrus rectus causing compression of the right optic nerve.

Methods:
N/A

Results:
N/A

Conclusion:
After the initial diagnosis of gyrus rectus syndrome was made the patient’s family decided to undergo a decompressive craniotomy. When the patient awoke after the craniotomy his vision in the right eye was normal. A few hours later his vision became bare LP in his right eye. An ophthalmology consult was ordered by the neurosurgical team. Dilated fundus examination of the right eye showed pale vessels with a cherry red spot in the macula. At this time it was felt that he had vision loss secondary to post-operative vasospasm resulting in central retinal artery occlusion. The next day he underwent fluorescein angiography showing vascular attenuation but near normal angiographic pattern. His vision remained LP in both eyes one month after surgery. The final diagnosis was gyrus rectus syndrome s/p decompressive craniotomy resulting in central retinal artery occlusion secondary to vasospasm.

References: None

Key Words: Gyrus Rectus Syndrome, Craniopharyngioma, Central Retinal Artery Occlusion, Vision Loss

Financial Disclosure: None
Vascular Risk Factors in Middle-Aged Men with and Without History of Migraine Headaches Experiencing Late-Life Migraine Accompaniments

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Introduction:
Migraine with aura is known to be associated with increased stroke and cardiovascular risk, especially in women, in whom 2-fold risk has been reported. Migraine aura occurring in the absence of headache (acephalgic migraine) in middle-aged men has been called “late-life migraine accompaniments” (LLMA). Several small studies have found no increased stroke risk in middle-aged men with this syndrome.

Methods:
A chart review of men seen in the Neuro-Op Clinic at Central Arkansas VAMC with onset of acephalgic migraine auras of onset in middle age revealed thirty-six patients, of whom 20 had a prior history of migraine headaches and 16 did not. Clinical features, such as scotoma morphology and duration, duration of symptoms, were recorded, as were vascular risk factors, including hypertension, diabetes, smoking history, hyperlipidemia, and coronary artery disease. History of stroke, TIA, ischemic optic neuropathy, or retinal vascular events were also recorded. Statistical analysis of comparison between the groups was carried out using Student’s two-tailed t-test.

Results:
There were no differences between the two groups with respect to age (mean-54), duration of symptoms (mean=7 years), and percentage with scintillating scotomata (60% versus 58%). Strokes and TIAs were more common in the non-migraineurs (25% vs. 13%), whereas hypertension was more prevalent in the migraineurs (60% vs. 30%). Prevalence of cardiac disease, diabetes, smoking history, and hyperlipidemia were all within ten percentage points of each other in the two groups. No differences between groups were statistically significant.

Conclusion:
This retrospective study did not find a significant difference in vascular risk factors or vascular events between migraineurs and non-migraineurs with LLMA. These findings support the hypothesis that LLMA represent an unusual type of migraine rather than being a manifestation of cerebrovascular disease.

References:

Key Words: Migraine, Stroke, Migraine Aura, Acephalgic Migraine, Vascular Risk Factors

Financial Disclosure: None
Management And Prognosis Of Diplopia Associated With Primary Divergence Insufficiency

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Introduction:
Divergence insufficiency is an unusual form of binocular dysfunction characterized by acute onset uncrossed diplopia only at distance and single binocular vision at near. It accompanies with full duction and version. Primary form is neurologically isolated and occurs in otherwise healthy person. Secondary form which is associated with neurologic disorders is another consideration. Our object of this study is to evaluate prognosis and treatment modality of diplopia with primary divergence insufficiency

Methods:
Retrospective case review of 15 patients who were diagnosed primary divergence insufficiency.

Results:
Fifteen patients with primary divergence insufficiency were identified and followed more than 6 months. At initial EOM evaluation, near and distance deviation disparity was 9.5 prism diopters. After 6 months observation, spontaneous regression of uncrossed diplopia showed 33% and after one year, 66%. To alleviate the residual diplopia, prism prescription (4 cases) and bilateral resection surgery (1case) was performed without any complication.

Conclusion:
Uncrossed diplopia in primary divergence insufficiency which is not associated with neurologic disorders revealed good prognosis in terms of spontaneous regression. In case of residual diplopia, prism therapy and customized strabismus surgery is safe and effective for restoring single binocular vision.

References:

Key Words: Divergence Insufficiency, Uncrossed Diplopia, Binocular Dysfunction, Prism, Bilateral Resection

Financial Disclosure: None
Isolated Foveal Hypoplasia: Morphological and Functional Changes

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Introduction:
To describe retinal morphological and functional changes associated with isolated foveal hypoplasia (IFH) in two brothers.

Methods:
Morphological documentation: color and red free (RF) retinography, fundus autofluorescence (FAF), spectral-domain optical coherence tomography (sOCT), (RF, FAF and sOCT: HRA Heidelberg Engineering). Functional tests: Full field and multifocal electroretinography (ERG, and mfERG) recorded according to ISCEV standards (Espion E2; Diagnosys LLC), Ishihara color test, visual filed tests (24-2 full threshold: Humphrey, Zeiss) and microperimetry (MAIA; CenterVue) were performed.

Results:
Fundus examination reveals typical absence of foveal reflex and depression. FAF shows the absence of normal hypoautofluorescence in the foveal area. Multifocal-ERG showed reduced, but present amplitude density increase in the foveal area. Mean P1 amplitude density was: ring 1: 13.8; ring 2: 10.7; ring 3: 9.7; ring 4: 6.9; and ring 5: 5.2 nV/deg², and microperimetry revealed a relative stable paracentral fixation with minor reduction of sensitivity thresholds in all 4 eyes.

Conclusion:
IFH causes foveal pigmentary abnormalities that can be highlighted by AF, but sOCT should be considered crucial evaluation for differentiation of this rare condition and other macular dystrophies, as described before (Querques et al., 2008). Functional changes appear to be related to the reduction of foveal photoreceptor density increase in the fovea, but interestingly, mfERG data indicates that there might be certain photoreceptor density increase at retinal centre, which could be related to the preference for a central fixation position.

References:

Key Words: Retinal Dystrophy, Electroretinography, Low Vision

Financial Disclosure: None
Poster 117

3T Assessment of Lateral Geniculate Nucleus Atrophy in Glaucoma and Correlation with Clinical Stage of Disease

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Introduction:
The aim of the present study is to examine the MR appearance of human glaucoma and to correlate the disease stage with the degree of lateral geniculate nucleus (LGN) atrophy. In addition, proton density weighted images (PD) are compared to images tailored for assessment of gray matter (GM) with respect to quantification of LGN atrophy, and measurements of LGN height and volume are compared with respect to their correlation with glaucoma stage.

Methods:
26 patients with known glaucoma and 26 age-matched healthy volunteers were included in this IRB-approved study. All subjects underwent imaging on a 3T MR system utilizing GM and PD sequences. LGN height and volume were measured by two blinded neuroradiologists. Measurements were compared between glaucoma and control patients and correlated with clinical disease severity as assessed by static threshold visual field parameters.

Results:
Average maximum LGN height in glaucoma patients on PD images was 4.36 mm (right) and 4.31 mm (left), significantly less (p<0.05) than respective measurements of 5.05 mm and 4.99 mm in volunteers. With the GM sequences such respective measurements were also less (p<0.05) in glaucoma patients, 4.2 and 4.0 mm, versus respective measurements in volunteers of 4.88 mm and 4.77 mm. Average LGN volumes in the patient group were 98.0 mm³ on the right and 93.7 mm³ on the left with the PD sequence versus respective measurements of 85.2 and 80.5 mm³ with GM sequence. All volume measurements were greater in volunteers (p<0.05). In the patient group, both maximum height and volume of LGN with both sequences were strongly correlated with clinical glaucoma stage (p <0.05).

Conclusion:
MR measurements of LGN height and volume are diminished in glaucoma patients and strongly correlate to clinical glaucoma stage, suggesting a novel imaging marker of disease severity.

References: None

Key Words: MRI, Glaucoma, 3 Tesla

Financial Disclosure: None
Poster 118

Role for Low-Contrast Acuity as Part of a New MS Disability Composite Measure

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Introduction:
In the AFFIRM trial of natalizumab vs. placebo for relapsing multiple sclerosis (MS), there was a reduction in sustained loss of low-contrast acuity (LCA) for patients on natalizumab. LCA captures information on visual function not entirely captured by the Expanded Disability Status Scale (EDSS) or the MS Functional Composite (MSFC), making these measures complementary for assessing disability. Using AFFIRM data, we evaluated a potential composite measure for sustained disability defined by progression in LCA, EDSS, and/or in any of the MSFC components (timed 25-foot walk [T25FW], 9-hole peg test [9-HPT], and Paced Auditory Serial Addition Test [PASAT-3]).

Methods:
AFFIRM was a 2-year pivotal, placebo-controlled, natalizumab monotherapy study. EDSS progression was defined as either a ≥1.0-point increase in score from baseline (if baseline ≥1.0), sustained for 3 months, or as a ≥1.5-point increase in score (if baseline EDSS=0), sustained for 3 months. For MSFC components, progression was defined as ≥15% worsening from baseline, sustained for 3 months. Visual progression was defined as a 7-letter reduction in LCA scores at 2.5% contrast, sustained for 3 months.

Results:
Baseline characteristics were balanced between natalizumab (n=627) and placebo groups (n=315). Proportions of patients who met individual criteria for 3-month sustained progression were lower in the natalizumab group vs. placebo for LCA (20% vs. 28%), EDSS (17% vs. 27%), T25FW (22% vs. 33%), 9-HPT (10% vs. 18%), and PASAT-3 (4% vs. 5%). Natalizumab significantly reduced risk of 3-month sustained composite progression (LCA, EDSS, and any MSFC component) vs. placebo by 33% at 2 years (HR=0.67; 95% CI=0.56, 0.80; P<0.0001).

Conclusion:
Adding visual function testing by LCA to EDSS and MSFC significantly increases the capacity of these measures to detect progression of disease in clinical trials of relapsing MS. These data will be useful in designing future studies with prospectively defined MS composite progression measures.

References:

Key Words: Low-Contrast Acuity, Vision, Multiple Sclerosis, Disability, Natalizumab

Financial Disclosure: Dr. Balcer has received consulting fees and honoraria from Biogen Idec and Bayer for development of visual outcome measures for MS trials and for currently serving on a clinical trial advisory board. Dr. Galetta has received honoraria from Biogen Idec and Teva.

Dr. Rudick has received royalties from Informa UK LTD, 2009 book: “Multiple Sclerosis Therapeutics”; and consulting fees from Biogen Idec and Bayhill Therapeutics.

Prof. Polman has received honoraria and consulting fees from Actelion, Bayer Schering, Biogen Idec, GlaxoSmithKline, Merck Serono, Roche, UCB, Teva, and Novartis.

Drs. Hotermans, Zhang, and Hyde are employees of Biogen Idec.
Poster 119

Idiopathic Intracranial Hypertension (IIH): Visual Outcomes in the Very Obese

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Introduction:
Increased body mass index (BMI) has been associated with increased risk of IIH, but the relationship of BMI to IIH outcomes is less clear. We hypothesized that increasing BMI was associated with less favorable visual outcomes in IIH.

Methods:
Retrospective chart review of all adult cases of IIH who satisfied the modified Dandy criteria seen at our institution between 1989 and 2010. Demographics, diagnostic evaluations, baseline visit and last follow-up examination data, treatment and visual outcome data were collected. Groups were compared using $\chi^2$, Fisher test, and Mann-Whitney U-test at the 0.05 significance level. Logistic regression was used to evaluate the relationship of BMI to severe visual loss (VA<20/200 or VF<20°), evaluating for interaction and controlling for potential confounders.

Results:
158/414 included IIH patients had BMI≥40 (WHO Obese Class III), and 172/414 had BMI 30-39. Patients with BMI≥40 were more likely to have more severe papilledema at first (p=0.02) and last (p=0.08) neuro-ophthalmology encounter than those with a lower BMI. They were more likely to have diagnosed hypertension (25 vs. 14%, p=0.002) and diagnosed sleep apnea (17 vs. 5%, p<0.001). They were also more likely to have severe visual loss in one or both eyes at last follow up (18 vs. 11%, p=0.067). Compared to patients with BMI<30, patients with BMI 30-40 had a 3.7 (95%CI:1.3-13, p=0.02) times greater odds, and those with BMI≥40 a 4.1 (95%CI:1.5-14.5, p=0.01) times greater odds, of severe visual loss after controlling for sex, race, diagnosed hypertension, and diagnosed sleep apnea.

Conclusion:
We found that BMI≥30 has an independent effect on visual outcome in IIH similar to black race[1] and male gender[2], emphasizing the importance of weight management in the treatment of IIH.

References:

Key Words: Idiopathic Intracranial Hypertension, Obesity, Vision Loss

Financial Disclosure: None
The Use of Pentoxifylline in the Prevention of Second Eye Involvement in Patients with NAION

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Introduction:
NAION is a common cause of acute optic nerve disease in patients over 55 years of age. There is no accepted method for the prevention or reduction of the likelihood of second eye involvement. The rate of second eye involvement according to the 5 year data from the IONDT was approximately 15%. We analyzed our institutional data for second eye involvement (SEI) in patients using pentoxifylline (rheologic agent that increases optic nerve head perfusion in animal models) as prophylaxis.

Methods:
A retrospective chart review was undertaken to look at the rate of second eye involvement in patients using pentoxifylline as prophylaxis. Due to practice patterns at our institution approximately half the patients seen with NAION had been placed on pentoxifylline prophylaxis (referred to as “treatment group”) and the other half served had not been thus serving as internal controls (control group).

Results:
456 patients were identified that had documented cases of NAION in one eye only. 235 patients were treated with pentoxifylline and 221 were not. 10% (23/235) of the treatment group had SEI vs. 3% (7/221) in the control group. At 3 years, 18.75% of the treatment group had SEI. Median time to second eye involvement was 7 months for the treatment group vs. 20 months for the control group.

Conclusion:
Our institutional data showed a lower SEI rate than that of the IONDT, however these differences can be accounted for due to variability of follow-up. However, the 3 year data shows that the treatment group had no difference from the IONDT rate. Therefore, it would seem that pentoxifylline plays no role in prevention of second eye involvement. Furthermore, when the time to second eye involvement is compared there appears to be a statistically significant trend, indicating that SEI may occur sooner in patients on pentoxifylline.

References:

Key Words: NAION, Second Eye Involvement, Pentoxifylline, IONDT

Financial Disclosure: None
Incidence of NAION among Medicare Beneficiaries with and without Diabetes

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Introduction:
Previous studies have identified a higher prevalence of diabetes mellitus (DM) among patient cohorts with non-arteritic anterior ischemic optic neuropathy. We sought to determine the development of incident NAION among a group of newly diagnosed patients with DM and to estimate the incidence of NAION among the elderly.

Methods:
Query of Medicare 5% claims files identified patients with new diagnosis of DM in 1994. A randomly selected control group was created using one-to-one propensity score matching. Patients with a diagnosis of giant cell arteritis, pre-existing DM, and age < 68 years or > 95 years were excluded. Diabetics and controls were followed for the development of incident NAION over the following 13 years.

Results:
25,515 diabetics and an equal number of controls were identified. Each group was 85% White with a mean age 76.4 years and 40% male. The mean follow-up time was 7.6 years. 188 patients with DM (0.7%) developed AION compared to 131 controls (0.5%; p<0.01). After adjusting for other covariates, the risk of developing AION among individuals with DM was increased by 40% (HR: 1.397; 95% CI: 1.115,1.750). Male gender increased an individual’s risk of developing AION by 32% (HR: 1.319; 95% CI: 1.052,1.654). No other covariate was significantly associated with developing AION. The annual incidence of NAION was 48 per 100,000.

Conclusion:
DM significantly increased the risk of receiving a diagnosis of NAION. The incidence of NAION among patients older than 67 years may be much higher than previously reported.

References: None

Key Words: NAION, Diabetes, Incidence, Medicare

Financial Disclosure: None
Water Disease

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Introduction: Thyroid related orbitopathy (TRO) is the most common cause of bilateral proptosis in adults. We report a previously-healthy 67-year-old man with progressive bilateral proptosis for 6 months, who was initially diagnosed with TRO but on follow-up was found to have bilateral neoplastic infiltrative orbital disease.

Methods: The patient underwent serial examinations and neuroimaging. VA was 20/30 RE and 20/25 LE. No APD was identified. Motility restriction was noted, but no diplopia in primary gaze. There was bilateral superior scleral show, conjunctival injection, 1mm lagophthalmos and proptosis. DFE and Humphrey 30-2 visual field test were normal. Orbital MRI without gadolinium revealed fusiform enlargement of extraocular muscles without tendinous involvement and with prominence of lacrimal glands bilaterally suggestive of TRO. TSH was mildly elevated at 4.8, and T4, free-T3 and antihyroglobulin antibody were normal. Diagnosis of TRO was made. At 3-month follow-up, patient presented with VA 20/50 RE and 20/30 LE. Marked worsening of edema and proptosis was noted bilaterally. RE showed APD, choroidal folds and normal optic nerve. Repeat orbital MRI with gadolinium illustrated diffuse infiltrative process involving the orbits with marked bilateral proptosis, right globe tenting and extension into skull base foramina.

Results: Anterior orbitotomy with orbital tissue biopsy was performed. Microscopic examination, immunohistochemical, flow-cytometric and gene-rearrangement studies revealed a monoclonal, atypical population of small lymphocytes which were CD20, CD79a, CD5 and Cyclin D-1 positive and CD23 negative.

Conclusion: In summary, this is a case of orbital lymphoma that initially simulated TRO with “thyroid like” symptoms, clinical, laboratory and imaging findings. Upon follow-up marked progression of infiltrative process was noted and repeat imaging studies were suspicious for malignant process. Orbital biopsy showed atypical monoclonal lymphocytic population consistent with mantle cell-type malignant lymphoma. Our case underscores the importance of follow-up and repeat neuroimaging when necessary in patients with signs and symptoms of TRO.

References: None

Key Words: Orbital Lymphoma, Thyroid Eye Disease, Proptosis

Financial Disclosure: None
Poster 123

Opsoclonus-Myoclonus with Exaggerated Startle Response

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Introduction:
Opsoclonus-myoclonus syndrome (OMS) is a rare neurological disorder that is usually seen in conjunction with tumors (most commonly breast, lung and gynecological), infections or toxic-metabolic processes. In addition to opsoclonus and/or myoclonus, the clinical features of OMS may include ataxia and encephalopathy. We describe a patient who presented with OMS and a strikingly exaggerated startle response, prompting an evaluation for an underlying malignancy which led to diagnosis of a small-cell lung carcinoma.

Methods:
A 64-year-old female presented with a 3-month history of dizziness and unsteadiness. The dizziness was non-vertiginous with a sense of light-headedness, and was associated with nausea and vomiting. These began as episodes that lasted a few minutes and evolved over weeks to become constant. Two weeks prior to her presentation, she described “jumping” vision, slurring of her speech and a staggering gait. Furthermore, the patient reported a weight loss of 15 pounds over the past months. Her past medical history was unremarkable apart from a history of tobacco use which she had ceased 20 years prior. Her family history was non-contributory. There had not been recent constitutional symptoms of fever, myalgia or arthralgia. She was on no regular medications.

Results:
She had high-frequency bursts of low-amplitude back-to-back, multi-directional saccades that were exacerbated during smooth pursuit. Smooth pursuit was occasionally punctuated by the oscillations. In addition she had spontaneous myoclonic jerks of her head and an exaggerated startle response to sound and light. There was mild dysmetria on finger-nose and heel-shin testing. Her gait was slightly wide-based, unsteady and she could not walk in tandem unassisted. MRI brain and CT temporal bones with superior canal dehiscence protocol were unrevealing. On PET-CT, there was increased FDG uptake in the cerebellum relative to the cerebrum. A 4.2cm lung mass was found in the left lower lobe which confirmed small-cell carcinoma on histopathology. She had a limited stage small cell lung carcinoma. Her paraneoplastic panel was normal.

Conclusion:
Opsoclonus-myoclonus syndrome associated with hyperekplexia secondary to paraneoplastic syndrome from a small-cell lung carcinoma.

References: None

Key Words: Opsoclonus, Paraneoplastic Syndrome, Lumbar Puncture, Magnetic Resonance Imaging, Vertigo

Financial Disclosure: None
A Case of Creutzfeldt-Jakob with Periaqueductal Grey Involvement and Upgaze Paresis

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Introduction:
- HPI: 60-year-old male ophthalmologist from Mexico c/o inability to perform cataract surgery b/c binocular oblique diplopia, headache & mental status changes.
- Onset 3 mos ago with progressive worsening
- Trouble remembering things & finding the right words
- Experiences imbalance w/ walking.
- Bitemporal headache, moderately severe, intermittent, lasting secs, 2-3/day & will occasionally awaken him.
- Previous workup
  - Cardiac: EKG, coronary angiogram, thallium stress test, Holter monitor & carotid dopplers negative.
  - Neurologic: MRI brain, EEG, EMG, audiometry, thoracic CT, visual auditory & somatosensory evoked potentials "negative."
  - PMHx, PSHx, FHx & SHx noncontributory
  - Medications: piracetam, telmisartan & aspirin.
- Ocular exam
  - VA 20/25 OD 20/25 OS
  - Pupils: isocoric, no APD
  - Confrontation fields: full OU
  - External exam: intermittent lid retraction, no lid lag, intermittent 5° chin-up position
  - Rest of anterior and posterior exam unremarkable
  - No nystagmus, no uveitis, no disc edema
  - Bilateral mild global ophthalmoplegia worse for upgaze
  - Orthoptic measurements: Primary gaze 7 ET; Right gaze 6 ET, Flick LHT; Left gaze 6 ET, 3LHT; Upgaze 4-6 LHT; Downgaze ortho

Methods: Case report.

Results:
- Inpatient workup
  - Structural MRI brain w/wo contrast: “normal"
  - EEG: normal
  - CT chest, abdomen & pelvis: no neoplasm or sarcoidosis.
  - SPECT brain scan: normal.
  - LP: normal cell count, protein & glucose.
  - BMP, CBC, Lyme titers, thyroid testing, SPEP, thyroid & anti-acetylcholine receptor antibodies: normal.
  - Pyridostigmine tried w/o effect.
- Between discharge and neuro-ophth f/u 2 mos later
  - Neurocognitive symptoms & diplopia worse.
  - Neuropsychological testing revealed: memory & executive function abnormalities w/ frontal lobe redominance.
  - Per family, ↑ confusion, dizziness & inability to walk.
  - Now wheelchair-bound w/ sporadic nocturnal myoclonic activity.
  - CSF study: positive for 14-3-3.
  - Repeat MRI brain: symmetric, mild diffusion restriction in the basal ganglia, thalami and mesial temporal regions bilaterally & subtle abnormally high signal in periaqueductal grey.

Conclusion:
Final diagnosis: Sporadic Creutzfeldt-Jakob disease1. Localization of upgaze paresis presumed related to FLAIR & DWI diffusion restriction in the brainstem (periaqueductal grey). Take home point: Diplopia associated with subacute progressive neurocognitive deficits should prompt consideration of Creutzfeldt-Jakob disease and DWI and spinal fluid (14-3-3, tau) might be helpful in diagnosis.

References:

Key Words: Opsoclonus, Paraneoplastic Syndrome, Lumbar Puncture, Magnetic Resonance Imaging, Vertigo

Financial Disclosure: None
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Cancer Associated Retinopathy (CAR) Presenting Two Years Prior to Diagnosis of Large Cell Neuroendocrine Carcinoma of Unknown Primary

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Introduction:
Cancer associated retinopathy (CAR) is a para-neoplastic disease affecting the retina leading to progressive visual loss. CAR is typically associated with small cell lung cancer and gynecologic malignancies and typically precedes the diagnosis of the malignancy.

Large cell neuroendocrine carcinoma (LCNEC) is typically a type of primary lung cancer with relatively aggressive behavior. Associated para-neoplastic syndromes have been observed with LCNEC including Lambert-Eaton, ectopic ACTH production, opsoclonus, and CAR. CAR associated with LCNEC has been rarely reported literature, and when it has, the malignancy was discovered at the time of evaluation of the patients’ symptoms.

A case of CAR in a 72 year old man diagnosed 2 years prior to the diagnosis of LCNEC is presented along with pathologic specimens.

Methods:
Single Case Study

Results:
A 72 year-old man presented with a one year history of “grainy” vision in both eyes and nyctalopia. Full-field ERG was performed which showed undetectable rod function and impaired cone function. Cancer-associated retinopathy was suspected. A full skin examination was performed by a dermatologist which did not reveal any melanoma. CT of the chest, abdomen, pelvis and whole body PET scan were performed which were normal. Western blot for the anti-retinal antibodies were positive for the 40kDa protein and negative for other anti-retinal antibodies including anti-recoverin. Since no cancer was found on CT, PET, and dermatological evaluations, the diagnosis of auto-immune retinopathy was made. Treatments with two courses of 5 days of IVIg were made without any response.

Around 26 months after onset of visual symptoms, the patient awoke with a mass on the left side of the neck. A lymph node biopsy was performed and was found to have large cell neuroendocrine carcinoma positive for pankeratin and CD 56, negative for CDX2, CEA, CK7, CK 20, P63, PAP, TTF1, and P3A.

Conclusion:
This is a case of cancer associated retinopathy with positive anti-retinal antibodies presenting over two years before the diagnosis of a large cell neuroendocrine carcinoma with initial negative cancer evaluation.

References:

Key Words: Paraneoplastic Syndromes, Visual Field Loss, ERG

Financial Disclosure: None
Pathologic Abnormalities in the Optic Nerves from Familial Dysautonomia

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Introduction:
Riley Day syndrome, or familial dysautonomia, has rarely been shown to be associated with optic atrophy. Recently we have evaluated a group of patients with this condition and have found evidence of maculopapillary optic nerve damage in all of them. One of these patients died and his eyes were available at autopsy.

Methods:
A 60 year-old man, diagnosed by Riley with familial dysautonomia underwent thorough neuro-ophthalmic examination, including visual field testing and optical coherence tomography. An MRI was done. He died several weeks later and his eyes were obtained at autopsy.

Results:
Paraffin-embedded, formalin-fixed histologic sections in the transverse plane of the right optic nerve showed a distinctive wedge of atrophy involving the maculopapular bundle. There was relative preservation of the other nerve fiber bundles. Immunohistochemistry for neurofilament showed the loss of axons involving the maculopapular bundle.

Conclusion:
This case demonstrates what we have observed clinically in a recent study of 30 patients with familial dysautonomia. In all patients there is selective maculopapular nerve fiber loss. Although many patients lose vision from corneal scarring, there have been a few reports describing optic atrophy without further clarification. In this case, the bitemporal nature of the visual field loss raised the possibility that he could have had a chiasmal lesion. This was not the case. The distinct nerve fiber bundle loss in the optic nerves seen at autopsy is typical of nutritional, toxic or hereditary optic neuropathy. Some patients with familial dysautonomia do have difficulty swallowing and some do end up having gastrostomies done prophylactically. However, in this case, there was no history of nutritional deficiency. Vitamin B12 levels were consistently normal throughout our patient’s period of observation. Our findings indicate that it is possible that the genetic abnormality associated with FD may affect other genes secondarily including those involved mitochondrial protein production leading to the type of optic neuropathy that is seen in other hereditary optic neuropathies, such as Leber’s hereditary optic neuropathy or autosomal-dominant optic neuropathy. These findings may have implications regarding the molecular biology of familial dysautonomia.

References:

Key Words: Familial Dysautonomia, Riley Day Syndrome, Optic Atrophy, Central Scotoma

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