



North American Neuro-Ophthalmology Society

42nd Annual Meeting

February 27 - March 3, 2016

JW Starr Pass Marriott • Tucson, Arizona

Poster Session II: Scientific Advancements in Neuro-Ophthalmology

Tuesday, March 1, 2016 • 6:00 pm - 9:30 pm

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

Odd-Numbered Posters: 6:45 pm - 7:30 pm

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Poster 117

Timing of Non-Arteritic Anterior Ischemic Optic Neuropathy (NAION) After Modern Cataract Extraction

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Introduction:

To study the prevalence of non-arteritic anterior ischemic optic neuropathy (NAION) and its temporal relationship with modern cataract surgery.

Methods:

Retrospective and prospective, chart review and patient historical assessment. SETTING: Single tertiary care center. All patients seen at The Wilmer Eye Institute between January 1, 2010 and December 31, 2014, with a documented episode of NAION were included in this study. Records were reviewed to identify which patients had previous cataract surgery during the 1-year period prior to developing an NAION. Patients with cataract surgery were reviewed and evaluated for differences in perioperative anesthesia, duration of surgery, and time interval between surgery and NAION onset.

Results:

188 patients were identified as having had an episode of NAION during the study period. Of these, 18 (9.6%) patients (22 eyes) had history of cataract extraction during the year subsequent to developing NAION. The median interval between cataract surgery and NAION was 173 days (range: 9-328 days). NAION occurred within 3 months, 3-6 months, 6-9 months, and 9-12 months after cataract surgery in 5 eyes (22.7%), 7 eyes (31.8%), 8 eyes (36.4%), and 2 eyes (9.1%) respectively (Figure 1). The temporal relationship between NAION onset and previous surgery was evaluated by using a chi-squared goodness of fit t-test to compare the observed with a uniform distribution across 3-month time interval, which suggested no significant temporal pattern ($P=0.28$).

Conclusions:

Our data suggest that there is no temporal relationship between modern cataract surgery and the occurrence of NAION.

References:

McCulley TJ, Lam BL, Feuer WJ. Nonarteritic Anterior Ischemic Optic Neuropathy and Surgery of the Anterior Segment: Temporal Relationship Analysis. *Am J Ophthalmology*, 136 (6), 1171-1172, 2003.

Keywords: NAION, Cataract Surgery

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 118

Getting Stoned From Being Too High: Accidental Discovery That Optic Disc Drusen May Arise From High Intracranial Pressure of Pseudotumor Cerebri / Idiopathic Intracranial Hypertension

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Introduction:

To evaluate the prevalence of optic disc drusen (ODD) in the setting of papilledema from pseudotumor cerebri/idiopathic intracranial hypertension (IIH), and to assess if the coexistence of both conditions may worsen the visual outcomes as compared with either condition alone.

Methods:

Observational retrospective review of 372 consecutive participants with ODD or resolved papilledema (rPAP) from IIH evaluated over a 25-year period. The prevalence of ODD among participants with rPAP was calculated. One eye from each of 372 participants was used to assess logMAR visual acuity and visual field mean deviation (MD) outcomes.

Results:

Participants included 170 ODD alone, 159 rPAP alone, and 43 combined ODD with rPAP (cODD-rPAP). The prevalence of ODD at 19% among eyes with rPAP alone and combined was almost 10 times greater and significantly higher ($P < 0.001$) than the prevalence of ODD in the general population. Eyes with exposed drusen had significantly worse visual acuity at 0.14 logMAR as compared with eyes with buried drusen at 0.029 logMAR ($P = .025$). The visual field MD of eyes with exposed drusen at -9.88 dB was worse than buried drusen at -3.35 dB ($P < 0.001$), rPAP at -4.80 dB ($P < 0.001$), and rPAP with exposed drusen at -6.02 dB ($P = 0.047$). Eyes with cODD-rPAP did not have worse visual acuity or visual fields than eyes with rPAP alone.

Conclusions:

The high prevalence of ODD with rPAP suggests a non-coincidental relationship, and that in some cases ODD may develop secondary to papilledema. Additionally, the presence of ODD in the setting of papilledema does not portend worse visual outcome. Our investigation supports the theory that conditions causing axonal distress, specifically optic disc edema (papilledema), may contribute to the formation of ODD.

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Keywords: Exposed Optic Disc Drusen, Idiopathic Intracranial Hypertension, Papilledema, Pseudotumor Cerebri, Vision Outcomes

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 119**To Evaluate Changes in Retinal Nerve Fibre Layer and Ganglion Cell Layer on SD-OCT in cases of Multiple Sclerosis and Optic Neuritis**

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Introduction:

To prospectively evaluate retinal nerve fibre layer (RNFL) and ganglion cell layer (GCL) changes over 6 months in multiple sclerosis and optic neuritis patients and correlate them with visual functions and disability.

Methods:

Patients with multiple sclerosis without optic neuritis (MS, n=40 eyes), optic neuritis (ON, n= 58 eyes), Multiple sclerosis with optic neuritis (MS+ON, n=48 eyes), and disease-free controls (n=40 eyes) were included. All subjects underwent visual testing(VA), contrast sensitivity, colour vision, visual evoked response, retinal nerve fibre layer thickness (RNFL) and ganglion cell layer thickness (GCL) measurement using spectral domain optical coherence tomography (SD-OCT) at baseline and 6 months. Expanded disability status scale (EDSS) score was calculated for multiple sclerosis patients.

Results:

Compared to controls average RNFL thickness was reduced significantly in affected eyes of MS+ON(p<0.005) unaffected fellow eye (p<0.005) and ON (p<0.005) but no significant difference was found in MS patients. Compared to controls GCL thickness was reduced significantly in eyes of MS+ON(p<0.005), unaffected fellow eye(p<0.005), ON (p<0.005) as well as MS patients (p<0.005). In MS patients GCL thickness correlated significantly with VA (p=0.0032), colour (p<0.001), EDSS (p=0.0047), VER amplitude (p=0.0078) and latency(p=0.0001). Significant correlation of GCL was observed with VA(p=0.0042), colour vision (<0.001) and contrast (p=0.02) in ON. GCL and RNFL loss in M.S+O.N and O.N patients and GCL loss in M.S patients over 6 months was significant as compared to age matched controls (p<0.005).

Conclusions:

Ganglion cell layer thickness may be a more sensitive marker of involvement in Multiple sclerosis and optic neuritis patients. Although eyes with history of optic neuritis demonstrate the greatest reduction in GCL thickness, MS non-ON eyes are also affected suggesting sub-clinical involvement causing chronic axonal loss in MS patients. Ganglion cell layer clinically correlated with EDSS and maybe used as a marker for disease progression.

References: None.

Keywords: SD-OCT, Ganglion Cell Layer Thickness, Visual Function, Optic Neuritis, Multiple Sclerosis

Financial Disclosures: The authors had no disclosures.

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Poster 120

Sensitive Assessment of Acute Optic Neuritis by a New Digital Flicker Test

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Introduction:

ON is primarily a clinical diagnosis but clinical tests with increased sensitivity, and tests which predict MS risk in the ON patients, are needed. The Aulhorn flicker test was shown in the 1980's to effectively diagnose acute ON¹. The aim of this study was to examine the applicability of a new, digitalized version of the analog test for examination of acute optic neuritis (ON).

Methods:

A psychophysical test where the subjective brightness of a steady field has to be adjusted to that of a flickering field of randomized varying frequencies 0-60 Hz. Recorded luminances are then plotted against frequencies. The normal curve shows a light-enhancement at medium frequencies (Brücke-Bartley-effect (BBE)^{2,3}) whereas the ON curve is hypothesized to show darkness enhancement and a cancelled BBE at these frequencies. Visual acuity (VA, ETDRS) and flicker test measurements were obtained in acute ON patients (within 1 month of onset). 52 consecutively referred, untreated ON patients were included (bilateral symptoms in 5/52).

Results:

Mean logMAR VA was 0.52 (SD:0.58) and mean age 37.1 year (SD:11.2). The flicker test was performed 22 days (SD:15) following ON onset. 49 of 52 patients showed abnormal response (cancelled BBE, darkness enhancement) corresponding to 94.23 %. 20/52 patients were reexamined 3 months following ON onset where 7/20 showed normal response (1/20 in the acute phase) which may indicate a dynamic response of the flicker test at different time points following ON onset.

Conclusions:

Preliminary results indicate very good sensitivity of the digital flicker test in ON. Follow up of the ON patients, and a general MS patient population, may further provide evidence of an accurate and easy to use tool in diagnosing acute ON and signs of demyelinating disease in the visual pathway.

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Keywords: Optic Neuritis, Clinical Testing, Psychophysics, Demyelinating Disease

Financial Disclosures: The authors had no disclosures.

Grant Support: The study was supported by the Danish Multiple Sclerosis Society.

Poster 121**Factors Associated with Papilledema in Children with Hydrocephalus**

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Introduction:

To report the factors associated with papilledema in children with hydrocephalus.

Methods:

The medical records of patients under 15 years old who were diagnosed with hydrocephalus and treated by ventricular drainage or ventriculo-peritoneal shunt operation from January 2005 to July 2015 were retrospectively reviewed. Factors including age, gender, etiology of hydrocephalus, duration of signs or symptoms, intracranial pressure (ICP) and presence of papilledema were investigated.

Results:

Thirty-eight patients were included. Their mean age was 5.5 ± 4.6 years and 19 patients (50%) showed papilledema. The mean age of patients without papilledema was 3.0 ± 3.3 years and 7.9 ± 4.4 years in patients with papilledema ($p=0.000$). The ICP was 19.8 ± 11.8 cmH₂O in patients without papilledema and 33.1 ± 9.3 cmH₂O in patients with papilledema ($p=0.000$). The mean duration of symptoms was 3.2 ± 4.5 months in patients without papilledema and 3.9 ± 4.4 months in patients with papilledema ($p=0.621$). The causes of hydrocephalus were tumor (50%), congenital anomaly (24%), hemorrhage (16%) and infection (10%). The groups with brain tumor had older age, higher intracranial pressure and showed papilledema more commonly than others.

Conclusions:

In patients with older age, higher intracranial pressure, and hydrocephalus induced by brain tumor, papilledema was more common. However, since there were 50% of patients without papilledema in children with hydrocephalus, the absence of papilledema does not ensure the absence of hydrocephalus, especially for younger patients.

References: None.

Keywords: Pediatric Neuro-Ophthalmology, High Intracranial Pressure/Headache

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 122

Postural Effects on Intraocular Pressure and Ocular Perfusion Pressure in Patients with Non-Arteritic Anterior Ischemic Optic Neuropathy

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Introduction:

The aim of this study was to investigate postural effects on intraocular pressure (IOP) and ocular perfusion pressure (OPP) in patients with non-arteritic ischemic optic neuropathy (NAION).

Methods:

Twenty patients with unilateral NAION underwent IOP and blood pressure (BP) measurements 10 minutes after changing to each of the following positions sequentially: sitting, supine, right lateral decubitus position (LDP), supine, left LDP, and supine. IOP was measured using a rebound tonometer. OPP was calculated using formulas based on mean BP. The dependent LDP was defined as the position when the eye of interest (affected or unaffected eye) was placed on the dependent side in the LDP.

Results:

In both the affected and unaffected eye, mean IOP was significantly increased and OPP significantly reduced when changing position from sitting to supine (all $P < 0.05$), but inter-eye comparisons showed no statistical difference (all $P > 0.05$). The affected eye showed a higher IOP in the dependent LDP compared to the unaffected eye-dependent LDP ($P < 0.001$). Compared with the IOP of the unaffected eye, the mean IOP of the affected eye increased significantly ($+2.89 \pm 4.40$ versus $+0.65 \pm 3.14$ mmHg, respectively; $P = 0.003$), and the mean OPP decreased significantly (-6.67 ± 9.37 versus -4.94 ± 7.98 mmHg, respectively; $P = 0.022$) after changing from supine to dependent LDP.

Conclusions:

Postural change from supine to dependent LDP may significantly increase IOP and decrease OPP in the affected eye. Posture-induced IOP changes could be a predisposing factor for NAION development.

References: None.

Keywords: Optic Neuropathy, Vascular Disorders, Visual Fields

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 123**Risk Factors for Radiation-Induced Optic Neuropathy: A Case-Control Study**

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Introduction:

Radiation-induced optic neuropathy (RION) is a rare complication of radiation therapy characterized by acute, painless vision loss many months after treatment. Various factors which appear to increase the risk of RION within determined safe-dosage thresholds have been described; however, these findings have been inconsistent and primarily based on descriptive case series.

Methods:

This study applied a case-control design to interrogate potential risk factors for RION. Patients with a diagnosis of RION established by ophthalmologic testing (n=14) were matched in a 2:1 ratio to controls according to maximum point dose to the affected optic nerve. Patient characteristics (age, gender, race, vascular risk factors, tumor profile) and treatment parameters (cumulative and fractionated dose volume and schedule) were analyzed for potential risk factors.

Results:

No significant association was found for diabetes (p=0.66, OR=1.84, 95% CI [0.35-9.61]) or hypertension (p=0.69, OR=1.3, 95% CI [0.35-4.8]). Odds ratios suggest hyperlipidemia may pose an increased risk (p=0.28, OR=2.31, 95% CI [0.56-9.48]), as may male gender (p=0.11, OR=2.85, 95% CI [0.77-10.57]). Age was not a significant risk factor (p=0.94, MD=1.04). A potentially increased risk was found for prior visual complaint (p=0.1, OR=3.04, 95% CI [0.78-11.81]) and tumor impingement of the optic nerve (p=0.95, OR=2.11, 95% CI [0.59-7.61]). Concurrent chemotherapy (p=0.74, OR=0.63, 95% CI [0.16-2.48]) and primary surgery (p=1, OR=0.86, 95% CI [0.23-3.23]) were not significant risk factors.

Conclusions:

More rigorously identifying risk factors for RION may decrease the incidence of this profound, irreversible outcome. Our study found a potential increased risk for male gender, hyperlipidemia, prior visual complaint, and tumor impingement of the optic nerve. While these individually may play some role, no single trait proved predictive when controlling for maximum radiation dose to the affected optic nerve, suggesting that radiation dose may be the overriding determinant.

References: None.

Keywords: Chemotherapy and Radiation Injury, Optic Neuropathy

Financial Disclosures: The authors had no disclosures.

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Poster 124**Number and Distribution of Causes of Optic Neuropathy in Patients Evaluated at a Tertiary Care Center**

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Introduction:

Optic nerve dysfunction is among the most common causes of vision loss in the industrialized world. The range of diseases that cause optic neuropathies is quite large, although the great majority of optic neuropathies are caused by a relatively small number of etiologies. The purpose of this study was to assess the total number and distribution of causes of optic neuropathies for patients who were evaluated at a tertiary care center, and to determine the frequency of patients who had optic neuropathies without identifiable cause despite current state-of-art diagnostic studies.

Methods:

A cross-sectional study of all patients who were evaluated by our Neuro-Ophthalmology service from July through October 2015 and found to have an optic neuropathy. For each patient, data was collected on the medical history, visual presentation, findings on neuro-ophthalmic examination, as well as the results of diagnostic studies that were obtained to search for an etiology. No diagnostic studies were performed that would not have otherwise been performed as part of our standard clinical evaluation.

Results:

A total of 165 patients were found to have an optic neuropathy. Of these patients, 41 (25%) had ischemic optic neuropathy, 30 (18%) had idiopathic intracranial hypertension, 23 (14%) had optic neuritis, and 8 (5%) had an identifiable genetic cause. In 15 (9%) patients, the etiology could not be determined. Of the 15 patients, 10 had bilateral optic nerve involvement.

Conclusions:

The most useful result of this study is the proportion of patients for whom an etiology of optic neuropathy could not be determined. This subgroup represents a potential target for future investigation, especially genetically, to identify new explanations for visual loss. The results of this ongoing study will be compared to results obtained in an identical way from other tertiary care centers around the world.

References: None.

Keywords: Optic Neuropathy, Ischemic Optic Neuropathy, Idiopathic Intracranial Hypertension, Optic Neuritis, Hereditary Optic Neuropathy

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 125**Short Follow-Up Bias Confounds Estimates of the “Typical” Clinical Course of Susac Syndrome**

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Introduction:

The prevailing view regarding the clinical course of Susac syndrome (SS) is that it is generally monocyclic and shows spontaneous resolution within approximately 2 years. However, the duration of follow-up in existing case reports may be inadequate to judge the veracity of these assumptions.

Methods:

We identified all reported cases of SS between 1973 and October 2015. If the duration of follow-up was not explicitly stated, we calculated it based upon provided information regarding the initial symptoms, clinical course during observation, and submission date of the published report. A course of up to 24 months without subsequent relapses was labeled “monocyclic”, whereas a relapse beyond 24 months indicated a “polycyclic” course. We performed descriptive analyses of the duration of reported follow-up for all patients.

Results:

We identified 185 reports, in which a total of 416 individual cases of SS were reported. Of these, the duration of follow-up was determined in 258 cases. It ranged from 0.5 to 312 months. The mean (\pm SD) was 42.2 ± 53.2 months, but the distribution was positively skewed. The median duration of follow-up was only 24 months. Classification of the clinical course as monocyclic or polycyclic was possible in 109 cases. A polycyclic course (60 cases, 55%) was associated with longer follow-up period (median 66 vs. 42 months, $P < 0.001$).

Conclusions:

This analysis of all published cases of SS shows that the median reported follow-up period is only 24 months, which is too short to establish a true estimation of the long-term risk of relapsing disease. Short follow-up in published case reports creates an inherent bias toward the impression that SS is a monocyclic, self-limiting disease. A multicenter patient registry with long-term follow-up is essential to determine the true clinical course and risk of relapse.

References: None.

Keywords: Susac Syndrome, Clinical Course

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 126**Neural Cell Adhesion Molecules in Acute Optic Neuritis: Relation to Clinical and Paraclinical Findings**

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Introduction:

Acute optic neuritis (AON) is an inflammatory condition of the optic nerve presumably of autoimmune origin. More than 50 % develop multiple sclerosis (MS). AON patients were chosen to achieve a homogenous patient group in which complete diagnostic work up was performed within one month from onset. No studies have thus far investigated the levels of Neural Cell Adhesion Molecules (NCAM) in the CSF in AON to determine whether NCAM is associated with demographic and paraclinical findings suggestive of MS.

Methods:

We performed a prospective study of NCAM level in the CSF in 50 adults with AON, median age 32 (range 18 to 62) years and healthy controls. Associations between NCAM levels and age, gender, results of brain MRI at 3.0 Tesla and routinely measured biomarkers in the CSF in patients with AON were assessed.

Results:

The median level of NCAM in the CSF was 348 ng/ml in AON, compared to a mean value 412 +/- 109 ng/ml in healthy controls. There was no age and gender difference. There was neither significant association between NCAM and presence of elevated leucocyte count in the CSF, nor elevated IgG index, nor presence of oligoclonal IgG bands in the CSF. No significant association was found between brain MRI and NCAM level, but patients with a normal NCAM level tended to be more likely to also have a normal brain MRI ($p=0.057$, Fisher exact test). The results showed a trend towards increased NCAM and the presence of Gadolinium enhancing lesions on brain MRI albeit not significant.

Conclusions:

The study showed no significant association between NCAM level in the CSF and the results of routinely measured biomarkers in the CSF and brain MRI without and with Gadolinium DTPA in 50 consecutive patients with AON. In a follow up study we will examine whether NCAM levels predict development of MS.

References: None.

Keywords: Demyelinating Disease, Optic Neuropathy, Neuroimaging

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 127

Radiographic and Systemic Prognosticators of Visual Acuity After Indirect Traumatic Optic Neuropathy

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Introduction:

Indirect traumatic optic neuropathy (iTON) is a well-described cause of post-traumatic vision loss thought to result from shearing forces transmitted to the optic nerve in the optic canal in the absence of a canal fracture. Visual recovery is unpredictable. The purpose of this study is to identify characteristics of cranial computed tomography (CT) and systemic comorbidities that may predict visual outcomes in eyes with iTON.

Methods:

This study is a retrospective case series of patients with iTON treated initially at a large, urban trauma center with follow-up at an affiliated ophthalmology clinic. In addition to detailed cranial CT characteristics and demographics, systemic comorbidities, co-injuries, blood products administered, and intracranial pressure (ICP) along with other factors were gathered. LogMAR visual acuity (VA) at initial presentation and up to 12 months follow-up was also collected.

Results:

Thirteen patients met inclusion criteria; 11 men and 2 women with mean age of 35.2 years +/- 15.2. One (8%) patient received high-dose IV steroids at the time of injury. Mean initial VA was 1.6 +/- 1.0 logMAR and final VA was 1.2 logMAR +/- 1.1 at the final visit. Three NLP eyes from 2 patients at the final visit were also NLP at presentation. Of the 25 predictors analyzed, three were found to significantly influence visual outcomes. Patients with worse presenting VA and ICP bolt placement had worse final vision, while patients with systemic immune response syndrome had better final vision. Orbital fractures and intra- and/or extraconal emphysema and/or hemorrhage were not found to significantly affect visual outcome.

Conclusions:

In this small case series, initial visual acuity and need for ICP bolt placement predicts poorer visual recovery in patients with iTON. The relationship between SIRS and vision after iTON maybe due to high correlation between ICP bolt placement and SIRS. Further studies are needed to investigate.

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Keywords: Optic Nerve Trauma And Treatment, Optic Neuropathy, Trauma, Neuroimaging

Financial Disclosures: The authors had no disclosures.

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Poster 128

Optic Atrophy in Classical Methylmalonic Acidemia

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Introduction:

Methylmalonic acidemia (MMA) is an autosomal recessive, metabolic failure to process various amino acids and lipids. Classical MMA results in methylmalonyl CoA mutase deficiency, preventing the vitamin B12-dependent conversion of methylmalonyl CoA to succinyl CoA, required in Krebs cycle. Patients typically present in early infancy with lethargy and failure to thrive. Long-term complications include renal failure (CRF) and encephalopathy. Only 4 cases of optic atrophy (OA) have been reported. With improved survival of patients offered advanced treatment, OA needs to be identified so that prophylactic/therapeutic intervention, when available, can be incorporated into management protocols. The purpose of this study is to identify and determine the prevalence of OA in classical MMA.

Methods:

22 patients diagnosed and genetically confirmed to have classical MMA were assessed neuroophthalmically. Diagnosis of OA was determined by a combination of visual acuity, pupil reactions, optic nerve appearances, optical coherence tomography (OCT) and VEP. Ophthalmic and medical data were tabulated and associations determined using Mann-Whitney U, Kruskal-Wallis, Chi-squared and Fisher's exact tests. Statistical significance was set at $p < 0.05$. Patients with propionic acidemia and intracellular cobalamin metabolism disorders, which have similar clinical features, were excluded.

Results:

8 patients were female and 14, male. Age range was 7 to 27yrs (median=14;IQR=11-16).13(59%) patients had OA; 85% of these were bilateral. 6 (46.15%) reported decreased vision and 7(53.85%) were asymptomatic.12 patients had CRF (median=16;IQR=14.5-20). Age was not significantly associated with OA ($p=0.17$) but was significantly related to CRF ($p=0.0067$). Patients with OA were more likely to have CRF ($p=0.0058$).

Conclusions:

Optic atrophy is a frequent finding in classical MMA and commonly is bilateral and sub-clinical. A positive correlation with CRF, known to be associated with OA, suggests a causal relation or common pathogenesis. These findings have important management implications. Early and periodic ophthalmic assessments should be performed in all, including asymptomatic patients with classical MMA.

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Keywords: Optic Atrophy, Methylmalonic Acidemia, Chronic Renal Failure

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Poster 129

Structure-Function Analysis in Chiasmal Compression Helps Predict Visual Prognosis

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Introduction:

Sellar mass compression of optic chiasm is often an emergency, and there is a relative lack of data helping clinicians predict visual outcome. To address this problem, we performed structure-function correlation of automated perimetry and optical coherence tomography measurements after treatment of chiasmal lesion.

Methods:

We performed a retrospective, cross sectional study of >100 patients with sellar mass at one institution from 2003 to 2015 and analyzed clinical characteristics and chronic, post-operative (range 3m-16y) paired perimetry and optical coherence tomography (OCT) measurements of retinal nerve fiber layer (RNFL) and macular ganglion cell complex (GCC), which includes the ganglion cell and the inner plexiform layers. For statistical analysis, we used SPSS ver.20.

Results:

We performed *post-operative* structure-function correlation with 64 eyes in 33 patients. Despite the relatively good visual acuity (81% of eyes with 20/30 or better), persistent visual field loss was found in 61% (bi-temporal 42%, unilateral-temporal 18%). Retinal thinning as defined by OCT measurement at 5-percentile of normal was found in 45% of eyes per RNFL (mean 80.2±1.9 microns, 64 eyes) and in 55% of eyes per GCC (mean 69.5±1.5 microns, 62 eyes), consistent with irreversible retrograde degeneration. Using re-calculated visual field mean deviation and GCC (nasal=crossed, temporal=uncrossed), we best fit the structure-function correlation using formula: thickness = slope * $10^{0.1 * (\text{mean deviation in dB})}$ + residual thickness.¹ The temporal-superior field had residual thickness of 43-microns and the temporal-inferior field, 48-microns, consistent with chiasmal compression from below. Using ROC analysis, nasal GCC ≥55-micron significantly predicted good temporal visual field outcome (mean deviation better than -10 dB)¹ (P<0.001, area under curve 0.914). The sensitivity and specificity using these criteria were 89.1% and 93.7%.

Conclusions:

Timely treatment of severe optic chiasm compression can often lead to good visual outcome. A GCC thickness of ≥55 micron correlated with good post-operative visual outcome.

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Keywords: Chiasmal Compression, Optical Coherence Tomography, Ganglion Cell Layer, Diagnostic Test, Visual Prognosis

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 130**Effect of Intraocular Pressure on Optic Nerve Structure and Function in Adults with Optic Nerve Head Drusen**

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Introduction:

To determine whether the intraocular pressure (IOP) in patients with optic nerve head drusen (ONHD) correlates to the mean deviation (MD) on automated visual fields (VFs) and the mean retinal nerve fiber layer (RNFL) thickness on optical coherence tomography (OCT).

Methods:

This retrospective chart review included adults with ONHD from two academic medical centers. Inclusion criteria were age over 18, definitive diagnosis of ONHD, and a VF within 3 months of diagnosis. Exclusion criteria were unreliable VFs and comorbidities that could alter VF results. Data were collected from the initial visit associated with the diagnosis of ONHD. Age, IOP, location of drusen (i.e. surface vs. buried), method of diagnosis, mean RNFL thickness, and MD were recorded.

Results:

A total of 239 patients were identified. Of 97 patients, 154 eyes met inclusion criteria. Mean age was 40.4 years (range, 19-76 years). Average mean deviation was -5.2 dB (range, -27.15 to +0.32 dB). Mean IOP was 15.9 mmHg (range, 6-23 mmHg). Forty eyes (25.9%) underwent Spectralis™ RNFL measurement; mean RNFL thickness was 80.8 micrometers (range, 43-117 micrometers).

Unexpectedly, there was a significant association between lower IOP and worse MD ($p=0.05$), however after adjusting for age this finding is no longer significant ($p=0.12$). There was no statistically significant association between IOP and RNFL thickness ($p=0.61$) in this study.

Conclusions:

Some have proposed that lowering IOP in patients with ONHD may prevent progression of optic neuropathy.¹ To our knowledge, this is the first large-scale study to investigate whether a correlation between IOP and optic nerve structure and function exists in patients with ONHD. This study suggests that lowering IOP may not be beneficial in this disease, as higher IOPs are not associated with either a worse visual field deviation or a thinner RNFL in this group of patients.

References:

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Keywords: Disc Drusen, Intraocular Pressure, Visual Fields, Ocular Imaging, Optic Nerve

Financial Disclosures: The authors had no disclosures.

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Poster 131**Electrical Fields Direct Retinal Ganglion Cell Axon Growth**

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Introduction:

Restoration of vision in patients with advanced optic neuropathies such as glaucoma requires regenerating the optic nerve (ON). A major hurdle to ON regeneration was the lack of available retinal ganglion cells (RGCs). Now that advances in stem cell biology readily allow for the production of RGCs, the rate-limiting step to ON regeneration becomes directing axons of newly transplanted cells from the retina their distant targets. While much of the current efforts to address this problem attempt to recapitulate ON development, studies suggest that an exogenous signal is needed. The body has naturally occurring electrical currents and many tissue culture experiments have shown that cells grow directionally in an electrical field. The effects of an electrical field on RGC growth, however, have never been tested. This project addresses the question of whether electrical fields can be used to direct the growth of transplanted RGC axons.

Methods:

Retina was isolated from post-nasal mice and cultured in an electrotaxis apparatus. One hour after plating, retina was exposed to various electrical fields for 6 hours. Time-lapsed microscopy was performed to record neuron and neurite movement.

Results:

In control cultures, cells and axons migrated away from the explant edge indiscriminately. In contrast, explants exposed to a continuous electrical field of 50mV/mm demonstrated no neuron or axon migration from the explant edge facing the cathode. Numerous cells and neurites were seen moving away from the explant facing the anode. Quantification of these experiments revealed a decrease in the number of RGCs that migrated towards the cathode compared to the anode.

Conclusions:

These experiments demonstrate that RGCs and/or RGC axons migrate directionally in an electrical gradient. Although further experimentation is needed, these experiments suggest that there may be a potential role for electrical currents in facilitating ON regeneration and the restoration of vision in patients with advanced optic neuropathies.

References: None.

Keywords: Orbital/Ocular Pathology, Optic Neuropathy

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 132**Long-Term Evolution of Optic Disc Drusen – 57 Year Follow-Up**

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Introduction:

Optic disc drusen (ODD) are hyaline deposits in the optic nerve head. The pathophysiology remains unknown, but the formation of ODD is probably based on alterations of axoplasmic transport in the optic nerve head. Long-term evolution of ODD anatomy and visual field defects in ODD patients is a key factor for learning more about the pathophysiology and prognosis of the condition. With a mean follow-up period of 57 years, this is the first study that evaluates optic disc anatomy and visual fields in patients with ODD over a life-span.

Methods:

Funduscopy pictures and visual fields from 7 ODD patients with known hereditary ODD were compared with 57 years between examinations. Digitalized quantification was used to assess progression of visual field defects examined by Goldmann perimetry.

Results:

Mean age at initial examination was 17.4 years. Mean age at follow-up examination was 73.9 years, resulting in a mean follow-up time of 56.7 years. When comparing funduscopy pictures from the initial and follow-up examinations, a minimal or non-existing change in ODD anatomy was seen in 12 out of 14 patients. However, there was a tendency towards more anatomical change in subjects younger at first examination. A 15.8% decrease in Goldmann visual field area (cm²) using digitalized quantification was found between initial and follow-up examination.

Conclusions:

Minimal or non-existing change in optic disc anatomy and visual fields in ODD patients oldest at the initial examination suggest that anatomical progression of ODD in patients with hereditary ODD happens primarily before adulthood and then ceases. A slow progression of visual field defects are seen throughout life, but mainly occurs in younger age.

References: None.

Keywords: Optic Disc Drusen, ODD, Visual Fields, Perimetry

Financial Disclosures: The authors had no disclosures.

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Poster 133

Early Treatment with Levodopa Promotes Visual Improvement in Nonarteritic Anterior Ischemic Optic Neuropathy

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Introduction:

To determine the clinical effectiveness and potential neuroprotection of levodopa in improving visual acuity, visual field, and retinal nerve fiber layer (RNFL) thickness in eyes affected by NAION.

Methods:

Retrospective cohort study involving 59 eyes of 59 participants with NAION evaluated within 15 days of NAION onset. Participants received 25mg carbidopa/100mg levodopa three times daily with meals for 12 weeks (Levodopa group) or untreated (Control group). Best corrected visual acuity converted to logMAR, mean deviation (MD) threshold sensitivity on automated perimetry, and mean RNFL thickness on optical coherence tomography (OCT) were assessed. The primary outcome was the categorization of eyes into improved visual acuity (by 0.3 logMAR difference), worsened visual acuity (by 0.3 logMAR difference), or no change in visual acuity. The proportions in each category were compared between the Levodopa and Control groups.

Results:

Among participants with 20/60 or worse initial visual acuity, Levodopa-treated participants had significant improvement ($P < 0.0001$) in the mean change from initial to final logMAR visual acuity of -0.74 ± 0.56 (95% CI, -0.98 to -0.50), while the mean change for the Control group at -0.37 ± 1.09 (95% confidence interval estimate, -1.00 to $+0.26$) was not significant ($P = 0.23$). A significant difference between groups was observed ($P = 0.0086$) such that 18/23 (78%) in the Levodopa group improved and none got worse, as compared with 6/14 (43%) in the Control group improving while 3 (21%) worsened. The change in visual field MD and RNFL thickness on OCT showed no significant difference at $P = 0.23$ and $P = 0.75$, respectively. No levodopa treated participant had any adverse event from the levodopa.

Conclusions:

Treatment within 15 days of onset of NAION with levodopa improved central visual acuity by an average of 6 lines on Snellen acuity chart. Levodopa may promote neuroprotection of the maculopapular retinal ganglion cell fibers in NAION.

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Keywords: Levodopa, Nonarteritic Anterior Ischemic Optic Neuropathy NAION, Neuroprotection, Dopamine, Optic Nerve

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Poster 134

Comparison of Critical Flicker-Fusion Threshold Between Patients with Either Demyelinating or Ischemic Optic Neuropathy

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Introduction:

Critical flicker-fusion (CFF) threshold is the frequency at which a flickering light is indistinguishable from a steady, non-flickering light. CFF is a well-established test to show optic neuropathy due to demyelination and other causes. The purpose of this study was to compare CFF threshold results between patients with demyelinating disease and ischemic causes of optic neuropathy. We hypothesize that demyelinating disease would have a more profound and more consistent effect on CFF compared to ischemic disease.

Methods:

We performed a retrospective review for eyes with demyelinating or ischemic optic neuropathies. Multiple independent 2-sided T-tests and Z-tests were used to compare eyes with demyelinating optic neuropathy (Group 1) from ischemic optic neuropathy (Group 2). A multivariate linear regression was performed for the dependent variable CFF.

Results:

The mean CFF value for Group 1 (20.7 Hz) and Group 2 (24.3 Hz) were not significantly different from each other (P=0.06). However, both CFF values were significantly less than fellow eyes from these patients (30.9 Hz) with no optic neuropathy (P<0.01). Roughly 71% of Group 1 and 37% of Group 2 had a CFF value of <24 Hz (P<0.01). The strongest factors found to contribute to the CFF value were: the type of optic neuropathy, age, duration, vision, and mean deviation by automated perimetry. Within this model Group 1 contributed to usually a CFF about 8.2 Hz lower than Group 2 (P<0.01).

Conclusions:

The mean critical flicker fusion value in eyes with demyelinating vs. ischemic optic neuropathies approach a statistically significant difference. A large majority of eyes with demyelinating disease have a CFF <24 Hz compared to eyes with ischemic disease. The variable with the greatest impact on CFF in our model was having demyelinating optic neuropathy. CFF may be a useful adjunct in distinguishing between demyelinating from ischemic optic neuropathy when the diagnosis is uncertain.

References: None.

Keywords: Demyelinating Disease, Diagnostic Tests, Optic Neuropathy, Neuro-Ophthalm & Systemic Disease

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Poster 135

Correlation Between Stereopsis and Reverse Stereopsis: "Turn Upside-Down the Stereo"

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Introduction:

Stereopsis is a binocular phenomenon dependent on higher order visual processing. We previously reported that although stereopsis can reliably suggest a range of visual acuity (VA) in patients with no history of strabismus¹, it cannot be used to establish a given VA. Michael Brodsky, M.D.² suggested that reverse stereovision testing in patients with bitemporal hemianopias demonstrates absence of stereopsis. The goal of this study was to compare the results of stereopsis and reverse stereopsis in patients without visual field (VF) defects before testing Dr. Brodsky's hypothesis.

Methods:

Patients seen in a neuro-ophthalmic service over a 6 month period underwent a detailed examination, including polarized vectogram stereoacuity measurements (Titmus) and VF. Patients were first tested with the stereoacuity book upright (traditional stereoacuity, "TS") and then with the stereoacuity book rotated 180 degrees (reverse stereoacuity, "RS").

Results:

Of 495 patients tested, we included the 98 (median age, 45 years [13-79]) who had normal VF, no ocular misalignment, and >0 circles on TS. Among these 98 patients mean score was 7.8/9 circles correct on TS and 7.5/9 on RS [p-value 0.07, paired two-tailed t-test]. 26/98 patients (26.5%) scored higher on TS than on RS (average difference, 2.3 circles), 16 patients (16.3%) scored higher on RS than on TS (average difference, 2.0 circles), and 56 patients (57.1%) scored the same on both. 64/98 patients (65.3%) scored 9/9 circles correct on TS; of those 64, 54 (84.4%) scored 8/9 or 9/9 circles, 5 (7.8%) scored 7/9 circles, 2 scored 6/9, and 1 patient each scored 5/9, 3/9, and 2/9 on RS.

Conclusions:

In a patient population presenting for neuro-ophthalmology evaluation with normal VF and without strabismus, there is no difference in TS and RS testing. Therefore, any difference found in patients with homonymous or heteronymous VF defects may relate to the VF defect itself.

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Keywords: Stereopsis, Perimetry, Visual Acuity

Financial Disclosures: The authors had no disclosures.

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Poster 136**Improved Visual Outcomes in Giant Cell Arteritis from Prior Use of Corticosteroids May Be Dose-Dependent**

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Introduction:

The literature suggests high-dose steroids have a protective effect in preserving vision in giant cell arteritis (GCA), however, the optimal dose has not been well elucidated. Clinical observation has noted that prior use of corticosteroids may result in less GCA-related visual dysfunction. Our hypothesis was that patients with corticosteroid use prior to visual dysfunction had a dose dependent benefit with improved visual outcomes in comparison to patients not on prior corticosteroids.

Methods:

A retrospective analysis was performed of GCA patients presenting to neuro-ophthalmology clinic between 2005-2013, all satisfying the 1990 ACR-GCA criteria. Patients were divided into two groups based upon whether they were on corticosteroids at the time of ocular manifestations or not. Data was collected from the most reliable baseline neuro-ophthalmic examination. Each eye was analyzed independently. Visual acuities were recorded using a modified LogMar scale. Visual fields were assessed by mean deviation and quadrants involved.

Results:

78 patients were analyzed (65% women, mean age 75 years), 69 of whom underwent temporal artery biopsy (84% positive). 13 patients (17%) were on corticosteroids at the time of visual manifestations. Those on greater doses of corticosteroids tended to have better visual acuity outcomes with a negative correlation between the LogMar scale and corticosteroid dose (spearman rho -0.31; P=0.002). This association remained in linear regression models when accounting for prior eye disease and aspirin use (p=0.019). There was also a positive correlation between visual field mean deviations and the corticosteroid dose (spearman rho 0.26; P=0.019), remaining when accounting for confounding aspirin use (p=0.024).

Conclusions:

Prior use of corticosteroids resulted in improved visual outcomes in GCA-related visual dysfunction. This appears to be a dose-dependent effect seems to be present as demonstrated in our analyses of the visual field mean deviations and visual acuity. Larger scale studies may provide supporting evidence for the optimal dose of steroids to protect from GCA-related vision loss.

References: None.

Keywords: Giant Cell Arteritis, Optic Neuropathy, Corticosteroid Use

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 137**Reconstructive and Destructive Process in Traumatic Injuries Optic Nerve After High Dose of Corticosteroids Use**

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Introduction:

The research on the possibilities of regeneration of the optic nerve turns attention by scientist for many decades. The aim was to activate regenerative processes in the optic nerve traumatic injury at high doses of corticosteroids.

Methods:

Six mature rabbits (group injured) males, weighing 3.5-4 kg Soviet Chinchilla breeds were used as an experimental model. Additionally, 6 rabbits (treated group) on the second day after the injury were treated using a / m administration of methylprednisolone 30 mg / kg for 3 days, then the dose is gradually reduced. The control group consisted of 15 intact rabbits. Both groups of animals injured removed from the experiment using the guillotine after 2 weeks. Cranial part of both eyes the optic nerve was morphological examined. As a control, use the appropriate structure of the control group.

Results:

There were manifested MNF, which MS consisted only of a few blades of myelin, that was expressly ordered in the study of ultrathin cross sections of the right optic nerve with an electron microscope in animals treated group. Young elongated mitochondria with dense matrix and ordered cristae were founded in axoplasm of MNF. The moderate amount of microtubule clearly structured and neurofilamentary were indicated the recovery of backbone and conductivity of the neural fibers. The large nucleus with dispersed chromatin and multiple clear nucleolemas shallow invagination of young mitochondria and clearly arranged tank granular endoplasmic reticulum detected in the cytoplasm of neurolemocytes. Many ribosomes situated on its surface. These morphological features are evidence of active electric utility and biosynthetic processes that occur in neurolemocytes. Nevertheless, it were present MNF with widespread MS else. There lamellar structure of myelin was disorder.

Conclusions:

Consequently, results showed reduce of microcirculation swelling and remyelination of MNF that improves of activation of damaged optic nerve regeneration by corticosteroids high doses at the 14th day.

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Keywords: Traumatic Injury Of The Optic Nerve, Regeneration, Remyelination, High Doses Of Corticosteroids, Microcirculation

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Poster 138

Optic Coherent Tomography Shows Anatomical-Functional Correlation with Visual Function of Children with Optic Pathway Glioma

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Introduction:

To describe long term follow-up of children with optic pathway glioma (OPG) and to investigate the anatomical-functional correlation between optic nerve function and nerve fiber layer thickness (NFL) as measured by optical coherence tomography (OCT).

Methods:

Medical files of 62 children, 35 with neurofibromatosis-1 (NF-1) and 27 without NF-1, diagnosed with OPG and followed between 2003-2015, were reviewed for demographic, clinical and neuro-imaging data. The correlations between visual function and OCT measurements were analyzed.

Results:

Among 258 NF-1 patients, 35 had OPG detected by routine imaging. Nine of them performed OCT. In the non-NF1 group, all were symptomatic and diagnosed with OPG at a mean age of 3.6 year (5m-17years). Mean follow-up was 6 years. Among 27 non-NF-1 children, thirteen performed OCT exam, 10 had more than one exam. In both groups, gliomas involved the chiasm(32), optic nerve(27), optic tract(10) or combination. Two of the NF-1 group and ten in the non-NF-1 had deterioration of vision during the follow-up. Imaging showed enlarged mass and intraventricular hemorrhage in two, and no change in imaging of the others. Mean OCT measurements of the 17 and 23 eyes examined in the NF-1 and non-NF-1 respectively were 85u and 62u. At the end of the follow-up, poor vision was observed in 34% of non-NF-1 (8/23) and 12% of NF-1 (2/17). Repeated OCT in non-NF1 showed thinning of RNFL in 18/20 eyes, associated with higher logMAR scores ($P < 0.001$), disc pallor, reduced color and visual field defects.

Conclusions:

The majority of NF-1 children with OPG are asymptomatic and stable, while all non-NF-1 were symptomatic. Deterioration of optic nerve function were in tight correlation with repeated OCT measurements but not with neuroimaging. OCT may serve as a better follow-up tool than MRI for children with OPG especially when considering initiating, continuing or stopping chemotherapy.

References: None.

Keywords: Optic Pathway Glioma, Optical Coherent Tomography, Neurofibromatosis, Pediatric Neuro-Ophthalmology

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Poster 139**Update on Treatment of Radiation Necrosis with Bevacizumab**

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Introduction:

Radiation Necrosis of the CNS at times involves the visual pathways, causing irreversible blindness. Standard treatment in the Ophthalmological literature includes treatment with IV Corticosteroids, Hyperbaric Oxygen treatment, and sometimes anticoagulation. Even with these treatments, the visual outcome is generally poor. The Oncologic Literature continues to support the consideration of treatment with Bevacizumab to reverse the effect of the radiation damage

Methods:

An International literature review from 2000 to 2015 was performed, and data collected, that supports the early use of IV Bevacizumab in a patient with CNS Radiation induced Necrosis. Analysis of case reports, small case studies, as well as a Double Blind Placebo Controlled Phase II trial were reviewed and then tabulated. The beneficial effect of this treatment was then compared to reports in the literature of patients treated only with Corticosteroids, versus patients treated with Corticosteroids as well as Hyperbaric Oxygen treatment.

Results:

There is a well-documented benefit of treating patients with IV Bevacizumab who suffer from Radiation Necrosis of the CNS. The improvement was both Radiologic as well as Clinical. The Oncology literature finds radiographic improvement on post treatment MRI imaging including on the post contrast T1-weighted MRI (reduction in abnormality by 48%), fluid-attenuated inversion-recovery sequences (by >50 decrease), and FLAIR images (by 50-64% reduction in volume and abnormality. The safety profile is also acceptable.

Conclusions:

Radiation optic neuropathy is a devastating form of vision loss that is usually treated with steroids and hyperbaric oxygen. Generally the visual outcome remains very poor. The Oncologic literature continues to support the treatment of this condition with IV Bevacizumab. Ophthalmologists and Neurologists, unaware of this literature, may miss an early treatment window for this condition with this alternative treatment. We believe it is the delay in treatment with this medication that limits its usefulness.

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Keywords: Radiation Necrosis, Blindness, Bevacizumab, Hyperbaric Oxygen Treatment, Optic Neuropathy

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Poster 140**A Photothrombotic Model of Optic Neuropathy Using Mesoporphyrin IX: Our Experience with Rats vs. Mice**

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Introduction:

We recently developed a photothrombotic model of optic neuropathy in rodents using mesoporphyrin IX as the photosensitizer. While originally considering similar laser parameters for both rats and mice, our experiments showed that the optic nerve (ON) response to the photodynamic treatment applied was very different between the two species. We present here some of the challenges faced, especially when trying to set up this model in mice.

Methods:

Laser (532nm) was applied to the ON of Brown Norway rats and C57BL/6 mice before or after an intraperitoneal injection of mesoporphyrin IX. Various laser parameters were tested. Funduscopy, fluorescein angiography (FA) and spectral domain optical coherence tomography (SD-OCT) were performed at different time points to evaluate the result of the laser applied.

Results:

The laser power output and duration of laser treatment were found to be the most significant factors in determining the type and extent of ON lesion induced. Laser parameters previously used to induce ON ischemia in rats - ie. 50mW, 12sec, 500um - successfully led to ischemic injury after mesoporphyrin IX injection, as determined by the fluorescein leakage and retinal edema shown on the FA and OCT, respectively. On the contrary, in mice, application of 50mW laser for 3 seconds using a 300um spot proved to be too powerful, leading to occlusion of major retinal vessels even without having administered any mesoporphyrin IX.

Conclusions:

Photodynamic therapy with Mesoporphyrin IX can lead to ischemic ON injury in rats without much difficulty. On the other hand, standardizing the laser parameters to be used in order to induce the desired ON lesion in mice proved to be quite challenging. Given that laser treatment alone can have extensive vasoocclusive effects in mice, it is advisable to thoroughly examine laser parameters prior to commencing projects using with photothrombotic models.

References: None.

Keywords: Ischemic Optic Neuropathy, Photodynamic Treatment, Mesoporphyrin IX, Animal Model

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 141**The Photopic Negative Response as an Objective Measurement of Visual Function in Patients with Leber's Hereditary Optic Neuropathy**

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Introduction:

The photopic negative response (PhNR) is a slow negative component of a flash photopic full field ERG which has been shown to be specific for retinal ganglion cell (RGC) activity. Direct evaluation of RGC function is important in patients with Leber's Hereditary Optic Neuropathy (LHON) in which functional improvement with Idebenone and EPI-743 can take years to be clinically measurable, presumably due to the time required for CNS remodeling. Thus there is a need for objective non-invasive clinical metric in patients with LHON which does not rely on fixation and has the promise of directly measuring RGC activity. The purpose of this study was to evaluate the use of PhNR as a potential clinical metric for patients with LHON.

Methods:

Full field ERG recordings using a sequence of 50 brief red flash stimulus on blue background were collected in eight subjects with LHON using standard ERG recording techniques. The PhNR was identified using a computer based automated detection system and data was manually examined to remove movement artifacts. The Humphrey Visual Field mean deviation and visual acuity was correlated to PhNR amplitudes in patients with LHON.

Results:

The PhNR was recordable in most patients with LHON. The PhNR amplitude decreased with decreasing HVF mean deviation. The PhNR amplitude and HVF were best fit by an exponential decay function ($r^2=0.71$). There was no meaningful correlation between decimal visual acuity and PhNR amplitude ($r^2=0.21$).

Conclusions:

PhNR may be a useful objective clinical metric in patient with decreased visual function and central scotomas from LHON. The strong correlation between HVF mean deviation and PhNR amplitude suggest that this may be a useful objective outcome measure for future clinical trials.

References: None.

Keywords: Optic Neuropahty, Visual Fields, Diagnostic Tests

Financial Disclosures: The authors had no disclosures.

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Poster 142**Neuro-Ophthalmic Manifestations of Costeff Syndrome**

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Introduction:

Type III 3-methylglutaconic aciduria (OPA III) is a neuro-ophthalmologic syndrome consisting of early-onset bilateral optic atrophy caused by mutation c.143-1G>C variant. Since Costeff described phenotype of 19 patients in 1989 several reports described approximately 40 patients, but most of them lack details about neuro-ophthalmic phenotype. Our aim was to characterize the clinical course of the neuro-ophthalmic manifestations of this syndrome.

Methods:

Consecutive patients seen at the neuro-ophthalmology unit were included. All patients underwent comprehensive neuro-ophthalmic evaluation including meticulous visual function history and medical documents review. Best corrected visual acuity, color vision evaluation, visual field testing, and motility, pupillary, slit lamp and dilated fundus examinations were recorded. Optical coherence tomography (OCT) performed whenever possible due to poor fixation.

Results:

Nine patients were evaluated (6 females). Poor vision was the presenting symptom in 4 patients. Average visual acuity (VA) was 1.5 logMAR. Humphrey visual field performed by 5 patients revealed generalized depression with temporal loss. All demonstrated dysmetric saccades. Four had strabismus, 3 exotropia, 1 esotropia. Seven patients demonstrated nystagmus. OCT testing was possible in 6 patients that revealed nerve fiber and retinal thinning. VA correlated with age possibly implying some progression over time.

Conclusions:

This study compiled information regarding neuro-ophthalmic manifestation of OPA III patients. Contrary to established literature poor vision was the presenting symptom in only 50% of our patients. All had decreased VA and variable degree of optic atrophy as previously reported. Nystagmus found as reported previously. Dysmetric saccades found might be the result of the cerebellar atrophy found on MRI of our patients. The thinning of RNFL was expected but the retinal thinning, even in the young patients, might be the result of ganglion loss found also in OPAI. The results may help informing the young patients about visual prognosis. Similarly, OCT may help monitoring experimental therapies when available.

References: None.

Keywords: Optic Neuropathy, Genetic Disease

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Poster 143

Pediatric Optic Neuritis Prospective Outcomes Study

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Introduction:

The Optic Neuritis Treatment Trial (ONTT) provided a vast amount of knowledge with regard to ON in adults. Despite this, there are little prospective data related to pediatric ON(PON). There are differences between adult and PON that may impact visual recovery and the diagnosis of MS. Furthermore, no studies have prospectively evaluated other metrics such as low contrast visual acuity (LCVA), retinal nerve fiber layer thickness (rNFL), or quality of life (QOL) in PON.

Methods:

This study will be conducted by the Pediatric Eye Disease Investigator Group (PEDIG) with cooperation of the Neuro-Ophthalmology Research Disease Investigator Consortium (NORDIC), funded through PEDIG. The pilot study includes two years of enrollment and two additional years of follow-up of up to 100 subjects (3 to <16 years) for prospective data collection to assess the ability to enroll sufficient patients for a future randomized controlled trial, and to evaluate clinical outcomes in this population. The primary clinical outcome measure is visual acuity. LCVA and, if performed, optical coherence tomography of the rNFL and ganglion cell thickness will be collected. In addition, NMO antibody and QOL measures will be analyzed. Treatment decisions will be at investigator discretion.

Results:

The primary outcomes are to (1) assess feasibility for enrollment into a PON protocol, and (2) assess the cohort's visual acuity at 6 months from enrollment. Secondary aims will include the characterization of PON in a multicenter cohort of children, visual acuity at 1 and 2 years, rNFL measurements, and satisfying the diagnosis MS at 2 years.

Conclusions:

This study represents an opportunity for our neuro-ophthalmic and the pediatric-ophthalmic communities to work together to collect data on a rare disease. We are excited at the prospect that this collaboration brings not only for PON but potentially for other rare pediatric neuro-ophthalmic diseases. We welcome participation from all interested members of both NORDIC and PEDIG.

References: None.

Keywords: Pediatric Optic Neuritis

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 144**Visual Outcomes of Diabetics and Prognostic Factors in Non-Arteritic Ischaemic Optic Neuropathy**

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Introduction:

Diabetics have a greater risk of non arteritic ischemic optic neuropathy (NAION) but visual outcomes compared with non-diabetics are unknown. Aims: To compare visual outcomes, prevalence of bilateral /sequential ION and risk factors for NAION in diabetics and non diabetics. To identify prognostic factors for poor visual outcomes in NAION

Methods:

This was a retrospective cohort study of diabetics and non-diabetics with a clinical diagnosis of NAION, presenting within 4 weeks of symptom onset with follow up for >3 months. Using population average logistic regression models, we assessed risk factors for visual prognosis.

Results:

The median follow-up duration was 24.6 weeks in diabetics and 34.5 weeks in non diabetics. 87.5% presented within 2 weeks of symptom onset. In non-diabetics, the most prevalent risk factor was a small disc:cup ratio (70%); for diabetics, they were hypertension (87%), hyperlipidaemia (84%), "disk-at-risk" (51%). 31% diabetics and 21% non-diabetics had sequential NAION. 50% of non-diabetics present/complete follow up with >20/40 VA. 33% of diabetics, present/complete follow-up with >20/40. Similar proportions of diabetics and non-diabetics (17%, 21%) present with or have a final vision of <20/100 (35%, 30%) Final VA in those with >6 months demonstrates no difference between diabetics and non diabetics (p=0.7). Ischemic heart disease (IHD) (OR 5.4 p=0.002) and age (OR 1.5 p=0.05) prognosticate for final VA <20/100. Presence of diabetes does not impact visual outcome. Patients with sleep apnea are less likely to have worse VA (OR=0.22, p=0.04).

Conclusions:

Only 50% of diabetics have a disk-at-risk configuration while this is present in 70% of non-diabetics. The natural history of vision in diabetics with NAION is not significantly different from non-diabetics although they have a higher prevalence of cardiovascular risk factors and higher prevalence of sequential NAION. Of all risk factors, including diabetes, only ischemic heart disease and age independently prognosticate for poor visual outcome.

References: None.

Keywords: Optic Neuropathy, Vascular Disorders

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 145

Categorization and Staging of Retinal Degeneration Using Pathway-Specific Alterations in Retinal Ganglion Cell Stimulus-Response Relationships

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Introduction:

To improve categorization and staging of retinal degenerations, we sought to identify disease-specific alterations in retinal signal processing pathways. We adapted a method to distinguish specific subclasses of ON and OFF responses in retinal ganglion cells (RGCs) using stimulus-response (S-R) relationships (Volgyi et al, J Neurosci 2004) and identifying specific shifts in sensitivity, gain, and dynamic range.

Methods:

We used *in vitro* multielectrode recording to sample ON and OFF responses of individual RGCs to full field flash stimuli of 17 intensities (0-34 $\mu\text{W}/\text{cm}^2\cdot\text{s}$, 1 sec) presented in randomized order, in the retina of 14 day old wild-type (*wt*) and *rd1* mice. For each RGC the number of spikes during the 1 sec following flash on- or offset was plotted vs. stimulus intensity and fit to a Michaelis-Menten relationship, and responses were classified into several types according to parameters extracted from the best fit curves for all cells in each strain.

Results:

In both *wt* and *rd1* mice, ON cells segregate into three and OFF cells into four groups, comparable to mature RGCs (Volgyi et al). For most response groups, *rd1* curves were right-shifted parallel to *wt* curves, but high-sensitivity ON and OFF *rd1* cells had decreased slope (gain) relative to *wt*, whereas low-sensitivity OFF *rd1* cells had a more markedly elevated threshold and steeper slope (gain). This pattern suggests differential changes in photoreceptor input to ON and OFF pathways, or in bipolar and amacrine cell circuits that modify them.

Conclusions:

Shifts in the parameters of RGC S-R curves occur as degeneration progresses in the *rd1* mouse. The nature and degree of these changes differ among subgroups of RGCs, suggesting distinctive signatures for particular forms of retinal degeneration. We are investigating characteristic shifts in S-R curves in other models of retinal degeneration. Ultimately, this method may help diagnose and stage patients with blinding diseases.

References:

Volgyi B, Deans MR, Paul DL, Bloomfield SA. Convergence and segregation of the multiple rod pathways in mammalian retina. J. Neurosci. 24:11182-11192, 2004.

Keywords: Retina, Retinal Degeneration, Ganglion Cells, Electrophysiology, Pediatric Neuro-Ophthalmology

Financial Disclosures: The authors had no disclosures.

Grant Support: March of Dimes Research Award # 12-FY11-200.

Poster 146**Meningiomas of the Optic Nerve. Lessons Learned Over Three Decades**

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Introduction:

Optic nerve sheath meningiomas, considered rare in the past, represent one-third of tumors affecting the optic nerve. The advent of imaging studies has dramatically increased our ability to make this diagnosis usually (without resorting to tissue biopsy). Published case series are small and it can be difficult to draw conclusions based on duration of follow up (especially as slow growing). The role of OCT is evolving.

Methods:

A retrospective review of 31 patients seen with optic nerve meningiomas with long-term (up to 34 year) follow-up. Attention was particularly directed to the results of natural history, radiation therapy, and the use of OCT.

Results:

Most patients were referred for progressive visual loss (several were picked up fortuitously by imaging). Visual acuity at the time of referral ranged from 20/20 to NLP. OCT often showed swelling and "green disease" with either normal or only minimally reduced nerve fiber layer thickness in spite of clear evidence of optic neuropathy (afferent pupillary defect, visual field changes, and decreased central acuity).

Conclusions:

Although the good news about meningiomas is they grow SLOWLY, the bad news about meningiomas is they GROW slowly. Although obviously variable in natural history, most of these patients over time (if untreated) will have worsening visual function. Fractionated radiation therapy (both conventional radiation therapy and proton beam) can result in dramatic improvement in function, reduction of disc edema, and resolution of optic chiasm vessels. While progression in spite of treatment is possible, the vast majority of cases do very well. In view of the immediate effect of radiation therapy on disc swelling, it is probably not necessary to wait for clear progression before instituting radiation therapy. Rare cases of involvement of the optic canal, if ectopic, may be treated with surgery, but this is unlikely to be of benefit in the majority.

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Keywords: Optic Nerve Sheath Meningioma, Sterotactic Radiation Therapy, Proton Beam Therapy, OCT

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 147

Autologous Bone Marrow Derived Stem Cells for Treatment of Neuro-Ophthalmic Disorders: Report of 25 Patients

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Introduction:

The new field of reparative and regenerative medicine has exploded with genetic therapy methods, stem cell therapy options (foreign or autologous fat, skin, bone marrow), and new drug therapies. We report the effect on the vision of 25 neuro-ophthalmology patients with bilateral visual loss using autologous bone marrow derived stem cell (BMSC) therapy, which requires no immunosuppression.

Methods:

Twenty-five patients from the Midwest with bilaterally blinding disorders entered the Stem Cell Ophthalmology Treatment Study (SCOTS), approved by the University of South Florida's IRB, registered with the NIH (NCT01920867 on www.clinicaltrials.gov) to recruit 300 patients (2013-2016). It is funded by the patients. These authors, not a part of that study, agreed to do pre-op and 3,6,9,12 month post-op exams, reducing families' expense and travel time. Data collected included: visual acuity by Snellen and ETDRS, anterior and posterior segment exams, visual fields (Humphrey, Goldmann or both), and OCT measurement of macular and peripapillary RNFL thickness. Each eye was assigned to: Arm 1: periocular (PO) injections BMSC, with intravenous (IV) injections; Arm 2: intravitreal intraocular (IO) injection, plus PO and IV; or Arm 3: vitrectomy plus intra-retinal or intra-nerve injections of stem cells plus PO, IV and IO as above.

Results:

25 patients (47 eyes) were treated: 20 patients with optic nerve disorders, 5 with retinal disorders. 27/47 eyes (58%) gained visual acuity, 13 (28%) neutral (+ or -3 letters), 3 eyes (6%) lost acuity, 4 eyes (8%) too soon to report. Visual fields improved in most eyes. OCT macular thickness peaked at 1 week to 7 months. 13 of 19 genetic disorder eyes improved (68%) with autologous DNA stem cells. 14 of 28 acquired disorder eyes improved (50%). Only 1 of 6 NLP eyes regained bare LP.

Conclusions:

SCOTS benefits: >60% improved vision, 22/25 patients more independent, active, happy.

References:

Weiss, Levy, Benes, Stem Cell ophthalmology treatment study (SCOTS) for retinal and optic nerve diseases: a case report of improvement in relapsing auto-immune optic neuropathy, Neural Regeneration Research, vol 10, pp 1507-1515, 2015.

Keywords: Optic Neuropathy, Retina, Genetic Disorders, Therapy, Autologous Stem Cells

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 148**Clinical Features of Optic Neuritis in Patients over 45 years-old.**

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Introduction:

Optic neuritis typically occurs in younger patients with a peak incidence in the third and fourth decade of life. Clinical and radiographic features of patients with optic neuritis older than age 45 have not been previously published.

Methods:

Retrospective review of cases of optic neuritis in patients older than 45 years-old treated at a tertiary care center from 2003 to 2014. Clinical features, MRI results, Humphrey visual field results were recorded and analyzed. Inclusion criteria are age > 45 years-old, first episode of optic neuritis in a given eye, and recovery defined as improvement of 2 lines of visual acuity or 3 dB on Humphrey automated perimetry. Exclusion criteria included bilateral optic neuritis, steroid dependence, and severe vision loss or mild vision loss without the previously defined recovery.

Results:

We project having 50 patients. To date, 11 patients met the criteria above, of which 7 were female. Average age was 56.7 years-old. 9 of 11 had eye pain. 1 had optic disc swelling. Initial visual acuity was $\leq 20/40$ in 4, 20/50 to 20/190 in 4, and $\geq 20/200$ in 3. Subsequent acuity was $\leq 20/40$ in 10, 20/50-20/190 in 1, and $\geq 20/200$ in 0. Initial automated perimetry in 8 patients showed an average mean deviation of -11.0 dB. Improvement on subsequent automated perimetry in 7 patients was 7.1 dB. MRI was done in 10 cases. The optic nerve showed abnormalities in 7 and the brain showed abnormal T2 lesions in the white matter in 7. Average follow up was 2.0 years with no diagnoses of multiple sclerosis.

Conclusions:

Compared to the Optic Neuritis Treatment Trial, the clinical characteristics were similar. Imaging showed a similar rate of optic nerve abnormalities. Although there were a higher percentage of patients with abnormal brain MRI, no patients progressed to develop multiple sclerosis.

References: None.

Keywords: Demyelinating Disease, Neuroimaging, Visual Fields

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 149

Transcorneal Electrical Stimulation in Optic Neuropathies

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Introduction:

Aim was to achieve improvement in vision and visual fields in optic neuropathies by transcorneal electrical stimulation (TES) .

Methods:

20 patients with non-arteritic anterior ischemic optic neuropathy and 10 patients with traumatic optic neuropathy were stimulated 40 minutes per day for 10 consecutive days by TES. Patients with optic neuropathy were treated 2 months after the acute event.

Results:

In both groups, improvement in vision and visual fields was achieved. The average visual acuity improvement in the two groups was 2 Snellen lines. Visual field improvements after 10 days of TES were documented. Expansion of fields reached approximately 25 percent. This was found to be highly significant. (p: 007)

Conclusions:

TES may be considered as a safe and effective treatment in certain optic neuropathies.

References: None.

Keywords: TES, AION, TON

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 150

Aquaporin 4 Antibody in the Korean Patient with Newly Onset Optic Neuritis

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Introduction:

To evaluate the prevalence of aquaporin 4 antibody (AQP4-Ab) in the newly onset optic neuritis patients and investigate the characteristics of seropositive patients.

Methods:

36 women and six men with newly onset isolated optic neuritis were included in this study between January 2013 and December 2014. AQP4-Ab was detected and all blood samples was obtained prior to treatment and within one week from attack. The patients were sub-grouped into either a seropositive group or a seronegative group according to AQP4-Ab. Differences in age, gender, initial visual acuity, and final visual acuity between groups were analyzed.

Results:

Six (14.3%) of these patients (five women and one man) exhibited AQP4-Ab. There was no significant difference in mean age between study groups (positive group: 38.7 ± 11.5 years, negative group: 42.3 ± 14.7 years, $P=0.548$). Bilateral simultaneous involvement was more common in seropositive patients than in seronegative patients (occurred in two out of six seropositive patients and in one out of 36 seronegative patients, $P = 0.007$). With regards to poor visual outcome (worse than 1.0 LogMAR), seropositive patients exhibited more severe visual loss than seronegative patients. None of the seropositive patients exhibited myelitis symptoms during the follow-up period (mean follow-up period: 8–32 months).

Conclusions:

The prevalence of AQP4 antibody was often detected in the newly onset optic neuritis patients. In the patients with bilateral involvement or poor initial visual acuity, the AQP4 antibody test should be considered.

References: None.

Keywords: Optic Neuropathy, Demyelinating Disease

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 151**Change of Th17 lymphocytes and Treg/Th17 in Typical and Atypical Optic Neuritis**

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Introduction:

Typical and atypical optic neuritis (ON) are two types of autoimmune inflammatory diseases of the optic nerve that causes acute vision loss, and which are difficult to distinguish in their early stages. The disturbance in the balance of Th17 and Treg lymphocytes is thought to play an essential role in these autoimmune inflammatory diseases. To detect the clinical relevance of Th17 and Treg in peripheral blood and the ratio of Treg/Th17 in patients with typical and atypical ON. To explore potential methods with which to distinguish typical and atypical ON based on their distinct pathogenesis.

Methods:

We studied consecutive patients aged 14-70 years with typical ON (n=30) or atypical ON (n=33) within 4 weeks of their acute attack. Routine clinical tests and ophthalmological examination were performed in all patients. Blood samples were collected from untreated patients and from gender- and age-matched healthy controls (n=30). The proportion of peripheral blood Th17 cells and Treg cells was determined by flow cytometry.

Results:

Patients with atypical ON had a higher proportion of Th17 cells than patients with typical ON (3.61 ± 1.56 vs 2.55 ± 1.74 , $p<0.01$) or controls (1.45 ± 0.86 , $p<0.01$). The proportion of Th17 cells in patients with typical ON was also markedly higher than in controls ($p<0.01$). The mean percentage of Treg cells in atypical ON (6.31 ± 2.11) and typical ON (6.80 ± 2.00) were significantly lower when compared to controls (8.29 ± 2.32 , both $p<0.01$). No significant difference in Treg frequency was observed between typical ON and atypical ON ($p>0.05$).

Conclusions:

The frequency of Th17 cells is higher in atypical ON than typical ON, and patients with atypical ON have a greater imbalance of proinflammatory and regulatory cells than patients with typical ON when compared with controls. These changes are indicative of distinct pathological mechanisms and may provide useful information to distinguish typical and atypical ON.

References: None.

Keywords: Demyelinating Disease, Optic Neuropathy, Th17 Cell, Treg Cell, Treg/Th17 Ratio

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 152**Vitamin A deficiency presenting as unexplained visual loss to neuro-ophthalmology clinic**

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Introduction:

Vitamin A is fat soluble nutrient, essential for conjunctival and corneal epithelial cells, retinal photo-transduction, and RPE cell viability. Vitamin A deficiency can cause night blindness, retinopathy and xerophthalmia. More common in developing countries due to malnutrition than in developed countries, where it is usually secondary to malabsorption. We present here a case series of vitamin A deficiency patients, presenting as unexplained visual loss to our neuro-ophthalmology clinic. We characterize the precipitating factors, demographics and workup leading to diagnosis and treatment.

Methods:

10 adult patients were seen over a 15 year period in a single neuro-ophthalmology practice presenting with visual loss. All patients had low vitamin A levels and vision improved after replacement treatment.

Results:

Of the 10 patients, 3 were male 3, 7 female. The average age was 58 years (range 42-72). The median number of encounters before neuro-ophthalmic exam was 2 (range 0-6). The average duration of symptoms was 6 months (range 1-12). 3 patient had an MRI, and 1 patient had a CT prior to the referral. Most common referral diagnosis was optic neuropathy. No patients had any other signs of vitamin A deficiency. All patients had recovery after treatment. The precipitating factors were: abdominal/bariatric surgeries (5 patients), pancreatitis (2 patients), cholecystectomies (2 patients), and in one patient the cause was believed to be caused by medication (Deferasirox-oral iron chelator).

Conclusions:

Vitamin A deficiency is an underdiagnosed cause of vision loss that can be seen in patients with no clear cause for vitamin deficiency and with no other signs of hypovitaminosis. Patients may not complain of night vision difficulties, which can delay the diagnosis. With normal exam, a long, time consuming and costly work up to the patient and the health system. Instead, a simple blood test and a trial of supplement vitamin A can improve or even resolve the symptoms quickly.

References: None.

Keywords: Vitamin A, Deficiency, Vision loss

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 153**Hemiatrophy of Macular Ganglion Cells in Patients with Retrochiasmal Lesions**

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Introduction:

Lesions in the optic tract or along the thalamocortical pathway cause a homonymous hemianopia. Irreversible retrochiasmal damage may lead to optic atrophy. As the macular area is overrepresented in the afferent visual system, a retrochiasmal lesion may be best detected at the level of the macula. We sought to determine the anatomic changes of the macular ganglion cell layer in patients with retrochiasmal lesions.

Methods:

In a consecutive retrospective case series we included 85 eyes from patients with homonymous hemianopia and macular optical coherence tomography (OCT). We used automatic layer segmentation to determine thickness of individual retinal layers.

Results:

We found that a significant portion of patients with lesions at the level of the optic tract, the optic radiation or the visual cortex displayed ganglion cell loss in the macula. The ganglion cell loss respected the vertical meridian leading to a characteristic macular ganglion cell 'hemiatrophy'. In contrast to the disc fovea angle, the vertical meridian separating the temporal and nasal hemiretina was not tilted.

Conclusions:

Macular ganglion cell layer analysis in patients with retrochiasmal lesions allows a better topical diagnosis with a higher sensitivity than can be achieved with assessment of the optic disc. The vertical respect that is important in the visual field analysis has an anatomic pendant in the macula.

References: None.

Keywords: Optical Coherence Tomography, Ganglion Cell, Optic Tract, Atrophy, Retrograde Degeneration

Financial Disclosures: The authors had no disclosures.

Grant Support: This study was supported by the Swiss National Science Foundation

Poster 154

Disorders of Higher Visual Processing in Patients after Stroke

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Introduction:

Damage to specific areas outside the primary visual cortex can cause selective deficits of higher visual processing. Patients with stroke may have undetected abnormalities of higher visual processing because these are not routinely assessed. If present, these could impact significantly on recovery and residual function. This pilot study aimed to determine the extent of these abnormalities in patients who had recovered from stroke.

Methods:

26 patients (65.5 ± 13.9 years; 12 female) who had recovered from stroke were compared with 29 age-matched controls (67.6 ± 13.1 years; 17 female). Ophthalmological causes of visual loss were excluded. Higher visual function was assessed using Ishihara charts and Farnsworth-Munsell D15 (FMD15), kinematography, random dot testing (depth), stereofly, line bisection, and the Cambridge facial memory test (CFMT).

Results:

Looking at the entire group, stepwise regression showed that random dot and CFMT were the most significant predictors of stroke ($F_{1,77} = 5.08$, $p = 0.027$). At a false positive rate of 10%, random dot classified 38.4% (± 17.6% SE) of patients and CFMT classified 28.0% (± 9.1% SE). Principal component analysis revealed two independent factors which accounted for 29.9% and 21.3% of the variance, respectively. Variables which contributed significantly to the first factor were random dot, FMD15, CFMT and kinematography. Line bisection, Ishihara and CFMT contributed to the second factor. The receiver operator characteristic yielded an area under the curve of 0.75 ± 0.071 (mean ± SE).

Conclusions:

Many patients with stroke had undetected abnormalities of higher visual processing. As a group, this was most obvious in terms of random dot testing for depth perception. These findings have potential relevance to the process of rehabilitation and to residual post-stroke function. Further investigation in the form of a larger trial is warranted.

References: None.

Keywords: Higher Visual Cortical Functions, Stroke

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 155**Measuring Axonal Loss with Serial OCT Testing in Multiple Sclerosis Patients: Methods and Yield**

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Introduction:

In patients with multiple sclerosis (MS), OCT demonstrates evidence of acute axonal loss after attacks of optic neuritis and chronic progressive axonal loss even without optic neuritis. We evaluated three methods of measuring retinal ganglion cell (RGC) axonal loss in the eyes of individual MS patients, using serial tests of optical coherence tomography (OCT).

Methods:

We performed a retrospective study of serial spectral-domain OCT tests in a cohort of 50 MS patients (92 eyes) from a single clinical center and laboratory. Tests were conducted at least six months apart to assess change over time (mean interval, 2.3 years). Standard assessments included peripapillary retinal nerve fiber layer (RNFL) thickness and macular segment volumes. We also quantified optic nerve head (ONH) volumes, as previously described.

Results:

Of 92 eyes, 23 eyes (25%, 18 patients) met at least one criterion for significant reduction between tests: mean or temporal RNFL thickness (≥ 7 microns), ganglion cell + inner plexiform layer (GC+IP) volume (≥ 0.09 mm³) or ONH volume (≥ 0.09 mm³). Of these 23 eyes, seven exceeded thresholds for significant loss for all three measures. Seven other eyes did so for two measures. RNFL thickness, GC+IP volume and ONH volume were equally sensitive in detecting change. Only 1/23 eyes had a clear interval history of optic neuritis, while 5/23 had vaguer histories of visual blurring or eye pain between tests.

Conclusions:

Serial OCT testing has a reasonably high yield for identifying evidence of RGC axonal loss over time in individual MS patients, even when there is no interval history suggesting optic neuritis. Studying ocular anatomy in more than one area may increase detection and certainty of change. North American Neuro-Ophthalmology Society | 315

Keywords: Diagnostic Tests, Demyelinating Disease, Optic Neuropathy Retina, Neuro-Ophth & Systemic Disease

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 156

Perceptual Learning Treatment of Amblyopia in Adults

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Introduction:

Recently a perceptual learning-based video game (ULTIMEYES) has been developed for treatment of various low vision conditions. The game task involves the detection of low contrast sine gratings of varying spatial frequency, where contrast is controlled adaptively to maintain performance at threshold levels. This program has previously not been utilized for amblyopia.

Methods:

This pilot study enrolled 9 adults (mean age 32 years; range 16-49) diagnosed with strabismic or anisometropic amblyopia, who had previously undergone patching treatment, and whose visual acuity in the amblyopic eye ranged from 20/40 to 20/200. Subjects underwent pre- and post-treatment testing using high and low contrast visual acuity optotypes and sensory measures including Worth-4-dot and stereoacuity at distance and near. Prescribed treatment consisted of four sessions of 20 minutes each per week for 8 weeks. The therapy was performed binocularly with the use of a polarizing filter over the dominant eye to minimize contribution ability of that eye.

Results:

Following treatment there was a statistically significant improvement in amblyopic eye ETDRS visual acuity averaging 5 letters ($p < 0.01$). Suppression on the Worth-4-dot tests at distance was reduced from 6/9 subjects pre-treatment to 3/9 subjects post-treatment.

Conclusions:

ULTIMEYES is a perceptual-learning based therapy. Pilot data shows that 8 weeks of therapy in adults leads to significantly improved visual acuity and reduced suppression. Further study is needed in order to determine long-term effects, and compare to placebo and established amblyopia treatments. ULTIMEYES shows promise as a perceptual-learning technique to improve visual acuity and binocularity in amblyopic adults.

References:

Deveau, Lovcik, and Seitz (2014). Broad-based visual benefits from training with an integrated perceptual-learning video game", *Vision Research*, 99:134-40.

Deveau and Seitz (2014). Applying Perceptual Learning to achieve practical changes in vision. *Frontiers in Psychology*, doi: 10.3389/fpsyg.2014.01166

Keywords: Adult Amblyopia, Perceptual Learning

Financial Disclosures: The authors had no disclosures.

Grant Support: NIH/NEI, Research to Prevent Blindness.

Poster 157

Imaging Features of Idiopathic Intracranial Hypertension (IIH) in Children

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Introduction:

MRI signs of intracranial hypertension are very common in adults with IIH in whom they are well characterized.¹ Our goal was to determine whether these findings are also present in children.

Methods:

50 children (<18yo) with definite IIH and an MRI performed at our institution (2007-2015) were included. Two expert neuroradiologists assessed MRIs for optic nerve head protrusion, scleral flattening, increased perioptic CSF, optic nerve tortuosity, sellar changes, enlargement of Meckel's cave, cerebellar tonsillar position, and transverse venous sinus stenosis. Findings were compared with those of 46 previously collected adult IIH patients meeting the same inclusion criteria.²

Results:

50 IIH children included 42 girls (26 black, 20 white, 4 other; mean age, 12.8yo [range 2-17]). Patients were analyzed in groups of prepubescent children (<11 years; n=10) and adolescents (11-17 years, n=40), and were compared to adults with IIH (>17yo, n=46). Compared to adolescents and adults, prepubescent children had a lower mean BMI (20.8 vs. 35.3 and 37.9, p<0.001), frequency of scleral flattening (50% vs. 89% and 85%, p=0.02), sellar changes (56% vs. 78% and 93%, p=0.007), perioptic increased CSF (60% vs. 84% and 89%, p=0.08), optic nerve tortuosity (20% vs. 46 % and 59%, p=0.07), and transverse sinus stenosis (67% vs. 93% and 96%, p=0.04). Adolescents had higher mean CSF-OP than prepubescent children and adults (47.8 vs. 37.0 and 36.7 cm, p<0.001). Median duration of symptoms prior to MRI was 5 weeks (range 4-12 weeks) in prepubescent children, 2 weeks (range 1-16 weeks) in adolescents, and 5 weeks (range 0-196 weeks) in adults (p=0.001).

Conclusions:

MRI findings of adolescents with IIH were similar to those described in adults. Prepubescent children (<11yo) with IIH have similar MRI findings as previously described in adults, although at a lower prevalence. This may suggest a different response of the brain and associated tissues in prepubescent children to elevated ICP.

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Keywords: Pediatric Neuro-Ophthalmology, Idiopathic Intracranial Hypertension, Neuroimaging, High Intracranial Pressure/Headache

Financial Disclosures: The authors had no disclosures.

Grant Support: Research to Prevent Blindness NIH/NEI:P30-EY006360.

Poster 158

Women with Idiopathic Intracranial Hypertension (IIH) Have a Distinct Andro-Metabolic Signature Compared to Polycystic Ovary Syndrome (PCOS) and Simple Obesity

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Introduction:

Idiopathic intracranial hypertension (IIH) is characterised by elevated intracranial pressure and occurs predominantly in obese premenopausal women. Polycystic ovary syndrome (PCOS) can coexist in IIH and patients have similar phenotypes. Here we compared the androgenic and metabolic phenotypes in IIH, PCOS and simple obesity.

Methods:

We studied 25 patients with IIH (mean age 34.4±9.2 years; mean BMI 37.8±5.2kg/m²), in comparison to 31 women with PCOS and 15 with simple obesity; all three groups were matched for age and BMI. Women with IIH were studied before and after a weight loss intervention (mean BMI change -5.8±3.0kg/m²). In all participants we performed comprehensive metabolic phenotyping and steroid profiling. Serum androgens were measured by liquid chromatography-tandem mass spectrometry; 24-hour urinary steroid excretion was analysed by gas chromatography/mass spectrometry. Urinary steroid profiles were correlated with clinical parameters of IIH severity.

Results:

Serum testosterone (T) in IIH was comparable to PCOS and significantly higher than controls (p=0.01). Serum androstenedione (A) was significantly increased in PCOS (p=0.008) but IIH did not differ from controls. Insulin resistance as assessed by HOMA-IR did not differ between IIH and controls. Total glucocorticoid excretion was significantly higher in IIH compared to controls (p=0.01) and decreased after weight loss (p=0.02). Similarly, the urinary ratio of 5α-THF/THF, a marker of systemic 5α-reductase activity, was significantly increased in IIH compared to controls (p=0.04). The An/Et ratio correlated significantly with baseline markers of papilloedema (Optical Coherence Tomography) in IIH (R=0.47, p=0.02).

Conclusions:

These results indicate a distinct andro-metabolic signature in IIH, characterised by increased testosterone but not androstenedione. In IIH, Testosterone and 5α-reductase activity falls following therapeutic weight loss alongside a significant reduction in disease activity (intracranial pressure and papilloedema). These findings maybe driven by increased activity of the enzyme aldo-ketoreductase type 1C3 (AKR1C3) which activates androstenedione to testosterone in adipocytes.

References: None.

Keywords: Idiopathic Intracranial Hypertension, Pathogenesis, Androgen Profile, Poly Cystic Ovary Syndrome

Financial Disclosures: The authors had no disclosures.

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Poster 159

Headache Characteristics in Pediatric Patients with Definite Pseudotumor Cerebri Syndrome and Intracranial Hypertension

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Introduction:

Revised diagnostic criteria for Pseudotumor Cerebri Syndrome (PTCS) were published in 2013[1]. This manuscript did not suggest a specific diagnosis for patients in whom there was clinical suspicion for PTCS where data did not support the diagnosis. Clinically it is unclear whether patients without papilledema who have headache features suggestive of increased intracranial pressure benefit from lumbar puncture and/or medications used to lower cerebrospinal fluid(CSF) pressure.

Methods:

In this study we performed a retrospective review to characterize the headache features of 59 subjects with Definite PTCS (47 primary, 12 secondary) and 30 with Intracranial Hypertension (IH) who did not meet criteria for PTCS.

Results:

Headaches were present in all subjects with Definite PTCS and 29/30 with IH. There was insufficient information in the chart to classify the headaches of 28 subjects with Definite PTCS & 6 with IH. This was related to primary symptom – there was less information about headache characteristics for subjects who presented with significant vision loss or papilledema. Headaches were consistent with migraine or probable migraine in 29/31 subjects with Definite PTCS and 21/23 with IH. The headaches were consistent with tension-type (TTH) in 2/31 subjects with Definite PTCS and 1/23 with IH. 1 subject with IH had headaches which did not fit criteria for either migraine or TTH.

Conclusions:

Consistent with prior studies, headache characteristics are similar across these groups. However, data analysis is ongoing. The final sample size will be 161. This will include subjects with Normal OP, and will characterize other features of headache, effect of LP, and response to treatments which could lower CSF pressure in all groups. The long-term goal of this research is to inform a prospective study which could clarify whether patients without papilledema who have headaches with features suggestive of increased ICP derive benefit from LP and/or treatments designed to lower CSF pressure.

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Friedman DI, Liu GT, Digre KB. "Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children." *Neurology*, 81(13): 1159-65, 2013

Keywords: Pseudotumor Cerebri, High Intracranial Pressure/Headache, Pediatric Neuro-Ophthalmology

Financial Disclosures: The authors had no disclosures.

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Poster 160

Radiographic Features of Pediatric Idiopathic Intracranial Hypertension

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Introduction:

Adult patients with idiopathic intracranial hypertension (IIH) may harbor characteristic radiographic signs on magnetic resonance imaging (MRI). These signs have not been extensively evaluated in children. The purpose of this study is to examine the radiographic findings of pediatric IIH to characterize salient features and gain a deeper understanding of disease pathophysiology.

Methods:

Retrospective, case-control study of pediatric patients with and without IIH from the ophthalmology department at single tertiary care center was performed. Clinical data including demographics, lumbar puncture results, and ophthalmic findings were obtained. Patient MRI/MRV was evaluated for presence of an enlarged perioptic subarachnoid space, posterior globe flattening, protrusion of the optic nerve head, empty sella turcica, prominent arachnoid granulations, skull base crowding, Chiari malformation, ventriculomegaly, and transverse sinus stenosis.

Results:

Neuroimaging and clinical findings of 49 patients with IIH and 30 control patients were evaluated. Compared to controls, IIH patients had significantly larger perioptic subarachnoid space ($p < 0.001$), and higher incidences of posterior globe flattening ($p < 0.001$), protrusion of the optic nerve head ($p < 0.001$), and empty sella turcica ($p < 0.001$). The presence of prominent arachnoid granulations, skull base crowding, Chiari malformation, ventriculomegaly, and transverse sinus stenosis did not reach significance. Multivariate models were developed for predicting IIH in children.

Conclusions:

Several highly sensitive key radiographic findings in pediatric IIH were identified and models were developed for predicting pediatric IIH. In the evaluation of pediatric patients, there are characteristic radiographic findings on MRI that should raise concern for IIH.

References: None.

Keywords: High Intracranial Pressure/Headache, Neuroimaging, Pediatric Neuro-Ophthalmology, Pseudotumor Cerebri

Financial Disclosures: The authors had no disclosures.

Grant Support: Fight For Sight-NANOS Research Award.

Poster 161**Idiopathic Intracranial Hypertension: A Retrospective Evaluation of the Management and Outcomes at One Large Tertiary Care Center**

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Introduction:

Idiopathic intracranial hypertension (IIH) is a common and potentially blinding disorder that affects most frequently young overweight women. The NORDIC IIH treatment trial defined the value of acetazolamide and weight loss in the management of IIH. The goal of this study was to reflect upon our outcomes based upon our traditional strategy for managing patients and also to explore outcomes across a greater range of visual disability than was studied in the IIH treatment trial study.

Methods:

Retrospective acquisition of data from the patients 18 years and older who were diagnosed with IIH at our center between 2010 and 2015. A total of 111 patients were identified. Data was entered into our customized database and analyzed with respect to patient demographics, history, clinical and radiological signs, results of lumbar puncture, and whether surgery was performed.

Results:

Our preliminary data on 35 patients showed headache as the major symptom in 83% and papilledema with concurrent vision changes in 85% of patients. The average mean deviation score among all patients in whom initial visual field testing could be performed (55 eyes in 35 patients) was -2.83 dB in the right and -3.71 dB in the left eye. Papilledema and headaches improved in more than 2/3 of patients after initiation of Diamox treatment (typical dose 1.0-1.5 g/d), none of these patients required surgery. The remainder of the analysis is in process.

Conclusions:

Surgery was only uncommonly required for management of our patients. Our standard treatment approach, which was usually acetazolamide (doses typically 1 – 1.5 grams), was adequate to control symptoms and visual function in the great majority of patients.

References: None.

Keywords: Pseudotumor Cerebri, High Intracranial Pressure, Headache

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 162**The Translaminar Gradient Influences Papilledema Severity**

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Introduction:

Recent studies have suggested that intracranial pressure (ICP) may play a role in the development of glaucoma. It has been specifically hypothesized that the translaminar gradient (TLG) between intraocular pressure (IOP) and the cerebrospinal fluid pressure in the optic nerve sheath may influence optic nerve conformation and axonal damage. This study aims to examine whether the TLG influences the degree of papilledema observed in patients with idiopathic intracranial hypertension (IIH).

Methods:

In this university-based retrospective study, the electronic medical record was used to identify patients being assessed for IIH from 2011-2015 who underwent lumbar puncture (LP), optical coherence tomography (OCT) scan of the peripapillary retinal nerve fiber layer (RNFL), and measurement of IOP, all within a three month period. Exclusion criteria included optic nerve gliosis, pallor, or anomalous shape, or interval surgery or change in medication that could affect ICP or IOP. Translaminar gradient (the difference between ICP and IOP) and ICP were plotted against average RNFL thickness, and the Pearson correlation coefficient was calculated.

Results:

34 patients (mean age 37 years; 15% were male) met the criteria. Among patients with LP opening pressure ≥ 25 cm H₂O (n=16), there was a significant correlation between TLG and OCT RNFL average thickness. TLG was more highly correlated with OCT RNFL thickness than absolute LP opening pressure (Pearson correlation $R^2 = 0.45$, $p = 0.004$ for TLG, compared to $R^2 = 0.38$, $p = 0.011$ for LP opening pressure).

Conclusions:

Our data suggests that the optic nerve translaminar pressure gradient correlates with the degree of papilledema more closely than ICP alone. This observation of the influence of IOP on papilledema carries significant pathophysiologic and clinical implications.

References: None.

Keywords: Pseudotumor Cerebri, Papilledema

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 163**Persistence of Transverse Sinus Stenosis After Lumbar Puncture in Idiopathic Intracranial Hypertension (IIH)**

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Introduction:

Diffuse or focal distal stenosis of both transverse venous sinuses is a classic sign of raised intracranial pressure (ICP), particularly IIH.^{1,2} Although some stenoses result from intrinsic sinus anomalies and are a primary cause of intracranial hypertension, others result from external compression of the distal transverse sinus and are reversible with treatment of increased ICP or after lumbar puncture (LP).^{3,4,5} Our goal was to evaluate transverse sinus stenosis (TSS) immediately after the diagnostic LP in patients with IIH.

Methods:

Twelve treatment-naïve IIH patients (11 women, 1 man, mean age 28y [range 18-42], mean BMI 36Kg/m² [range 24-53]) seen at our institution after 12/2009 who underwent a high-quality standardized contrast-enhanced brain magnetic resonance venogram (MRV) less than 30 minutes after their diagnostic LP were included. The degree of TSS was calculated by dividing the width of the stenosis by the apparent normal width of the sinus, as previously described.²

Results:

All patients had elevated CSF-opening pressure (median 35cmH₂O [IQR 32-41]). Median CSF-closing pressure was 17cmH₂O (IQR 16-19). All patients had residual bilateral TSS on the post-LP MRV (median 61% [IQR 50%-74%]). Three of these patients had a pre- and a post-LP MRV, which demonstrated a reduction in the degree of stenosis in all 3 patients after the LP, but only by less than 25%.

Conclusions:

As previously suggested², all IIH patients have some degree of bilateral TSS, which may contribute to elevated ICP, but is not associated with clinical outcome. Substantial bilateral TSS persists immediately after acute reduction of the ICP by LP. Possible explanations include: 1) resolution of TSS is delayed after a decrease of ICP; 2) some degree of TSS persists indefinitely in some IIH patients as previously suggested.⁶ Our findings reinforce that clinical features, not the presence or degree of TSS, should be used to determine management in IIH.²

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Keywords: Pseudotumor Cerebri, Neuroimaging, Transverse Sinus Stenosis, High Intracranial Pressure/Headache, Venous Stenting

Financial Disclosures: The authors had no disclosures.

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Poster 164

Comparative Detection of Optic Disc Abnormalities with Digital Images Obtained with an iPhone Camera attached to a PanOptic Ophthalmoscope vs. Standard Fundus Photography

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Introduction:

Neurogenic visual loss often causes optic disc abnormalities that need to be evaluated urgently by non neuro-ophthalmologists. The ability to transmit digital photographs to a specialist makes it possible for a neuro-ophthalmologist to remotely evaluate optic nerves in real time. The purpose of this study was to evaluate the relative diagnostic accuracy of detecting optic disc abnormalities in photographs taken by an iPhone4S camera attached to the PanOptic Ophthalmoscope (ICPO) vs. those obtained with standard fundus camera photography.

Methods:

New patients ≥ 18 years of age referred to the MEEI Neuro-ophthalmology service for acute vision loss or headache were enrolled, which was approved by the local IRB. Fundus photographs were taken of all patients before and after pupil dilation with ICPO and post-dilation with a standard fundus camera. The photographs were evaluated by two neuro-ophthalmologists who were masked to the clinical impression and the reason for referral.

Results:

Twenty-nine patients (58 eyes) were enrolled. Roughly 20% of the cases were clinically diagnosed with either optic disc edema (20.7%) or optic disc pallor (19.0%). When reviewing photographs, the mean false positive rate was 9.3% (undilated ICPO), 22.9% (dilated ICPO), and 10% for dilated standard fundus photography. The mean false negative rate was 41.3%, 33.7% and 31.5% for the three groups respectively. The mean accuracy in detecting optic disc edema in the three groups was 45.8% (95%CI, 25.6 – 67.2), 50.0% (95%CI, 29.1 – 70.9), 58.3(95%CI, 36.6 – 77.9) respectively. The mean accuracy in detecting optic disc pallor was 27.3% (95%CI, 10.7 – 50.2), 45.5% (95%CI, 24.4 – 67.8), 63.6% (95%CI, 40.7 – 82.8) respectively.

Conclusions:

Blinded reviewers of ICPO photography allows for limited accuracy in detecting optic disc edema that was not statistically different from the standard fundus camera photography. There was a trend to higher accuracy with Frisen grade 2-4 versus Frisen grade 1 edema.

References: None.

Keywords: Optic Disc Abnormalities, Diagnostic Tests, Digital Images

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 165

Quantitative Analyses of Retinal Microvascular Network in Alzheimer's Disease

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Introduction:

Alzheimer's disease (AD) is a progressive neurodegenerative disorder with cognitive symptoms appearing long after neural degeneration has begun. Accumulating evidence implicates vascular dysfunction in AD pathogenesis, but assessing the cerebral microvasculature is difficult. The microvasculature in retina and brain are similar anatomically and physiologically. In this project, optic coherence tomography angiography (OCTA) was used to quantitatively analyze the retinal microvascular network density in patients with AD or mild cognitive impairment (MCI).

Methods:

The macula (3×3 mm² centered on the fovea) was imaged by Cirrus OCTA prototype (Carl Zeiss Meditec, Dublin, CA). Five AD, 5 MCI and 10 age-matched normal controls (NC) were recruited. Image processing was performed to separate microvessels (width <25 μ m) from the *en face* angiograms. Fractal analyses using box counting (D_{box}) representing vessel density were performed. The relationships between the microvascular density and macular ganglion cell layer (GCL) thickness were also analyzed.

Results:

The retinal microvessel networks of all subjects (mean age, 50 – 85 yrs; men: women = 10:10) were clearly visualized. D_{box} (AD median=1.763, range=1.665 to 1.776; MCI median=1.749, range=1.737 to 1.767) was highly correlated with GCL thickness in AD ($r = 0.90$, $P < 0.05$) and MCI patients ($r = 0.71$, $P < 0.05$). The D_{box} of parafoveal microvessels of the 2.5 mm annular zone was 1.746 (SD: 0.046) in AD and did not differ significantly from MCI (1.751, SD: 0.011) ($P = 0.40$). The D_{box} in AD and MCI subjects combined did not differ from controls (1.752, SD: 0.025) ($P > 0.05$).

Conclusions:

This preliminary study demonstrated the feasibility of quantitative analysis of retinal microvascular network in patients with AD or MCI. While retinal microvascular network density did not differ across groups in this study, further research in a larger sample is needed to investigate if retinal microvascular dysfunction exists in AD.

References: None.

Keywords: Alzheimer's Disease, Mild Cognitive Impairment, Retinal Microvascular Network Density, Fractal Analysis (D_{box}), Optic Coherence Tomography Angiography

Financial Disclosures: The authors had no disclosures.

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Poster 166

Neuro-ophthalmology Training in Ophthalmology Residency Programs in the United States

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Introduction:

Significance: The status of neuro-ophthalmology education in ophthalmology residency training programs is unknown in the United States. **Background:** There is literature detailing resident experiences for surgical procedures, however there are no articles that detail the teaching of a non-procedural specialty within ophthalmology. There are no ACGME requirements for neuro-ophthalmology (NO) training within ophthalmology residency programs. Each program independently determines the amount of NO training and the residency level of training within ophthalmology. Recent studies of ophthalmology residents on satisfaction with their residency NO training rate ranged from 55% to 91%.

Methods:

A survey was sent to residency directors and/or neuro-ophthalmologists at all ophthalmology residency programs in the United States using the Ophthalmology Residency Matching Program to determine the amount of NO training that residents receive.

Results:

One hundred surveys were obtained from a total of 113 ophthalmology programs (88% response rate). Ninety two percent of ophthalmology residency programs had a NO rotation although every program except one had neuro-ophthalmology training during residency. Neuro-ophthalmology training consisted of a solid block of time or scattered days, with the total number of days ranging from 4 to 80. Most rotations occurred within post-graduate year 2 or 3 with an average of just over 1 resident/clinic (1.18).

Conclusions:

This is the first evaluation of the amount of NO training that occurs within ophthalmology residencies participating in the matching program in the US. Future studies could evaluate if there is any correlation between resident satisfaction in NO training and the amount of training. Determining extent of NO training within neurology residency programs is another area of interest.

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Keywords: Neuro-Ophthalmology, Residency, Education

Financial Disclosures: The authors had no disclosures.

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Poster 167

Relationship Between Cyclotorsion and Visual Vertical: Dependence on Sighting Dominance

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Introduction:

Eye torsion appears as a major determinant of the subjective visual vertical (SVV). Apparent paradox is that monocular vision is spontaneously used in some visuospatial skills while monocular SVV was shown to be unreliable. Based on the large interindividual variability of eye cyclotorsion, this study compares the dependence of monocular SVV on torsion in sighting and non sighting eyes.

Methods:

84 healthy volunteers, 25 M, 59 F, aged 20 to 50, Normal Acuity and Binocular vision. RE dominant :57, LE dominant : 27. Ten measures of SVV each eye and each participant. Ten eye fundus photographs with Scanning Laser Ophthalmoscope (SLO). ANOVAs were carried out separately on the measures of eye torsion and SVV, including sighting eye, preferred hand and sex as between-subject factors, and eye (left, right) as repeated factor. A Bravais-Pearson coefficient was calculated for each sighting dominance group and each eye.

Results:

Averaged on both eyes, excyclotorsion also appeared slightly greater ($F_{1,76} = 4.69$; $p = 0.033$) in the right sighting group (6.12°) than in the left sighting group (5.44°), but the difference between LE and RE did not change according to sighting group ($F_{1,76} = 2.06$; $p = 0.155$). Value of SVV was greater for RE than for LE in 70.2 % of the participants. Mean difference between monocular SVV of LE (-0.19°) and RE ($+0.34^\circ$), was significant ($F_{1,76} = 5.95$; $p = 0.017$). It was not significantly affected by sighting side ($F_{1,76} = 0.27$; $p = 0.605$). When the non sighting eye was considered, SVV correlated significantly with OT whether the non sighting was the RE ($r_{25} = 0.40$; $p = 0.039$) or the LE ($r_{25} = 0.27$; $p = 0.043$). It was not the case for the sighting eye, right ($r_{25} = 0.13$; $p = 0.345$) or left ($r_{25} = 0.08$; $p = 0.692$).

Conclusions:

The torsion of non sighting eye correlated with monocular SVV, while that of sighting eye did not

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Keywords: Cyclotorsion, Visual Vertical, Sighting Dominance

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 168**Medication Reconciliation in a Neuro-ophthalmology Clinic**

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Introduction:

The purpose of this study was to quantify and categorize the type of medication errors we may encounter in the patient record, and also to evaluate the relevance of these errors with respect to neuro-ophthalmologic care. We aimed to evaluate the accuracy of the medication list prior to the patient being examined in our clinic. Time-stamping on the electronic medical record allowed us to distinguish medication changes from medication errors (changes which should have been documented at the previous reconciliation).

Methods:

We prospectively evaluated consecutive patients meeting inclusion criteria of having a time-stamped confirmed medication reconciliation prior to evaluation, having full recall of their medications, and having pharmacy staff available. Medications were reviewed and errors recorded, categorized, and rated by expected relevance to the forthcoming neuro-ophthalmologic evaluation (1=possibly, 2= probably, 3=definitely relevant).

Results:

Seventy-seven patients were evaluated. Sixty-two had at least one medication error. There were 296 medication errors total. The type of error included: Not on list: 112 (38%); No longer taking: n=93 (31%); Dosage error: n=76 (26%); and Duplicate: n=15 (5%). The errors were classified as having: Possible: n=248 (84%), Probable n=42 (14%), and Definite (n=6, 2%) relevance to forthcoming neuro-ophthalmologic care.

Conclusions:

Despite documentation that a medication list is updated and accurate, there remains a significant chance that an error is present. Six percent (4/77) of our patient charts contained a highly relevant medication error.

References: None.

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 169

Visual Quality of Life in Migraine

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Introduction:

Previous research has shown that patient-reported quality of life (QOL) data can be useful in determining the severity of disease burden in migraine, but previous studies have not assessed how migraine affects visual QOL. In this study, we are assessing vision-specific QOL in migraine patients.

Methods:

We assessed individuals with chronic and episodic migraine using the National Eye Institute Visual Function Questionnaire-25 (NEI-VFQ-25), the 10-item NEI-VFQ-25 Neuro-Ophthalmic (NO) Supplement, and the Headache Impact Test (HIT-6). We compared these results to those from previously published disease-free controls, and to results from other neuro-ophthalmic disease QOL studies. We intend to enroll at least 120 participants.

Results:

Our preliminary results reveal that among 21 participants with chronic migraine (17 women and 4 men; mean age \pm SD of 50.8 \pm 13.2 years), vision-specific QOL scores were significantly decreased compared to previously published disease-free controls ($p=0.0001$). Among 19 participants with episodic migraine, with or without aura (13 women and 6 men; mean age \pm SD of 51.7 \pm 18 years), vision-specific QOL scores were also decreased compared to disease-free controls. Results for the NO supplement were statistically significant ($p=0.0013$), but these results may not be statistically significant for the NEI-VFQ-25 ($p=0.0867$). Chronic migraineurs had decreased visual QOL scores compared to those with episodic migraine. Scores for the NEI-VFQ-25 and the NO supplement for chronic migraine were similar to previously published results for patients with other neuro-ophthalmic disorders such as MS, myasthenia gravis, and ischemic optic neuropathy. Subjects with episodic migraine had QOL scores that were better than patients with other neuro-ophthalmic disorders.

Conclusions:

Migraine affects visual QOL. Patients with chronic migraine may have visual QOL similar to patients with other neuro-ophthalmic disorders. These findings suggest closer attention to, and study of, vision in migraine is indicated, and could have important implications in the treatment of migraine.

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Keywords: Headache, Quality of Life, Photophobia, Aura

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Poster 170

Cognitive Enhancement For Visual Field Testing

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Introduction:

Abnormal Humphrey Visual Field (HVF) testing in healthy adults can be the result of decreased mental alertness and vigilance, even with normal reliability indices. Methylphenidate is a stimulant intended for the treatment of Attention deficit and Hyperactivity Disorder (ADHD) which is also used for cognitive enhancement. Our aim was to evaluate the effect of methylphenidate on HVF testing in normal adults without ADHD.

Methods:

This was a prospective randomized controlled study. Sixteen patients with an unexplained visual field defect were randomly assigned into an interventional and control groups. Both groups repeated the same HVF test (8 subjects in each group). The interventional group was given 10mg methylphenidate one hour before the test. Patients completed the Adult ADHD Self-Report Scale (ASRS-v1.1) and Montreal Cognitive Assessment (MoCA) questionnaires. One eye was randomly selected for statistical analysis. The Mean deviation (MD), Pattern Standard Deviation (PSD) and reliability indices were compared. Three experienced ophthalmologists that were masked to the subjects' identity, group assignment and the order of the tests reviewed both tests for a clinically significant difference between the first and second HVFs. Wilcoxon Rank Sums test and Fisher Exact test were used for statistical comparisons.

Results:

Median age was 65.5 for the methylphenidate group (interquartile range [IQR]: 51-77) and 53.5 for controls (IQR: 47-67, $p=0.24$). MD improved in the methylphenidate group (median +3.96 dB, IQR: 2.43-5.39) compared to no change in controls [median -0.15 dB, IQR: (-1.0) – (+2.22), 1-sided Wilcoxon, $p=0.046$]. PSD decreased in the methylphenidate group [median -3.32 dB, IQR: (-4.43) – (-0.82)] compared to no change in controls [median +0.16 dB, IQR: (-0.48)–(+2.13), $p=0.006$]. Clinical assessment of the visual fields yielded improvement in 7 (87.5%) of the methylphenidate group compared to 2 (25%) of the controls ($p=0.04$).

Conclusions:

This preliminary (ongoing) study may suggest that methylphenidate improves HVF testing in non-ADHD adults.

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Adler LA, Spencer T, Faraone SV, et al. Validity of pilot adult ADHD self-report scale (ASRS) to rate adult ADHD symptoms, Ann Clin Psychiatry 18, 145-148, 2006.

Keywords: Diagnostic Tests, Perimetry, Visual Fields

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 171**Assessing the Cost Inefficiency Attributable to Surgical Instrument Trays Used in Adult Strabismus Surgery**

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Introduction:

This study aims to quantify the cost to the healthcare system attributable to inefficient use of instrument trays during adult strabismus surgeries performed at our institution. We plan on predicting the potential savings involved in developing a standardized adult strabismus instrument tray that excludes unnecessary instruments.

Methods:

This is a single-site observational study. Data on frequency of instrument use will be obtained through a survey distributed to the three ophthalmologists who routinely perform strabismus surgery at our institution. The survey includes a list of all instruments present on the three most commonly used instrument trays for this type of surgery at this site. The surgeons are asked to place each instrument into one of the following three categories: (a) always used, (b) occasionally used (once every 4 OR days), or (c) never used. To evaluate the costs associated with tray usage, the following data will be obtained from the central sterile processing (CSP) unit of the hospital: tray decontamination and reassembly time, costs associated with missing or damaged instruments, and the CSP operative cost. Potential cost savings associated with eliminating from the trays those instruments that are deemed never used and developing a standardized adult strabismus tray that excludes these instruments will be calculated.

Results:

Data collection is currently under way.

Conclusions:

Pending collection and analysis of data.

References: None.

Keywords: Strabismus, Cost Analysis

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 172

Comparison of the Surgical Treatment of Superior Oblique Palsy: Inferior Oblique Myectomy Versus Combined Contralateral Inferior Rectus Recession

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Introduction:

To compare inferior oblique (IO) myectomy with combined contralateral inferior rectus (IR) recession for the treatment of superior oblique (SO) palsy.

Methods:

A retrospective review of the medical records of patients with SO palsy who underwent surgical treatment with either IO myectomy or IO myectomy and contralateral IR recession between 2006 and 2015. Group 1 (n=21) underwent IO myectomy alone to correct hypertropia, while group 2 (n=22) underwent IO myectomy plus contralateral IR recession. Analysis of the magnitude of corrected hypertropia in primary position and contralateral gaze, magnitude of postoperative drift and, success rate (defined as residual hypertropia less than 4 PD) was performed.

Results:

Preoperatively, there was no statistical difference in mean vertical deviation in primary position between the two groups (11.0 PD and 13.7 PD, respectively (p=0.17)). However, there was a significant difference in mean vertical deviation in contralateral gaze between group 1 and 2 (13.7 PD and 17.9 PD, respectively, p<.000). Mean vertical deviation measurements at postoperative months 3 were 1.3 PD and -1.5 PD (p=0.02) in primary position and 2.0 PD and 1.2 PD in contralateral gaze (p=0.034). Mean corrected hypertropia in primary position was 9.3 PD and 15.2 PD in group 1 and 2 (p=0.006), and in contralateral gaze was 11.8 PD (group 1) and 16.7 PD (group 2) (p<.000). Success rate of group 1 was 90.5% and that of group 2 was 72.7% (p=0.058).

Conclusions:

Both IO myectomy and IO myectomy combined with contralateral IR recession were effective in the treatment of SO palsy. IO myectomy alone lead to a slight undercorrection whereas a combined procedure lead to a slight overcorrection in primary position. Both procedures often result in a slight undercorrection in contralateral gaze and have a similar surgical success rate.

References: None.

Keywords: Ocular Motility Disorders And Nystagmus, Neuro-Ophthalmic Disorders Of Neurologic And Systemic Diseases, Superior Oblique Palsy, Inferior Oblique Myectomy, Contralateral Inferior Rectus Recession

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Grant Support: None.

Poster 173

MRI Findings in Children with Pseudotumor Cerebri Syndrome (PTCS), Intracranial Hypertension, and Normal Opening Pressure Without Papilledema.

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Introduction:

Revised diagnostic criteria for pseudotumor cerebri syndrome (PTCS) include findings on neuroimaging, which can be used if papilledema is not present. We compared neuroimaging in children suspected to have PTCS, in order to facilitate the identification and treatment.

Methods:

We reviewed 237 pediatric subjects from our PTCS database and from recent lumbar punctures (LPs) performed for clinical suspicion of PTCS. We retrospectively classified 123 subjects with MRIs in the following comparison groups: 61 with Definite PTCS, 13 with Probable PTCS, 24 with Intracranial Hypertension (IH) and 25 with normal opening pressure (OP). A neuroradiologist, who was blinded to the diagnostic group, assessed MRIs for empty sella, posterior globe flattening, distention of perioptic subarachnoid space (>5.5mm) and tortuosity of the optic nerve and transverse sinus stenosis.

Results:

Empty sella was found almost exclusively in Definite PTCS (88%). Posterior globe flattening and transverse sinus stenosis were found most commonly in the Definite PTCS group at 36% and 38%, respectively. Distention of the perioptic subarachnoid space and optic nerve tortuosity were found in 25% of those with Definite and Probable PTCS and in 1% of those with IH. In those with normal OP, no more than 6% had any one of the criteria examined. Three of 4 criteria were present in 72% with Definite PTCS, 16.7% with Probable PTCS, 4.8% with IH, and 6.7% with normal OP. Between-group comparisons with Fisher's exact test for each of the 4 criteria, and for the combined score, were significant with p-value <0.001. The positive predictive value of 3 or more MRI criteria for definite or probable PTCS is 95%. The negative predictive value is 58.6%.

Conclusions:

In children, three of four neuroimaging criteria have a robust positive predictive value for PTCS. Of these, empty sella was found the most often in definite PTCS.

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Friedman DI, Liu GT, Digre KB. "Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children." *Neurology*, 81(13): 1159-65, 2013
Mallery, R.M., et al. MRI findings in pseudotumor cerebri syndrome (PTCS) with and without papilledema. in American Academy of Neurology 2014. Philadelphia, PA.

Keywords: Pseudotumor Cerebri, Neuroimaging, Pediatric Neuro-Ophthalmology

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 174**Hyperintense Optic Nerve Heads on Diffusion Weighted Imaging: Is this Sign Specific to Papilledema?**

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Introduction:

Hyperintensity of optic nerve heads on Diffusion Weighted Imaging (DWI) has been described for papilledema. The purpose of this study was to evaluate if similar changes are associated with other causes of disc edema specifically optic neuritis and anterior ischemic optic neuropathy.

Methods:

In this retrospective study, 18 controls, 16 patients with optic neuritis, 18 patients with papilledema, and 4 patients with anterior ischemic optic neuropathy who had undergone magnetic resonance imaging of brain within 4 weeks of presentation were included. Two neuro-radiologists blinded to the diagnosis independently reviewed the DWI for the presence of hyperintense signal at each optic nerve head as well as the retrobulbar optic nerve. These groups were compared for the prevalence of optic nerve head or retrobulbar nerve hyperintensity.

Results:

For both readers independently, presence of likely or definite hyperintensity of optic nerve head on DWI had sensitivity of 5.9% for detection of papilledema and 35.3% for detection of optic neuritis. This finding was not specific for papilledema or optic neuritis based on the first reader who reported likely hyperintensity in 2/36 (5.5%) control eyes; the second reader did not report this in any control eyes. Also, none of the six eyes with anterior ischemic optic neuropathy showed this finding. Sensitivity of presence of hyperintensity of retrobulbar optic nerve on DWI in detection of optic neuritis was 0% for the first reader and 17.6% for the second. This finding of hyperintensity of retrobulbar optic nerve was specific for optic neuritis, with none of control eyes, eyes with papilledema, or those with anterior ischemic optic neuropathy demonstrating this finding.

Conclusions:

Hyperintensity of the optic nerve head on DWI can be seen with both papilledema and optic neuritis. In optic neuritis, hyperintensity of optic nerve head may be seen in absence of corresponding hyperintensity in retrobulbar optic nerve. In our study, we found hyperintense signal on DWI in retrobulbar optic nerve to be specific for optic neuritis.

References:

Viets R, Parsons M, Van Stavern G, Hildebolt C, Sharma A. Hyperintense optic nerve heads on diffusion-weighted imaging: a potential imaging sign of papilledema. *AJNR Am J Neuroradiol.* ;34(7):1438-42, 2013.

Keywords: Neuroimaging, Optic Neuropathy, Pseudotumor, Demyelinating Disease

Financial Disclosures: The authors had no disclosures.

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Poster 175**Cerebral Correlates of Melanopsin-Mediated Retinal Photoreception**

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Introduction:

Intrinsically-photosensitive retinal ganglion cells (ipRGCs) expressing melanopsin, mediate non-visual photic responses, subserving circadian functions and pupillary reactions to light. The cerebral correlates of the retinal ipRGCs reception are not known, in healthy individuals.

Methods:

In order to specifically select the responses of the ipRGCs, we have performed a fMRI study, and scanned 17 healthy subjects, who were exposed to perceptually similar white lights (metamer-like lights) with high and low levels of melanopic excitation (High_Mel versus Low_Mel), allowing minimal excitation differences for cones and rods.

Results:

The contrasts between High_Mel and Low_Mel revealed significant bilateral activation of the frontal eye fields, suggesting that ipRGCs stimulation might enhance cortical signaling in areas of the brain that are known to be involved in attention and ocular-motor responses. Other activated regions included areas in the inferior temporal gyri, caudate nuclei and the pineal gland.

Conclusions:

Our study suggests that ipRGCs stimulation activates not only the expected cerebral areas involved in circadian rhythms, but also cerebral regions classically involved in attentional and ocular motor responses.

References: None.

Keywords: Melanopsin, fMRI

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 176

Optic Nerve Magnetic Resonance Imaging Characteristics in Inherited Optic Neuropathies

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Introduction:

Inherited optic neuropathies include dominant optic atrophy (DOA), Leber's hereditary optic neuropathy (LHON), and Wolfram syndrome (DIDMOAD). DOA is the most common of these, and mutations in *OPA1* account for 40-60% of DOA cases. Few studies have examined or compared findings on neuroimaging of the optic nerves in inherited optic neuropathies.

Methods:

Using an updated retrospective database of 111 patients with bilateral optic atrophy referred for genetic testing, magnetic resonance (MR) images were analyzed and compared across genotype groups. Patients were screened using next generation sequencing of 243 genes (including *OPA1*, *WFS1*) and the mitochondrial genome (includes all LHON mutations). T2 signal was quantified in MR images (3T) of the orbits and/or brain and normalized to internal standards within each slice. A sample of patients without ocular, central nervous system or visual diagnoses was used to validate the T2 quantification methods.

Results:

Eight patient with and 19 without *OPA1* mutations had MR images available for analysis. There were 2 patients with DIDMOAD and 1 patient with LHON in our sample who also had MR images available. The optic nerves of optic atrophy patients appeared smaller with increased normalized T2 signal compared to controls. There was a wide range of variability in the intensities of optic nerve T2 signals among optic atrophy patients.

Conclusions:

Increased T2 signal intensity is not a typical feature of neurodegenerative disease and may reflect nuances in optic atrophy specific to this group of conditions. Differences in T2 signal could represent differences inherent to distinct hereditary optic neuropathies and/or the degree of atrophy or gliosis. Further characterization of these differences on MR imaging can help guide diagnostic genetic testing and provide insight into differences in the pathophysiology of hereditary optic neuropathies.

References: None.

Keywords: Optic Neuropathy, Genetic Disease, Neuroimaging

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Grant Support: None.

Poster 177**Sarcopenia and Radiographic Changes in the Aging Orbit**

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Introduction:

Sarcopenia is defined as age related muscle wasting and is in part characterized by fat infiltration of skeletal muscle. This study investigated age related magnetic resonance imaging (MRI) changes to assess for fatty infiltration of the extraocular muscles as evidence of sarcopenia.

Methods:

In this retrospective study, the electronic medical records were used to identify 108 patients (216 orbits) who underwent orbital MRI. Patients with pathology involving orbit muscle or fat were excluded. For coronal, non-contrast enhanced, T1 weighted images, the region of interest (ROI) tool was used to measure signal intensities within the medial rectus muscles and intraconal fat. The muscle-fat proportion was calculated by dividing fat by muscle signal strength. Patients were divided into four groups: age 18 to 30 years (G1), 31-49 (G2), 50-69 (G3) and ≥ 70 (G4) and mean muscle-fat proportion was calculated for each. Analysis of variance (ANOVA) was used to compare the means between groups.

Results:

Mean proportion of muscle-fat signal increased significantly with advancing age: 0.27 ± 0.02 (G1), 0.29 ± 0.02 (G2), 0.32 ± 0.03 (G3), 0.40 ± 0.06 (G4). Group four was significantly different than all other groups ($P < 0.01$), indicating the medial rectus muscle signal in older individuals more closely matched that of surrounding fat.

Conclusions:

This preliminary data suggests that sarcopenia related fat infiltration of the extraocular muscles may occur and is detectable with MRI. This has implications regarding age related changes in extraocular motility, such as reduced supraduction and blepharoptosis.

References: None.

Keywords: Sarcopenia, MRI, Fatty Infiltration

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 178

A Comparison of Patients with Isolated Diplopia and Giant Cell Arteritis Versus Those from Other Causes

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Introduction:

Diplopia as a presenting feature of GCA is uncommon and reported to occur in 3-6% of all cases [1-3]. Clinicians often face an older patient presenting with acute or sub-acute onset of diplopia. Although GCA is an uncommon cause for double vision [3,5], many individuals obtain a GCA work up including laboratory evaluation to rule out GCA due to its potential for vision loss. This study will evaluate the differences in patients presenting diplopia, subsequently diagnosed with biopsy-proven giant cell arteritis with patients presenting diplopia from other causes.

Methods:

This is a retrospective multi-center study that will enroll patients presenting with diplopia with biopsy proven GCA. For each study group patient, 5 age-matched (+/- 5 years) controls presenting with diplopia without a diagnoses of GCA will be collected. Data on the demographics, presence of other ocular signs and symptoms, other systemic and laboratory markers will be compared between subjects and controls.

Results:

Preliminary data includes 12 patients and 33 controls that were matched with respect to age and race. Of the patients with GCA, third nerve palsies (n=6) and sixth nerve palsies (n=5) were most common. Patients with GCA were more likely to present with headache (100%), jaw claudication (75%), malaise (42%), myalgia (58%), scalp tenderness (58%), neck pain (42%), and PMR (33%) as well as elevated markers (ESR 75%,; CRP 83%). Ocular ischemic lesions were common in patient with GCA (n=4) presenting as AION, PION, cotton wool spots, and choroidal ischemia. Risk factors such as hyperlipidemia, hypertension, type 2 diabetes and smoking history were not significant among the two groups.

Conclusions:

Patients with diplopia from GCA are more likely to present with systemic symptoms, ocular ischemia, and elevated markers of inflammation and should be promptly evaluated and treated when GCA is suspected.

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Keywords: Giant Cell Arteritis, Diplopia, 6th Nerve Palsy, 3rd Nerve Palsy, Skew

Financial Disclosures: The authors had no disclosures.

Grant Support: Research to Prevent Blindness.

Poster 179

Prevalence of Thymoma in Ocular Myasthenia Gravis

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Introduction:

The prevalence of thymoma in ocular myasthenia gravis has not been explicitly investigated in the medical literature. Published data vary and chiefly consist of smaller case series. The purpose of this study is to determine the prevalence of thymoma in a large cohort of patients with ocular myasthenia gravis.

Methods:

A retrospective multicenter analysis was conducted. We reviewed charts of 158 patients who met diagnostic criteria for ocular myasthenia gravis without generalized disease at initial presentation. The main outcome measure was the percentage of patients with thymoma as diagnosed by chest imaging. Conversion to generalized disease and duration of follow-up also were evaluated. These data will be added by review of additional charts. The medical literature was searched for articles or abstracts in the English language containing data regarding the prevalence of thymoma among other cohorts of patients with ocular myasthenia gravis.

Results:

Of the 158 patients in our cohort, 8 (5.1%) were found to have thymoma. Of these, 3 (37.5%) had disease progression to generalized myasthenia gravis. Nine case series of patients presenting with ocular myasthenia gravis were analyzed. Cohort size varied from 15 to 147, with prevalence of thymoma ranging from 0% to 20%.

Conclusions:

Among our patients presenting with ocular myasthenia gravis, approximately 5% were found to have thymoma. This was distinct from the next largest case series published but not inconsistent with the body of literature. Previous studies have suggested that thymoma may have prognostic implications indicating increased likelihood of generalization of disease and early identification may allow resection prior to the onset of invasive thymoma.

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Keywords: Myasthenia, Neuro-Ophth and Systemic Disease, Paraneoplastic Syndromes

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Grant Support: None.

Poster 180

Absence of Bacteria in the Temporal Arteries of Patients with Giant Cell Arteritis

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Introduction:

An infectious trigger for giant cell arteritis (GCA) has been considered but no organism has conclusively been linked to GCA. We have previously presented data at NANOS linking GCA to a strain of bacteria. We performed confirmatory experiments to determine if GCA is caused by a bacterial infection. We cultured superficial temporal artery biopsies (STABs) and analyzed the cultures for bacterial growth. We also used 16S rRNA sequencing technology to identify bacterial genomic sequences in STABs.

Methods:

STABs were collected from 3 groups: Subjects with histopathologic features of GCA (“positive”), subjects with normal STABs and without clinical features of GCA (“negative”), and subjects with normal STABs and with clinical features of GCA (“indeterminate”). A portion of the specimen was sent for histopathologic analysis. Homogenized arteries were plated onto 3 different growth media. DNA samples were used as a template in PCR reactions using bacterial 16S rRNA primers. Investigators performing experiments were masked to the affected status of the samples.

Results:

Eighteen arteries were analyzed: 10 positive; 5 negative; and 3 indeterminate. The only bacteria that could be cultured was *S. epidermidis*. 16s rRNA sequencing failed to identify bacterial DNA sequences in any of the specimens.

Conclusions:

It is unlikely that *S. epidermidis* has a pathogenic role in GCA. PCR sequencing did not identify any bacterial DNA sequences in any of the arteries. These results suggest one of two possibilities, either bacteria do not reside in the temporal arteries of subjects with GCA or 16S rRNA sequencing is not sensitive enough to detect bacterial genomic material from temporal arteries. Although this study does not rule out the possibility that a bacterium causes GCA, it does suggest that it is unlikely that GCA is caused by a live bacterial strain residing in the artery walls of these patients.

References: None.

Keywords: Giant Cell Arteritis, Bacteria, 16s rRNA

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 181**Incidence and Evaluation of Third Nerve Palsy Using a Population-Based Method**

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Introduction:

Among cases of diplopia caused by cranial nerve palsies, oculomotor paresis is the most worrisome because some are the result of life-threatening aneurysms. Although there exists a number of studies on third nerve palsy from academic centers, all are subject to referral bias. The purpose of this study was to determine the population-based incidence and causes of third nerve palsies using a well-established medical records linkage system designed to capture data on patient-physician encounters.^{1,2}

Methods:

The medical records of all patients who were newly diagnosed with third nerve palsy from January 1, 1978, through December 31, 2014, were retrospectively reviewed. Incidence rates were adjusted to the age and gender distribution of the 2010 US white population.

Results:

We identified 150 new cases of third nerve palsy over the 37-year period, yielding an age- and gender-adjusted annual incidence of 4.2/100 000 (95% confidence interval, 3.5-4.9/100 000). The incidence was low among children and young adults. There was a significant increase among patients over the age of 60 ($P < 0.001$), predominantly due to presumed microvascular third nerve palsies. The causes of third nerve palsy in order of frequency were presumed microvascular (40%), trauma (12%), compression from neoplasm (11%), post-neurosurgery (9%), compression from aneurysm (6%), idiopathic (5%), stroke (4%), congenital (3%), pituitary apoplexy (2%), Tolosa-Hunt syndrome (2%), giant cell arteritis (1%) and other (1%). Ten (17%) patients with presumed microvascular third nerve palsies had pupil involvement, while pupil involvement was seen in 16 (64%) patients with compressive third nerve palsies.

Conclusions:

This population-based cohort demonstrates a higher incidence of presumed microvascular third nerve palsies and a lower incidence of aneurysmal compression than previously reported. While compressive lesions had a significantly higher likelihood of pupil involvement, pupil involvement did not exclude microvascular third nerve palsy and lack of pupil involvement did not rule out compressive third nerve palsy.

References:

1. Kurland LT, Molgaard CA. The patient record in epidemiology. *Sci Am.* Oct 1981;245(4):54-63.
2. Melton LJ, 3rd. History of the Rochester Epidemiology Project. *Mayo Clin Proc.* Mar 1996;71(3):266-274.

Keywords: Third Nerve Palsy, Epidemiologic Study

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Grant Support: None.

Poster 182

The CHOP Sign: Saccadic Correction With Horizontal Prism. A Novel Method to Identify Functional Monocular Vision Loss

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Introduction:

This case series describes a novel exam tool and its utility in distinguishing functional monocular vision loss (FMVL) from organic monocular vision loss (OMVL).

Methods:

Patients with monocular vision loss were evaluated. The patient was directed to view a distant target and asked if they could see it with each eye independently. Those who denied seeing the target with the lower acuity eye were included. Alternate cover testing was performed, initially without prism and then a 12-diopter base out prism was introduced over the normal eye. Patients who demonstrated no corrective saccades (- CHOP sign) were presumed to not see the target with the lower acuity eye. Those that demonstrated corrective saccades (+ CHOP sign) were presumed to see the target with both eyes.

Results:

Five patients who demonstrated corrective saccades and five who did not were identified. Pertinent history, exam (acuity, colors, RAPD, fundus, vertical prism dissociation, OKN) and testing results (VFT, OCT, ERG, imaging) are described and led to a diagnosis of FMVL or OMVL for each case. Corrective saccades (+ CHOP sign) reliably identified patients with FMVL and absence of corrective saccades (- CHOP sign) identified patients with OMVL.

Conclusions:

In this case series, the CHOP sign provided an objective method that reliably distinguished FMVL from OMVL.

References: None.

Keywords: Non-Organic Visual Disorders

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 183**Risk Factors of Ischemic Optic Neuropathy in Korean population**

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Introduction:

To analyze the clinical features and risk factors of Korean patients with ischemic optic neuropathy (ION).

Methods:

Medical records of 39 Korean patients (study group) who were diagnosed as having ION and 54 patient (control group) who underwent healthy inspection were retrospectively reviewed and the risk factors of ION including age, sex, associated systemic disease, past medical history, social history, hematologic findings were investigated using statistical analysis.

Results:

The mean age of the 39 patients was 62.44 ± 14.69 years (range, 28-91 years). There were 31 patients (79.4%) with associated systemic diseases such as hypertension (56.4%), hyperlipidemia (41.1%), diabetes mellitus (33.3%), cerebrovascular disease, migraine, and heart disease with a decreasing order. 14 patients (35.8%) with a mean age of 63.41 years were smoker. Risk factor for ischemic optic neuropathy were diabetes mellitus (odds ratio; OR = 4.000, $p=0.009$) and hyperlipidemia (OR = 6.817, $p < 0.001$) smoking history (OR = 3.220, $p < 0.018$). None of risk factors differed significantly between the two groups (hypertension $p=0.064$, IHD $p=0.396$, LVH = $p 0.307$)

Conclusions:

Ischemic optic neuropathy is closely related to diabetes mellitus, hyperlipidemia and smoking in Korean patients, as compared to the control group

References: None.

Keywords: Ischemic Optic Neuropathy, Diabetes Mellitus, Hyperlipidemia, Smoking, Korean Patients

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 184

Neuromyelitis Optica Spectrum Disorder: Disease Course and Long-Term Visual Outcome

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Introduction:

NMO spectrum disorder (NMOSD) is an autoimmune disease that classically manifests as attacks of optic neuritis (ON) and transverse myelitis (TM). The prevalence, course, and severity of NMOSD varies considerably. Few studies report the neuro-ophthalmological disease course and visual outcome. |We describe the course and long-term visual outcome in a cohort of NMOSD patients treated in a single tertiary referral center

Methods:

The database was searched for all patients with NMOSD who were treated in our center from 2005 through 2014. Data collected included detailed visual outcome, grade of final visual disability, neuroimaging and optical coherence tomography results. Details on relapses, acute episodes and maintenance therapies were recorded

Results:

.Of the 18 patients with NMOSD who were followed for a mean duration of 7.5 years, 14 (78%) were female. Mean age at presentation was 37.6±17.7 years. Patients with acute attacks were treated with high-dose intravenous methylprednisolone and offered immunosuppressive maintenance. Optic neuritis (ON) occurred in 12 patients, with a cumulative total of 37 ON episodes. At the end of the follow-up period, no patient had become legally blind and only one patient had lost her driver's license. Pain associated with acute ON was common (83%) whereas optic disc edema was a rare finding in our cohort (6%).

Conclusions:

In this retrospective series of 18 patients with NMOSD, followed for a mean of 7.5 years, acute-phase treatment was given within 8 days of relapse, followed by maintenance therapy. Good functional vision and driver's license were preserved in 17/18 patients

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Keywords: Neuromyelitis Optica Spectrum Disorder, Optic Neuritis, Visual Outcome

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 185**Repeatability and Validity of the Ice Test in the Evaluation of Ptosis in Myasthenia Gravis**

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Introduction:

The ice test has been reported to show a sensitivity of 90% and a specificity of 100% in the diagnosis of ocular myasthenia gravis (MG). However, there is lack of consensus on the criteria of positive response and little is known of the repeatability of the ice test in myasthenic ptosis and controls. This study was conducted to determine the repeatability and validity of the ice test in diagnosing MG according to different criterias.

Methods:

Thirty-one patients with ptosis related to MG and 38 controls with non-myasthenic ptosis were included. All patients were tested with the ice test twice on separate days in the afternoon. The margin reflex distance (MRD) was measured before and immediately after 5-minute application of ice on the eyelids. The ice test was judged 'positive' if improvement of MRD was ≥ 1 mm (criteria 1) or ≥ 2 mm (criteria 2) after the ice test.

Results:

Repeated ice tests showed consistent results in 68% of MG, and 97% of controls with ptosis. Repeated ice tests increased the sensitivity of diagnosing MG by 18% (criteria 1) and 21% (criteria 2) compared to a single test. The sensitivity and specificity of repeated ice tests were 97% and 87% by criteria 1, and 65% and 97% by criteria 2. There was no significant difference in the results between ocular MG and generalized MG ($p = 0.558$).

Conclusions:

The results of the ice test was variable in one third of myasthenic ptosis. Repeated ice tests may enhance the sensitivity of the test. MRD improvement of 1 mm or more after repeated ice tests is very sensitive for screening MG, and further evaluation is necessary in these patients.

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Keywords: Myasthenia Gravis, Ice Test

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 186

Case Series of Posterior Cortical Atrophy

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Introduction:

Posterior cortical atrophy (PCA), a visual variant of Alzheimer's disease, is an increasingly prevalent disease with today's aging population. Distinguishing between PCA and other causes of visual impairment is essential in the early diagnosis and treatment. Clinical presentation is heralded by subjective vision loss, yet a normal ophthalmic examination - there is not yet definitive diagnostic criteria and it may be underdiagnosed. Through the advances in understanding the early signs and symptoms of this disease, the prospect of early diagnosis, and thus eventually, prevention and treatment is optimistically hopeful.

Methods:

First we conducted a literature review to better understand the signs and symptoms that have been reported. Then, we established a repository to collect patient data to evaluate for commonalities. We have identified 30 patients from 3 academic institutions with the disease thus far and have begun preliminary data analysis.

Results:

Preliminary data analysis demonstrates that patients reported trouble reading (78%) and difficulty with depth perception (44%) as the most common concerns. Recurrent signs included abnormal color plates (100%), simultanagnosia (75%), hemianopsia (44%), decreased stereopsis (100%), and abnormal Amsler's grid (44%). We will presents at a table describing the most common findings in our cohort of patients.

Conclusions:

Earlier diagnosis is key to developing new treatments, hopefully aimed at impacting PCA in a disease modifying state. Additionally, patients can better understand their diagnosis at an earlier stage and make the necessary life style adjustments. The only way to ensure earlier diagnosis is by developing more concrete diagnostic criteria and increasing awareness. We developed the only PCA repository to our knowledge and as we continue to collect cases, we hope this will lead to firm diagnostic criteria and increased awareness.

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Keywords: Posterior Cortical Atrophy, Alzheimer's Disease, Simultanagnosia, Benson's Disease, Visual Impairment

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 187**Evaluating the Incidence of Arteritic Ischemic Optic Neuropathy and Other Causes of Vision Loss From Giant Cell Arteritis**

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Introduction:

Giant cell arteritis (GCA) is a potentially blinding condition, so determining the incidence of permanent vision loss, especially arteritic ischemic optic neuropathy (AION), is important and may help guide the screening and treatment of this disease. The goal of this study is to evaluate the population-based incidence of AION and other causes of permanent vision loss from GCA using a well-established medical records linkage system designed to capture data on patient-physician encounters.^{1,2}

Methods:

The medical records of all patients who were newly diagnosed with GCA from January 1, 1950, through December 31, 2009, were retrospectively reviewed to identify and determine the incidence and causes of permanent vision loss. Systemic symptoms of GCA and visual outcomes were also determined.

Results:

Among the 245 new cases of GCA over the 60-year period, 20 (8.2%) patients suffered permanent vision loss due to GCA. The occurrence rate of AION was 6.9% (95% CI: 4.0% – 11.1%) accounting for 85% of cases of permanent vision loss. The occurrence rate of central retinal artery occlusion (CRAO) was 1.6% (95% CI: 0.4% – 4.2%) and cilioretinal artery occlusion was 0.4% (95% CI: 0.01% – 2.3%). The population-based age- and sex-adjusted annual incidence of AION among persons age ≥ 50 years was 1.3 (95% CI: 0.7 – 2.0) per 100,000 population. 20% of patients with permanent vision loss from GCA had an occult presentation without constitutional symptoms. Overall, there was no significant difference between presenting and final visual acuities.

Conclusions:

Using population-based data, we provide the most accurate incidence of permanent vision loss from GCA. This study confirms that visual outcomes from GCA-related vision loss are poor and that 20% of patients with permanent visual loss from GCA can present without systemic symptoms of GCA.

References:

1. Kurland LT, Molgaard CA. The patient record in epidemiology. *Sci Am.* Oct 1981;245(4):54-63.
2. Melton LJ, 3rd. History of the Rochester Epidemiology Project. *Mayo Clin Proc.* Mar 1996;71(3):266-274.

Keywords: Arteritic Ischemic Optic Neuropathy, Giant Cell Arteritis, Occult

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 188**Yield of the Clinical Neuro-Ophthalmologic Examination in Patients with Concussion**

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Introduction:

Due to the widespread nature of ocular motor anatomic pathways, traumatic brain injury and its mildest form, concussion, affect eye movements in many ways. However, the extent to which concussion results in ocular motor deficits detectable on clinical examination is unclear. Existing literature suggests a high incidence of convergence insufficiency, saccadic deficits, and smooth pursuit impairment. We sought to identify the incidence of ocular motor dysfunction in a multidisciplinary concussion center-based neuro-ophthalmologic practice.

Methods:

We performed a retrospective chart review of all patients with a concussion-related diagnostic code seen in the practice of four neuro-ophthalmologists between 8/1/2014 and 8/1/2015. Those with pre-existing ocular motor deficits or significant positive neuroimaging findings (i.e. subdural or intracranial hemorrhage, orbital fractures) were excluded from the analysis.

Results:

Seventy patients with a concussion-related diagnosis were identified. Fifty-eight subjects (mean age 30.8 ±14.7, age range 11-65, 30 males, 28 females) met inclusion criteria. Loss of consciousness was documented in 13 (22.4%). Twenty-four (41.4%) reported a history of multiple concussions. The most common symptom was headache (n=36, 62.1%), followed by photosensitivity (n=19, 32.8%). Difficulty reading or using screens were reported in 11 (19%) and 9 (15%), respectively. Ocular motor dysfunction was seen on neuro-ophthalmologic examination in 16 (27.6%) patients; findings predominantly included convergence insufficiency (n=13, 22.4%). Reduced stereopsis, ocular flutter, impaired VOR cancellation, and gaze-evoked nystagmus were each found in single patients. One patient had square wave jerks, but it was unclear if these were pathologic. Deficits of saccades were not identified by clinical examination.

Conclusions:

Results of this neuro-ophthalmic concussion study are concordant with existing literature showing a fairly high incidence of convergence insufficiency. Interestingly, other eye movement abnormalities were rare and saccadic deficits were not detected, emphasizing the need for additional performance measures or eye movement recordings to capture concussion-related efferent visual deficits.

References: None.

Keywords: Trauma, Ocular Motility

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 189

Visual Performance of Non-Native versus Native English Speakers on a Sideline Concussion Screen: An Objective Look at Eye Movement Recordings

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Introduction:

The King-Devick (KD) test of rapid number-naming, a sensitive and rapid performance measure, adds a visual dimension to sideline testing for concussion. We performed a laboratory-based eye movement study during performance of the KD test. We sought to determine if having English as a second language results in slower KD reading times or changes in eye movements.

Methods:

We tested 27 native English speakers (NES) (mean age 32) and 27 subjects for whom English was a second language (ESL) (mean age 34). Participants had no history of concussion. Participants performed a computerized version of the KD. Simultaneous infrared-based videoculographic (VOG) recordings were performed using the Eyelink1000+. A Bilingual Dominance Scale survey, which quantifies primary versus secondary language dominance, was completed by all ESL subjects.

Results:

Digitized KD reading times were significantly prolonged for ESL participants, compared to NES (54.4 ± 15.4 sec vs. 42.8 ± 8.6 sec, $p=0.001$, t -test). Average intersaccadic intervals (ISI), a combined measure of saccadic latency and fixation duration, were significantly longer for ESL participants (402 ± 116.9 msec vs. 317.7 ± 53.9 msec, $p=0.002$, t -test). The total number of saccades for ESL participants was significantly higher (149 ± 28 vs. 135 ± 18 , $p=0.03$, t -test).

Conclusions:

This study highlights performance disparities that linguistics may impose on rapid number-naming tasks. Concussion screening is best implemented by establishment of pre-season baselines to allow for intra-subject comparisons after impact in sport. If pre-season baseline data are unavailable, caution should be taken in comparing non-native English speaker reading times to a NES normative control KD time database.

References: None.

Keywords: Trauma, Ocular Motility

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 190**Historical Trends in the Diagnosis of Peduncular Hallucinosi**

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Introduction:

Peduncular Hallucinosi (PH) refers to dream-like visual hallucinations during wakefulness that are typically caused by a focal lesion in the rostral brainstem. The conventional understanding of PH is that associated eye movement abnormalities are frequently, if not always, present. However, in the era of modern neuroimaging, PH has been increasingly diagnosed in patients without any objective deficits on physical examination. We sought to determine how modern neuroimaging has impacted the diagnosis of PH.

Methods:

We reviewed all available cases of PH, beginning with Lhermitte's original description in 1922. Papers reported in languages aside from English were translated. Patients with afferent visual loss, toxic exposure, and pre-existing psychiatric disease were excluded from the analysis. We grouped the patients according to whether or not a neuroimaging study had been performed. We then determined the frequency of objective clinical abnormalities among patients in each group.

Results:

A total of 95 cases were collected, of which 84 cases met criteria for inclusion in the study. Twenty-eight reports were translated from 7 different languages. Among cases diagnosed without a neuroimaging study, 16/21 (76%) had associated eye movement abnormalities. However, among modern cases that included CT or MRI imaging, associated eye movement abnormalities were significantly less frequent, occurring in only 22/63 (34%; $p=0.001$).

Conclusions:

Although eye movement abnormalities have historically been considered a key clinical feature supporting the diagnosis of PH, we found that in the era of modern neuroimaging, co-occurring eye movement abnormalities are far less frequent and are not a requisite feature of the diagnosis.

References: None.

Keywords: Peduncular Hallucinosi, Hallucinations, Magnetic Resonance Imaging

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 191

Opening Pressure Assessment Following Venous Stenting for Idiopathic Intracranial Hypertension

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Introduction:

Increasing evidence supports a role of venous sinus stenosis in the pathogenesis of idiopathic intracranial hypertension (IIH). Numerous retrospective studies have demonstrated improvement in symptoms and papilledema after stenting, but few have diligently compared pre and post opening pressure (OP). We measured the pre and post OP in 19 patients who underwent stenting for IIH, 10 of whom were enrolled in a prospective trial evaluating the effect of stenting on visual fields and optic nerve morphology

Methods:

All patients fulfilled the modified Dandy criteria for IIH and were either refractory or intolerant to medical therapy or presented with fulminant vision loss. Lumbar puncture (LP) was performed prior to venous stenting, and again at approximately 3 months.

Results:

Mean age at presentation was 27.5 and all patients were female. Mean BMI increased from 36.0 to 36.8. Stenosis was bilateral in 10 (52.6%) and unilateral on the dominant side in 9. Mean OP reduced from 40.4 (SD 11.3) to 20.9 (SD 5.72) with a mean change of -19.47 cm H₂O. Median OP reduced 45% from 39 to 21 cm H₂O (P<0.0001, Wilcoxon signed rank-sum test). Bivariate analysis found a lower pre-stent BMI in those patients who demonstrated a post-stent absolute change in OP > 25. (38.6 vs. 28.9, P<0.03). Pre-stent BMI correlated with %change in OP (Pearson Correlation coefficient: -0.63, P = 0.004). 2/19 (10.5%) patients developed a new stenosis during follow up.

Conclusions:

The clinical improvement observed with venous stenting in IIH patients is accompanied by an objective reduction in OP. The % improvement in OP weakly correlates with pre-stent BMI (but not pre-stent OP), suggesting that the degree to which the stenosis contributes to the elevation in OP may be smaller in more severely obese patients. A prospective trial comparing venous stenting to shunting is needed to confirm its efficacy in OP reduction.

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Keywords: Idiopathic Intracranial Hypertension, Venous Stenting, Lumbar Puncture, Opening Pressure

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 192**Multiple Sclerosis Patients with Depression are More Likely to Subjectively Rate Their Vision Poorly**

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Introduction:

Depression is a common condition in patients with multiple sclerosis (MS), and is routinely assessed in our MS center using the Patient Health Questionnaire-9 (PHQ-9). Patient-reported outcomes (PRO) are increasingly utilized in clinics and in clinical trials to assess the impact of neurological symptoms on patients' function. The aim of this study was to determine if depression negatively affects a patient's rating of their visual function.

Methods:

This was a retrospective, observational study of patients diagnosed with MS or clinically isolated syndrome (CIS). A clinic database and electronic medical record were used to identify patients with a PHQ-9, multiple sclerosis performance scales (MSPS) vision subscore, and visual acuity performed within 3 months. MSPS vision subscore was dichotomized into two categories (0 or 1 indicating better vision vs 2, 3 or 4 indicating poorer vision) and PHQ-9 was dichotomized into two categories (≥ 10 indicating depression vs < 10 , no or mild depression). Visual acuity was measured using near card assessment of best corrected visual acuity. Logistic regression models were used to assess the relationship between dichotomized MSPS vision subscore, visual acuity, and PHQ-9.

Results:

363 patients met the inclusion criteria. In the univariate analysis, patients with a dichotomized PHQ-9 score ≥ 10 were 4.1 times more likely to have a MSPS vision in the higher categories ($p < 0.001$). In the multivariate analysis, visual acuity (worst eye) and PHQ-9 score (continuous) were both predictive of a worse vision rating. For every 1 point increase in PHQ-9 score, the odds of being in the poor MSPS vision subscore category was 1.122 times that of the better category ($p < 0.0001$).

Conclusions:

This study shows that patients with depression may overestimate their visual disability on patient-reported outcomes. Future studies should evaluate for this effect in more comprehensive patient-reported measures of visual function.

References: None.

Keywords: Demyelinating Disease, Depression, Visual Acuity

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Poster 193

Causes of Eye Pain: A Multicenter Center Experience

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Introduction:

Eye pain is a common presenting complaint caused by eye disorders such as keratitis and dry eye as well as neurological disorders such as tumors and headache. Although a common symptom, no previous study has reviewed the causes of eye pain in ophthalmology and neurology clinics from large referral institutions. We aimed to determine the causes of eye pain presenting to two tertiary eye and neurologic centers.

Methods:

This cross-sectional study analyzed patient records from the Departments of Ophthalmology and Neurology at two major universities. Entry criteria included all patients seen in those departments between January 2012 to December 2013 with the main complaint of eye pain. We had IRB approval.

Results:

We screened 16,482 patient files at U2 and 62,532 files at the U1. We analyzed the causes of eye pain in 1488 patients from U2 and 814 patients from U1. 1385/1488 (93%) of the U2 patients and 584/814 (72%) U1 were from ophthalmology; 103/1488 (7%) U2 patients and 230/814 (28%) U1 patients were from neurology. The three most common ophthalmologic diagnoses at U2 were conjunctivitis (27%), keratitis (11%), and blepharitis (8%), and the three most common neurologic diagnoses were optic neuritis (2.5%), zoster ophthalmicus (1.1%) and migraine (0.6%). The three most common ophthalmologic diagnoses at U1 were dry eye (13%), migraine (9%), and blepharitis/keratitis (6%), and the two the most common neurologic diagnoses were migraine/headache (61%) and optic neuritis (5%).

Conclusions:

Eye pain was caused by ocular inflammation in the ophthalmology clinic in both institutions; however, more migraine was identified at U1. In the neurology clinics, optic neuritis and migraine were the most frequent causes in both institutions. These findings may help neurologists and ophthalmologists look for the most frequent causes of eye pain in their populations. Different referral bias and patients may account for some of the differences between institutions.

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Keywords: Eye Pain, Dry Eye, Migraine, Optic Neuritis, Conjunctivitis, Keratitis

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 194

Retinal Nerve Fiber Layer Thickness in HTLV-1 Patients

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Introduction:

The Human T-cell leukemia virus type 1 (HTLV-1) is the first discovered human retrovirus. Although the vast majority of infected individuals are asymptomatic carriers (a.c.HTLV-1), HTLV-1 can induce Tropical Spastic Paraparesis/HTLV-1 Associated Myelopathy (TSP/HAM), a progressive, inflammatory, and demyelinating myelopathy. HTLV-1 intermediate uveitis is the most frequent ophthalmic manifestation, in the setting of which optic disc edema can be observed. An actual acute optic neuropathy (ON) can also occur during the course of TSP/HAM, the whole clinical presentation mimicking a progressive form of multiple sclerosis. Previous reports have suggested subclinical involvement of the optic nerve in HTLV-1 patients using visual evoked potential. The goal of our study was to determine whether there is subclinical ON in a.c.HTLV-1 and TSP/HAM patients using retinal nerve fiber layer (RNFL) thickness.

Methods:

We prospectively included a.c.HTLV-1 and TSP/HAM patients between January 1st,2014 and March 31st,2015. All patients had a complete eye examination. Visual acuity (VA) and RNFL thickness were measured and compared to age- and sex-matched control groups including patients seen in our refraction clinic with no previous medical or surgical history.

Results:

Twenty-nine a.c.HTLV-1 (group1) and 29 TSP/HAM patients (group2) were included. Average RNFL thickness was 99.9+/-14.3 µm in group1 and 87.8+/-19.2 µm in group2. Average RNFL thicknesses were lower in both groups, when compared to controls. The difference was significant in patients with TSP/HAM (87.8+/-19.2 vs. 97+/-7.8 µm;p=0.003) who also had significantly decreased VA.

Conclusions:

We report here the first study about RNFL thickness in patients with TSP/HAM. In these patients, there is decrease of the RNFL thickness with subtle but definite decrease of VA. This suggests that subclinical ON occurs in the natural history of the disease. The diagnosis of TSP/HAM must be evoked as a differential of primary progressive multiple sclerosis in a population at risk. Moreover, RNFL thinning with no evidence of glaucoma should raise suspicion for HTLV infection and TSP/HAM in a population at risk.

References: None.

Keywords: Optic Neuropathy, HTLV-1, TSP/HAM

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 195

Sabril Vision Study: Retinal Structure and Function in Adult Patients with Refractory Complex Partial Seizures (rCPS) Treated with Vigabatrin

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Introduction:

Visual deficits have been reported in retrospective, cross-sectional studies in vigabatrin-treated patients. This study evaluated visual-field (VF) and retinal-structure changes following adjunctive vigabatrin treatment in vigabatrin-naïve adults with rCPS.

Methods:

Prospective, longitudinal, single-arm, open-label study (NCT01278173). Eligible patients (≥ 2 seizures/month who failed ≥ 3 therapies) performed reliable perimetry (Humphrey automated static) and retinal-structure assessment (spectral-domain optical coherence tomography) pre-vigabatrin exposure. Following vigabatrin initiation, testing occurred within 1 month (reference) and at 3, 6, 9, and 12 months. Endpoints included: mean change from reference in mean deviation (dB) and average retinal nerve fiber layer (RNFL) thickness, visual acuity (VA) changes from baseline, and number of patients who met pre-defined vision-parameter changes at 2 (confirmed) or 3 (persistent) consecutive visits.

Results:

Sixty-five of 91 screened patients received ≥ 1 vigabatrin dose (all-patients-treated set [APTS]); 55 had ≥ 1 valid reference and post-reference assessment (full-analysis set [FAS]). Thirty-six APTS patients with valid pre-/post-reference values completed all planned visits (per-protocol set [PPS]). Thirty-eight (59%) APTS patients completed the study; 27 (42%) withdrew (none for VF changes); 32% and 15% had abnormal RNFL thinning and VA at baseline, respectively; 20% had abnormal near VFs vs reference. No significant mean near-VFs changes were observed (PPS); mean change in average RNFL thickness *increased* significantly (1-year data: Left-eye: 6.37 μ m, CI:[4.66,8.09]; Right-eye: 7.24 μ m CI:[5.47,9.01]; PPS). No confirmed 3-line decreases in VA (FAS) were observed; 3 patients had pre-defined confirmed/persistent near-VF changes (FAS). All vision-related AEs were non-serious; the most common was vision blurred (9%).

Conclusions:

Prior to vigabatrin initiation, rCPS patients may already exhibit vision deficits. Adjunctive vigabatrin treatment (up to 1-year) did not significantly change population near VFs (despite 3 patients meeting pre-defined near-VF-change criteria). RNFL thickening (unknown clinical significance) was observed. Limitations include: single-arm, open-label design; patients' inability to perform ophthalmic/VF examinations; and short-term vigabatrin-exposure duration. Funding: Lundbeck, LLC

References: None.

Keywords: Vigabatrin, Refractory Epilepsy, Vision Study, Perimetry, Visual Field

Financial Disclosures: Sergott: Consultant and speakers' bureau: LundbeckWechsler: Ad boards and consulting: Cyberonics, Eisai, Gerson Lehrman, Lundbeck, Sunovion, UCB, Upsher-SmithDribinsky:Salary, LundbeckWhittle: Salary, LundbeckFeng: Salary, LundbeckLee: Salary, LundbeckIsojarvi: Salary, Lundbeck.

Grant Support: None.

Poster 196

Patterns of Change in Macular Thickness on OCT in Normal Pregnancy and Pregnancy Complicated by Hypertension: A Tool for Assessing Dynamic Function of the Microcirculation

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Introduction:

The macula is a capillary bed with a microcirculation homologous to that of CNS tissues. Unlike arterioles of most organs, the eye and brain autoregulate (AR) capillary transudation such that it remains within physiologic limits despite large variations in pressure and flow within the systemic circulation. Pregnancy is accompanied by 40-60% greater blood volume and cardiac output and increased capillary permeability. These test the integrity of the AR reflex. Our study assesses whether SD-OCT measurements of retinal macular thickness can profile dynamic changes in AR response in pregnancy.

Methods:

Sequential SD-OCT measurements of macular thickness were obtained in over 100 pregnant women; 32 have complete data at gestational ages < 20 wks, 20-40 wks, at delivery and ≥ 2 wks post delivery. Women with normal pregnancies, pre-existing hypertension and pregnant, and hypertensive disorders of pregnancy were included. Differences in retinal thickness from the internal limiting membrane to the retinal pigment epithelium in each ETDRS segment were compared. Clinically meaningful change was defined as $\pm 4\mu\text{m}$ (Test-Re-test Coefficient of Repeatability) in 3 or more contiguous segments.

Results:

The retina thins 5-15 μm in early normal pregnancy (14 patients). Two patients with pre-existing, poorly controlled hypertension had delayed thinning, but only with progressive elevations in blood pressure (BP). Retinae thickened in 10 patients with preeclampsia/severe hypertension with capillary leak occurring into macular tissues even before BP started rising. As retinal edema worsened near term, BPs rose precipitously.

Conclusions:

Preliminary data show SD-OCT is able to characterize changes in retinal thickness in response to hemodynamic events accompanying normal pregnancy. Arteriolar constriction due to capillary permeability left-shifts the AR curve in early normal pregnancy. This differs from renal AR. AR fails in most preeclampsia. Insights gained from this work may enhance understanding of a broad spectrum of disorders involving the microcirculation, including preeclampsia, hypertension, diabetes and dementia.

References: None.

Keywords: Pregnancy, Hypertension, Preeclampsia, Autoregulation, Microcirculation

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 197

The Association Between Multiple Sclerosis and Uveitis

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Introduction:

Uveitis and multiple sclerosis (MS) are considered to be somewhat associated. What is the prevalence of MS-associated uveitis? We set out to study this question through a systematic search of contemporary literature.

Methods:

A systematic search of the database Pubmed.com using the following MeSH-terms '[Multiple Sclerosis]' AND '[Uveitis]' was undertaken. All studies with a prevalence of MS-associated uveitis were included. Of the 215 original hits 26 studies were included. Additional 6 studies from the related citations were included.

Results:

The prevalence of MS in patients with uveitis differs from 0,7% to 30,4%, while the prevalence of uveitis in patients with MS differs from 0,65% to 36,7%. All types of uveitis are seen among the patients with multiple sclerosis.

Conclusions:

The 32 studies are difficult to compare due to differences in the size of the cohort, differences in study design as well as different diagnostic criteria for MS and especially uveitis. Based on the biggest retrospective studies the prevalence of MS-associated uveitis seems to be approximately 1% - both for patients with uveitis as well as for patients with MS. To further investigate the association between MS and uveitis bigger prospective studies are needed.

References: None.

Keywords: Multiple Sclerosis, Uveitis

Financial Disclosures: The authors had no disclosures.

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Poster 198**Asymmetric Fundus Autofluorescence Findings in Parkinson's Disease**

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Introduction:

Motor asymmetry is one of the criteria for the diagnosis of Parkinsons disease (PD). Interocular asymmetry in spectral domain optical coherence tomography (SD-OCT) of the retina and possibly nerve fiber layer has also been documented.

Methods:

The files of 20 PD patients diagnosed at other neurology clinics were examined. Fundus autofluorescence (FAF) images of these patients were observed by two ophthalmologists in a masked fashion. Hyper or hypofluorescent lesions which suggested neurodegeneration were taken into consideration. The mean age of the patients was 69.

Results:

In 15 patients hypo and hyperfluorescent lesions were on the nasal side of the retina. In patients who had early PD, degenerative lesions were on the nasal side. Diffuse degeneration on FAF was detected in the middle stages of the disease. 5 patients had this kind of distribution. This asymmetric appearance was in parallel with the asymmetric findings in motor functions.

Conclusions:

Since FAF detects lipofuscin in the retina, the images were consistent with retinal damage. Lipofuscin gives damage to the tissues by mechanically obstructing the flow into and out of the cells and slowing down the elimination of waste materials. The predilection of neurodegeneration for the nasal retina and and the asymmetric appearance on FAF was not reported before, to the best of our knowledge. Studies with larger series may give important information about the early detection of PD by ophthalmologic examination. Our study suggests that imaging of the nasal retina early in the disease may reveal findings consistent with the asymmetric nature of PD.

References: None.

Keywords: FAF, Parkinson's, Asymmetry, Retina

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 199

Visual Symptoms and Movement Disorders: Establishing a Neuro-ophthalmology Clinic Designed for Patients with Movement Disorders

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Introduction:

We aim to share knowledge gained with the development of a subspecialty clinic within neuro-ophthalmology for patients with visual complaints in the setting of a movement disorder, including: Idiopathic Parkinson's Disease (IPD), Progressive Supranuclear Palsy, dementia with Lewy Bodies, Parkinsonism, ataxia, and others. Barriers to the delivery of high-quality, neuro-ophthalmic care that is patient-centered include the multifactorial, fluctuating, and progressive nature of the visual symptoms and the need for extended appointments to accommodate fatigue and cognitive impairment. To address these barriers, we established a monthly neuro-ophthalmology clinic dedicated to patients with movement disorders.

Methods:

Based on input from ophthalmic technicians, movement disorder specialists, optometry, and patients, we began a monthly, neuro-ophthalmology clinic with specialized facets of care for patients with movement disorders. The clinic is staffed by a neuro-ophthalmologist, an optometrist, a movement disorders fellow, and an ophthalmic technician. Standardized procedures were introduced and are being refined and developed, including the use of the Self-Administered Gerocognitive Exam (or SAGE) and disease-specific rating scales.

Results:

Ease of scheduling, defined goals for visits, and patients' perception of care were subjectively improved. Standardized data collection is ongoing. Most patients referred have IPD or Parkinsonism and complain of problems driving, reading and/or double vision, while others are referred specifically to aid in the diagnosis of the primary movement disorder. Cognitive impairment is common.

Conclusions:

We believe the establishment of a neuro-ophthalmology clinic to address the unique needs of patients with movement disorders will ease the inherent burdens associated with evaluating, diagnosing, and treating visual complaints in this population. Cognitive impairment may be more common than expected and/or contributing to visual complaints in this unique population. Standardized data collection will aid in the delivery of high-quality, patient-centered, neuro-ophthalmic care and increase our understanding of the causes and management of visual complaints in this population.

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Keywords: Quality of Care, Visual Complaints and Movement Disorders, Multispecialty Neuro-Ophthalmology Clinic, Patient-Oriented Care, Cognitive Impairment

Financial Disclosures: The authors had no disclosures.

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Poster 200

Pseudotumor Cerebri Secondary to Venous Sinus Occlusion

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Introduction:

Pseudotumor cerebri (PTC) is characterized by increased intracranial pressure (ICP) without any focal neurological sign and symptom with normal cerebrospinal fluid constituents. We are presenting cases of pseudotumor who had cerebral venous sinus occlusion as a cause of raised ICP.

Methods:

40 cases of PTC were followed up for a period of one to two years. In all patients a detailed neuro-ophthalmic examination and lab investigations were performed. Contrast enhanced MRI & MRV, MRA were carried out in all cases. In cases of venous sinus stenosis the pressure gradient across the stenosis was recorded. Finally all cases of venous sinus occlusion were subjected to digital subtraction angiography (DSA).

Results:

Out of 40 cases of PTC there were 16 cases where cause of raised ICP was venous sinus occlusion. Out these 16 cases, 3 had right transverse sinus stenosis with significant pressure gradient >10cm water while 13 cases had superior sagittal sinus and / or transverse sinus thrombosis. Out of these 13 cases of venous sinus thrombosis, 4 cases underwent venous sinus stenting with glued occlusion of secondary dural AVM (Arterio venous malformation) as they had rapidly falling vision which could not be controlled on maximal medical treatment. In 9 cases of venous sinus thrombosis PTC resolved over a period of 6 months on medical treatment.

Conclusions:

After stenting of venous sinus occlusion if signs and symptoms of increased ICP reappear then one should suspect secondary dural AVM formation. When the medical treatment fails and visual loss is rapid then lumboperitoneal shunt or optic nerve fenestration may be considered prior to stenting of venous sinus. As some of our cases could not undergo stenting on time due to financial constraint, had irreversible visual loss.

References:

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Keywords: Pseudotumor Cerebri, Cerebral Venous Sinus Thrombosis, Cerebral Venous Sinus Stenosis, Venous Sinus Stenting

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 201

Comparison of the Surgical Treatment of Superior Oblique Palsy: Surgery Versus Combined Preoperative OnabotulinumtoxinA Injection

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Introduction:

To compare extraocular muscle (EOM) surgery with combined preoperative OnabotulinumA injection to the ipsilateral inferior oblique (IO) muscle for the treatment of superior oblique (SO) palsy

Methods:

A retrospective review of medical records of patients with SO palsy who underwent surgical treatment between 2006 and 2015. Group 1 (n=12) underwent preoperative OnabotulinumtoxinA injection to the ipsilateral IO and then EOM surgery to correct hypertropia, while group 2 (n=12) underwent EOM surgery only. Comparisons were made of preoperative and postoperative vertical deviation angles in primary position and the amount of corrected hypertropia, postoperative drift and success rate (defined as less than 4 PD).

Results:

Preoperatively, there was no statistically meaningful difference in mean vertical deviation in primary position between the two groups (13.0 PD and 13.5 PD, respectively ($p>0.05$)). Mean vertical deviations in primary position at postoperative 1 week measurements were -0.2 PD and 1.8 PD in group 1 and 2 respectively, and -0.7 PD and 0.3 PD in group 1 and 2, respectively, at postoperative 3 months. Mean amount of postoperative drift toward hypertropia in primary position was 0.5 PD and 1.6 PD in group 1 and 2, respectively. Success rate of group 1 was 85% and that of group 2 was 77% ($p>0.05$).

Conclusions:

Both EOM surgery and EOM surgery combined with preoperative OnabotulinumtoxinA injection to the ipsilateral IO muscle were effective in the treatment of SO palsy. Our findings support the condition that there is no reason to avoid preoperative OnabotulinumtoxinA injection while waiting for the EOM surgery in patients with SO palsy.

References: None.

Keywords: Superior Oblique Palsy, OnabotulinumtoxinA, Inferior Oblique Muscle, Extraocular Muscle Surgery, Hypertropia

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 202

Ocular Alignment After Bilateral Medial Rectus Recession in Esotropic Patients with Spinocerebellar Atrophy

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Introduction:

To compare the surgical results of esotropic patients with spinocerebellar atrophy (SCA) and controls without neurologic disorders.

Methods:

Five patients with esotropia and SCA (group 1) and 10 age matched controls without neurologic disorder (group 2) who underwent bilateral medial rectus (BMR) recession were retrospectively included. All patients underwent BMR recession by one surgeon. Surgical success was defined as esotropia less than 6 PD and exotropia less than 9 PD. Preoperative and postoperative deviation angle measurement and success rate were analyzed.

Results:

Preoperatively, there was no statistical difference in mean horizontal deviation in primary position between the two groups (18 PD and 16.4 PD, respectively ($p>0.05$)). Mean esodeviation angles in primary position at postoperative 1 week measurements were 4.8 PD and 1.1 PD in group 1 and 2 respectively, and 6.4 PD and 3.2 PD in group 1 and 2, respectively, in postoperative 3 months. Success rate of group 1 was 40% and that of group 2 was 60% ($p<0.05$).

Conclusions:

: SCA patients with esotropia demonstrated slight undercorrection of their esodeviation when compared with that of controls after BMR recession. Our findings support the condition that surgeon should consider increasing the amount of BMR recession when planning the surgery for esotropic patients with SCA.

References: None.

Keywords: Spinocerebellar Atrophy, Esotropia, Bilateral Medial Rectus Recession, Postoperative Undercorrection, Success Rate

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 203

Capturing the Efferent Side of Vision in Multiple Sclerosis: New Data from a Digitized Rapid Number Naming Task

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Introduction:

Visual function in multiple sclerosis (MS) has been well characterized from an afferent standpoint using low-contrast acuity, optical coherence tomography (OCT) and quality of life (QOL). Compared to controls, patients with MS also demonstrate slowed reading times on the King-Devick (K-D) test, a rapid number naming task that captures widely distributed aspects of efferent function, particularly saccades. Slowed times in MS are associated with neurologic dysfunction and reduced vision-specific (QOL). However, the ocular motor underpinnings of such slowing have not been determined. We sought to determine ocular motor performance and characterize deficits leading to slowed KD reading times using recorded eye movements during a digitized King-Devick (K-D) test.

Methods:

We tested 13 patients with MS (mean age 37) and compared their results to those of our normative database of control participants without MS (n=38, mean age 31). Participants completed the digitized K-D task with simultaneous video-oculographic eye movement recording (EyeLink 1000+). Data were analyzed off-line in Matlab and Stata 14.0.

Results:

Digitized K-D completion times in the MS cohort were longer (worse) relative to controls (52.6 ± 14.1 sec vs. 43.7 ± 8.5 sec, $p=0.02$, t -test). Intersaccadic intervals (ISI), which represent a combination of saccadic latency and fixation duration between saccades, were prolonged in MS patients (397 ± 137 msec vs. 312 ± 53 msec, $p=0.02$, t -test). Within the MS cohort, test times were longer for the digitized vs. spiral-bound hand-held K-D test (52.0 ± 9.2 sec vs. 43.7 ± 9.7 sec, $p=0.01$, linear regression).

Conclusions:

In this ongoing study of ocular motor performance in MS, we have demonstrated that K-D reading times are slower secondary to prolonged ISI. The K-D test captures efferent visual dysfunction in MS, and is likely to be a sensitive performance-based outcome measure for future research, practice and clinical trials.

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Moster S, Wilson JA, Galetta SL, Balcer LJ. The King-Devick (K-D) test of rapid eye movements: a bedside correlate of disability and quality of life in MS. *J Neurol Sci*, 15, 1-2, 2014.

Keywords: Ocular motility, Multiple Sclerosis, King-Devick, Saccades

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 204**Exploring the Oculomotor Effects of Sustained-Release Fampridine in Multiple Sclerosis Patients with Gait Impairment.**

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Introduction:

Fampridine is a potassium channel blocker that has been shown to suppress downbeat nystagmus effectively. Its effect upon other eye movement abnormalities is largely unknown. We sought to evaluate the immediate and late effects of sustained-release fampridine 10 mg twice a day upon eye movement abnormalities in MS patients with gait impairment and to ascertain the utility of fampridine's oculomotor response in predicting gait response.

Methods:

A total of ten MS patients undergoing treatment with sustained-release fampridine to improve gait disability were submitted to binocular video-oculography and video-head impulse test (vHIT) prior to (visit 1), after 2 hours (visit 2), and after 2 weeks (visit 3) of fampridine treatment.

Results:

Among 10 enrollees, median age was 51 (41–63) years and 9 were females. The number of patients evidencing mild spontaneous nystagmus in darkness in the upright position markedly increased from visit 1 (2, 20%) to visits 2 (6, 60%) and 3 (4, 40%). Similarly, the mean number of patients showing positional nystagmus in darkness in 5 different positions was significantly higher at visit 2 (7.6) than visit 1 (5.1) ($p=0.02$). Six patients evidenced internuclear ophthalmoplegia (INO) (left, 6; right, 4) at visit 1. Importantly, the velocity versional dysconjugacy index (ratio of abducting to adducting eye mean velocity) significantly improved from visit 1 (1.62) to visit 2 (1.23) and 3 (1.23) ($p=0.03$), albeit only for left INO patients. In patients evidencing low horizontal canal (HC) gain (<0.8) at visit 1, vHIT responses significantly improved at visit 3 for the left HC ($p=0.04$) only. None of the ocular parameters and/or their change between visits was able to predict gait response to fampridine.

Conclusions:

Sustained-release fampridine seems to improve eye velocity dysconjugacy and high-frequency vestibular loss in MS patients. The development of spontaneous and positional nystagmus in darkness after treatment with fampridine deserves further investigation.

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Keywords: Aminopyridines, Downbeat Nystagmus, Multiple Sclerosis, Internuclear Ophthalmoplegia, Video-Head Impulse Test

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 205

Diagnostic Ability of the Three-step Test According to the Presence of the Trochlear Nerve in Superior Oblique Palsy

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Introduction:

To compare the diagnostic ability of the three-step test in unilateral superior oblique palsy (SOP) according to the presence of the trochlear nerve on high-resolution MRI.

Methods:

Diagnosis of unilateral SOP was made by the following 3 absolute criteria, (1) hyperdeviation in the primary position on alternating prism cover test, (2) unilateral underaction of the ipsilateral SO muscle with or without inferior oblique overaction, and (3) lack of evidence of other ocular motility disorders causing vertical deviation, plus 1 of 2 supportive criteria (4) a positive Bielschowsky head-tilt test, (5) unilateral atrophy of the SO muscle on MRI. Eighty-seven patients with a trochlear nerve (present group) and 79 patients without a trochlear nerve (absent group) were included. The sensitivities of each components of the three-step test were evaluated as well as factors related to sensitivities.

Results:

All three steps were positive in 78% of the present group and 72% of the absent group ($P = 0.471$). Step 1 plus 2 was positive in 83% of the present group versus 73% of the absent group ($P = 0.187$). Step 1 plus 3 was satisfied in 95% of the present group versus 99% of the absent group ($P = 0.370$). Patients who were positive in all three steps (complete group) had a larger amount of hyperdeviation during contralateral gaze ($P < 0.001$), ipsilateral tilt ($P = 0.038$) and adduction ($P < 0.001$) compared to those who were negative in at least one step (incomplete group). Superior rectus muscle contracture was more frequent in the incomplete group (24%) compared to the complete group (9%) ($P = 0.014$).

Conclusions:

The diagnostic ability of the three-step test in SOP was equivocal irrespective of the presence of the trochlear nerve. The sensitivity of the three-step test increased when only two steps were applied.

References: None.

Keywords: Superior Oblique Palsy, Three-Step Test, Bielschowsky Head-Tilt Test

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 206

A Detailed Analysis of Oculomotor Function in 16 Patients with Spinocerebellar Ataxia Type 3 (SCA 3).

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Introduction:

Oculomotor abnormalities in SCA 3 patients include the presence of square wave jerks (SWJ), saccade dysmetria, gaze-evoked nystagmus (GEN), positional nystagmus (pN), and caloric hyporreflexia. However, eye movement static and dynamic disconjugacy and high acceleration vestibular responses have been scarcely studied.

Methods:

Sixteen patients with genetically confirmed SCA 3 underwent neurological assessment applying the scale for the assessment and rating of ataxia (SARA), neuro-ophthalmological examination, binocular video-oculography and video-head impulse test (vHIT).

Results:

Patients' median age was 53.5 (26-77) years, median disease duration was 9 (2-25) years and 11 patients were female. The majority of patients (81%) showed esotropia at far. SWJ (mean number, 40/min) were present in all patients except one with concomitant horizontal saccade slowing. Dynamic overshoots within SWJ were asymmetric between eyes, giving the appearance of monocular disconjugate adducting "nystagmus" in two patients. Mean ocular pursuit gain (0.8) and saccade velocity (>300°/sec in 81% of patients) were relatively preserved. However, horizontal saccades were markedly disconjugate between eyes, being shorter and slower in the abducting eye (amplitude and velocity disconjugacy ratio, 0.95). Phase-plane analysis further showed that saccade velocity disconjugacy persisted during the entire eye excursion in 2/3 of patients. Spontaneous nystagmus in dark was an unusual finding (18%) while GEN and pN were both present in 75% of patients. vHIT evidenced right and/or left horizontal canal (HC) low gain (<0.8) in 75% of patients, which was negatively correlated with SARA score (right HC, $r=-0.7$, $p=0.001$; left HC, $r=-0.6$, $p=0.005$).

Conclusions:

A vergence abnormality possibly induced by cerebellar pathology is hypothesized to be an additional cause of disconjugate saccade velocity between eyes that remains throughout the whole eye displacement. Convergent strabismus may equally help to explain fixation instability disconjugacy in patients with SCA 3. Horizontal canal gain may be a potential biomarker of SCA 3.

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Keywords: Eye Movements, Strabismus, Spinocerebellar Ataxia Type 3, Video-Oculography, Video-Head Impulse Test

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 207**Measurement of Ocular Torsion with Digital Image Analysis in Face Turn and Head Tilt for Assessment of a Superior Oblique Palsy**

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Introduction:

The superior oblique intorts the globe, and its paresis results in pathologic excyclotorsion. Abnormal torsion can be clinically detected by fundus assessment. Typically fundus torsion is assessed in primary position. We hypothesized that additional assessment of fundus torsion in face turn and head tilt would improve diagnostic accuracy.

Methods:

Study participants underwent a full neuro-ophthalmic and sensorimotor examination. Fundus images of each eye were taken for primary position, 45 degrees face turn (right and left), and 15 degrees of head tilt (right and left). Torsion was assessed by an experienced strabismus specialist and by digital analysis software, measuring the angle between the fovea and the center of the optic nerve.

Results:

Fifteen normal subjects and twenty-five patients with clinical features of a unilateral superior oblique palsy were recruited. In primary position the mean torsion for normal subjects was 6.4 degrees of excyclotorsion (SD 3.6). Only 9 of the 21 superior oblique cases demonstrated abnormal torsion in primary position, and an additional 4 cases had abnormal torsion only in the contralateral eye. There was significant correlation with double-maddox readings (p 0.01) and high correlation with the subjective fundus assessment of the expert (p <0.0001). The degree of torsion in side gaze was not significantly different from primary position in either population. Relative to the sagittal axis of the skull, ipsilateral and contralateral head tilt produced a significant but slight change in torsion for both normal subjects and superior oblique palsy cases but there was no significant variation between the two populations.

Conclusions:

A significant amount of patients with clinical findings of a superior oblique palsy have normal fundus torsion. An experienced strabismus surgeon is able to accurately assess fundus torsion. Additionally evaluating torsion in side gaze or head tilt does not add diagnostic value.

References: None.

Keywords: Superior Oblique Palsy, Fundus Torsion, Double Maddox

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 208**Dental Malposition in Patients with Congenital Superior Oblique Muscle Palsy**

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¹Center of Dental Medicine, Dept. of Orthodontics, University of Zurich, Zurich, Switzerland, ²Dept. of Ophthalmology, University Hospital Zurich, Zurich, Switzerland, ³Dept. of Ophthalmology, Cantonal Hospital St. Gallen, St. Gallen, Switzerland

Introduction:

Patients with congenital superior oblique palsy tend to adopt a head tilt to the contralateral side to maintain binocular single vision. Facial asymmetries as a consequence of a long-standing head tilt have been reported previously. The aim of this study was to analyze the effect of habitual head tilt due to congenital superior oblique palsy on dental occlusion.

Methods:

The study was designed as a descriptive cohort study. Ten patients with congenital superior oblique palsy (3 female, 7 male; mean age 51.7 [y] \pm 15.8 SD, ranging from 19 to 69 [y]) underwent an orthodontic examination. Preoperative amounts of vertical, torsional and horizontal deviation were measured using Harms tangent screen and the results were correlated with orthodontic findings.

Results:

Three orthodontic parameters were found to correlate significantly or at least as trend with orthoptic parameters. Midline deviation of the upper jaw to the face (Mili UJ/F; $\rho=0.623$; $p=0.054$) as well as anterior positioning of upper first molar in the sagittal plane (6+6 ant; $\rho=0.594$; $p=0.07$) correlate with vertical deviation in primary position. Vertical distance between tips of upper and lower incisors, so called overbite, correlates with horizontal deviation measured in primary position ($\rho=0.768$; $p=0.016$).

Conclusions:

Based on our results we propose that in some patients with congenital superior oblique palsy the upper jaw rotates slightly towards the non-paretic side, inducing an anteriorization of the upper molar teeth on the paretic side. In our preliminary study three orthodontic parameters could be found to correlate with orthoptic measurements. Further studies should investigate whether in patients showing these orthodontic parameters a congenital superior oblique palsy can be found more frequently than in subjects without such characteristics.

References: None.

Keywords: Adult Strabismus, Ocular Motility, Head Tilt, Consequences Of, Dental Occlusion, Orthodontics

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 209

Rapid Number Naming and Quantitative Eye Movements May Reflect Contact Sport Exposure in a Collegiate Ice Hockey Cohort

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Introduction:

The King-Devick (K-D) test of rapid number naming is a reliable visual performance measure that is a sensitive sideline indicator of concussion when time scores worsen (lengthen) from pre-season baseline. We determined the relation of rapid number naming time scores on the K-D test to video-oculographic eye movement performance during pre-season baseline assessments in a collegiate ice hockey team cohort.

Methods:

Athletes from collegiate ice hockey team received pre-season baseline testing as part of an ongoing study of rapid sideline/ rinkside performance measures for concussion. These included the K-D test (spiral bound cards and tablet computer versions). Participants also performed a laboratory-based version of the K-D test with simultaneous infrared-based video-oculographic recordings using EyeLink 1000⁺. This allowed measurement of temporal and spatial characteristics of eye movements, including saccade velocity, duration and inter-saccadic intervals.

Results:

Among 13 male athletes, aged 18 to 23 years (mean 20.5±1.6 years), prolongation of the inter-saccadic interval (ISI, a combined measure of saccade latency and fixation duration) was the eye movement measure most associated with slower baseline KD scores (mean 38.2±6.2 seconds, $r=0.88$, $p=0.0001$). Older age was a predictor of longer (worse) K-D baseline time performance ($r=0.57-0.70$, $p=0.008-0.04$) as well as prolonged ISI ($r=0.62$, $p=0.02$) in this collegiate cohort. Slower baseline K-D times were not associated with greater numbers of reported prior concussions.

Conclusions:

Rapid number naming performance at pre-season baseline is best correlated with ISI when eye movements are recorded. Baseline K-D scores notably worsened with increasing age but not with number of prior concussions in this small cohort. These findings suggest that duration of contact sport exposure, rather than concussion history, may influence pre-season baseline rapid number naming performance.

References: None.

Keywords: Rapid Number Naming, Concussion, Eye Movements, Saccades, King-Devick Test

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 210**Adaptation to New Onset Incomitance**

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Introduction:

Incomitance is characterized by gaze dependent deviations of ocular alignment. The physiological mechanisms that maintain equal horizontal ocular alignment in all gaze directions (concomitance) in healthy individuals are poorly explored. We investigate adaptive processes in the vergence system that are induced by horizontal incomitant vergence stimuli (stimuli that require a gaze dependent vergence response in order to re-establish binocular single vision).

Methods:

We measured horizontal vergence responses elicited after healthy subjects shifted their gaze from a position that required no vergence to a field of view that required convergent eye movement.

Results:

Repetitive saccades into a field of view with a convergence stimulus rapidly decreased phoria (defined as the deviation of ocular alignment in absence of binocular stimulus). This change of phoria was present in all viewing directions, but was more pronounced in the gaze direction with a convergence stimulus. We also found that vergence velocity rapidly increased and vergence latency promptly decreased. We found gaze dependent modulation of phoria in combined saccade-vergence eye movements, and also in pursuit-vergence eye movements.

Conclusions:

Acute horizontal, gaze-dependent change of vergence, such as may be encountered in new onset parietic strabismus, will rapidly increase vergence velocity, and decrease latency. Gaze specific (concomitant) and gaze independent (incomitant) phoria levels will adapt and thus reduce induced phoria and promote concomitance.

References: None.

Keywords: Vergence, Saccade, Pursuit, Adaptation, Incomitance

Financial Disclosures: The authors had no disclosures.

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Poster 211

Decreased Binocular Summation in Strabismic Amblyopes and Effect of Strabismus Surgery

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Introduction:

Binocular summation (BiS) is the improvement in visual function using binocular vision surpassing the better eye alone. Previous studies have demonstrated diminished BiS in non-amblyopic strabismic patients. We examined effects of strabismic amblyopia and strabismus surgery on BiS.

Methods:

We prospectively enrolled 13 strabismic patients with amblyopia, 26 age-matched normal controls, and 26 strabismic patients without amblyopia. Strabismic patients were tested before and after realignment surgery. Visual acuity was tested binocularly and monocularly using high-contrast ETDRS charts and Sloan low-contrast charts at 2.5% and 1.25% contrast. BiS was calculated as the difference between the binocular and better eye scores.

Results:

Patients with strabismic amblyopia had subnormal BiS at both 2.5% (1.7 letters in strabismic amblyopes vs. 6.2 letters in normal controls, $p=0.001$) and 1.25% contrast (-0.46 vs. 5.9 letters, $p = 0.0075$). There was no difference in BiS between strabismic amblyopes and non-amblyopic strabismic patients. After strabismus surgery, BiS improved in amblyopic patients who had measurable stereoacuity preoperatively, from -2.6 to 6.0 letters on 2.5% contrast charts ($p = 0.03$), but did not improve after surgery in patients without stereo. Among patients with different strabismus subtypes, there was no significant difference in postoperative improvement in BiS.

Conclusions:

Both strabismic amblyopes and non-amblyopic strabismic patients have decreased BiS, but amblyopia may not further degrade BiS in the setting of strabismus. BiS may improve after strabismus surgery in amblyopic patients with measurable stereoacuity.

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Keywords: Strabismus, Amblyopia, Binocular Summation, Strabismus Surgery

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Poster 212**Stability of Human Binocular Alignment: A Video-Oculographic Luminance Study**

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Introduction:

To evaluate the stability of human binocular alignment under conditions of altered fixation and luminance.

Methods:

We measured horizontal binocular alignment in examined 8 healthy orthotropic subjects using infrared video-oculography (VOG) under conditions of binocular fixation and luminance change. Each testing condition was preceded by a binocular fixation period in room light (475 lux) to define the baseline binocular alignment. We then measured binocular alignment in darkness without fixation, in darkness with a distant fixational target, and in room light through a semi-translucent filter that precluded fixation. We determined whether these experimental conditions induced significant binocular alignment change from each baseline binocular alignment statistically using the signed rank test.

Results:

The mean horizontal binocular alignment in the dark was similar to baseline binocular alignment ($0.2^{\circ} \pm 2.8^{\circ}$; $p=0.4$). The mean horizontal binocular alignment when fixing in a room light was similar to baseline binocular alignment ($-1.4^{\circ} \pm 1.6^{\circ}$; $p=0.08$). The mean horizontal binocular alignment in the dark when a fixational target was provided showed an exo-drift compared to baseline alignment ($2.3^{\circ} \pm 1.0^{\circ}$; $p=0.0004$).

Conclusions:

The human brain does not require visual input to maintain binocular alignment on a short-term basis. The resilience of binocular alignment reflects the presence a subcortical memory system or tonus mechanism which is probably calibrated by phoria adaptation, a sensorimotor process that resets the baseline phoria toward zero to eliminate binocular disparity. Ocular proprioception may play a secondary role.

References: None.

Keywords: Ocular Motility, Higher Visual Cortical Functions, Higher Visual Functions

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 213**Pitfalls and Rewards for Implementing Emergent Bedside Oculomotor Testing in Patients with Acute Vestibular Syndrome**

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Introduction:

Patients with Acute Vestibular Syndrome (AVS) commonly presents to the Emergency Department and pose management challenges given concerns for posterior fossa strokes. A three-step bedside oculomotor examination called HINTS Plus (Head-Impulse, Nystagmus, Test-of-Skew Plus acute hearing loss) was shown to accurately identify small infarcts as a cause of AVS. We studied the feasibility of implementing HINTS testing in the ED, aiming to reduce unnecessary neuroimaging and improve posterior circulation stroke detection.

Methods:

A Quality Improvement project was launched in April 2015 after education about HINTS testing significance, performance and documentation. ED physicians were encouraged to obtain immediate neurological or stroke consultation in patients with acute vertigo. Brain imaging was not performed if HINTS suggested peripheral vestibular etiology. If central process was suspected, thin brainstem cuts MRI and vascular imaging were performed. Head CT was performed if presentation was within the time-window for thrombolysis. All patients were assessed for follow-up at 7 days.

Results:

Encountered and defeated pitfalls in the QI process were the performance of oculomotor examination, findings' documentation and interpretation and protocol applicability in patients with vertigo characteristics different than AVS. 37% of AVS patients had peripheral HINTS examinations and definitive peripheral vestibulopathies. 40% of patients had central HINTS exams with various underlying pathologies. tPA was administered in 2 cases, with excellent outcomes. 44% of patients underwent acute brain imaging. Direction-changing nystagmus and skew deviation were sensitive markers for central pathology. 57% of patients that became asymptomatic after vestibular suppressant's administration before HINTS testing or had episodic vertigo, demonstrated lack of corrective saccade.

Conclusions:

HINTS ED evaluation is feasible and valuable, by identifying patients in need of further CNS imaging or acute stroke therapy and safely avoiding neuroimaging for patients with peripheral HINTS exams. The impact on the cost of care and repeat consultation rates is pending determination.

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Keywords: Vertigo, Nystagmus, Skew Deviation, Head Thrust, Stroke

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 214
Head Tilt Observation Study

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Introduction:

Vertical ocular misalignments can produce significant symptoms even when the degree of strabismus is small. Those can be difficult to detect and are frequently missed. We observed that most patients who have vertical misalignment often have a resting head tilt. Thus, we performed a study to assess the sensitivity and specificity of the presence of a resting head tilt correlating to a vertical strabismus.

Methods:

Patients were included if they presented to our Neuro-ophthalmology clinic and were observed to have a resting head tilt. In such cases, data was recorded regarding a vertical/oblique diplopia, an awareness of the head tilt, history of head trauma, whether the referring provider noted the head tilt, and the results of a sensorimotor exam including a Maddox rod evaluation. Thirty-five patients were identified with a head tilt.

Results:

Our preliminary results showed that 94% of those patients (33/35) had a vertical ocular misalignment, 76% (25/33) of whom described diplopia. The misalignment was detected by alternate cover test in 25% of cases. When this test was negative, Maddox rod identified 75% of the remainder. Of those, 18% were aware of the head tilt, which was recognized by 17% of the referring healthcare providers.

Conclusions:

This preliminary study revealed a very high sensitivity and specificity for the predictive value of a resting head tilt to be associated with a vertical ocular misalignment. An important outcome was the value of using a Maddox rod test. Its use, however, induces phorias, and the next phase of our study will include an assessment of a false positive yield with the Maddox rod test (i.e. how often is it positive in patients without a head tilt). Attention to the presence of a resting head tilt should prompt clinicians to search for even subtle vertical misalignment, as correction with prism often provides symptomatic improvement.

References:

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2. Muthusamy B, Irsch K, Peggy Chang HY, Guyton DL. The sensitivity of the bielschowsky head-tilt test in diagnosing acquired bilateral superior oblique paresis. *Am J Ophthalmol.* 2014 Apr;157(4):901-907

Keywords: Head Tilt, Ocular Vertical Misalignment

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 215**Factors Influencing Success of Prisms In The Management of Diplopia From Various Causes**

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Introduction:

Prospective studies evaluating prism satisfaction amongst patients with various kinds of ocular deviations are lacking. Our objective was to evaluate systematically various factors such as type and amount of ocular deviation, fusional ability, amount of prism given, type of glasses, that can influence the success of prisms in patients with diplopia. By means of a questionnaire patients were queried about their overall satisfaction with prism use as well as the side effects they experienced.

Methods:

Prospective, consecutive, single center study of adult patients with binocular diplopia. Details of ocular misalignment and prisms prescribed were collected. A survey questionnaire was administered specifically inquiring into advantages and disadvantages of prism glasses.

Results:

Of the 110 patients enrolled in the study, there were 69 females and 41 males with a mean follow up of 7 months. Prisms were prescribed for esodeviation (41), exodeviation (12), hyperdeviation (35) and combined deviations (22) from various etiologies. The mean horizontal alignment was 7.6PD and mean vertical alignment was 6.3 PD. The average horizontal prism prescribed was 6.4PD and mean vertical prism prescribed was 5PD. The overall satisfaction with prism was 88 % with improvement noted in diplopia (90%), enhanced depth perception (44%), improved driving (57%) and reading (51%). The side effects of prisms reported that were bothersome to the patients were increased weight (5%), altered depth perception (16%), distortion of vision (10%) and seeing halos (7%).

Conclusions:

Overall the satisfaction of prisms in our prospective cohort of patients was high across all ocular misalignments and for wide range of deviations. Improvements were reported by patients with regards to driving, reading and other day today tasks. Noticeable side effects in this cohort included altered depth perception and visual distortion.

References: None.

Keywords: Diplopia, Prisms, Ocular Motility

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 216

Screening for Thymoma in Suspected Ocular Myasthenia Gravis: A Retrospective Study

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Introduction:

Thymoma can be associated with ocular myasthenia gravis (OMG). The yield of thymic imaging to screen for thymoma in suspected OMG patients is not known.

Methods:

A retrospective study of consecutive patients with suspected OMG seen in a neuro-ophthalmology clinic from Jan 2013-Sept 2015

Results:

42 patients (57% men) with a median age of 50 years (range 20-86) were included. 43% were seropositive for anti acetylcholine receptor (anti-AChR), one seropositive for muscle specific kinase antibody. 36 patients had thymic imaging by magnetic resonance (MR) or computed tomography (CT). 2/36 (6%) had thymoma confirmed on imaging and histology. Both were seropositive to anti-AChR. Anti-striated muscle antibody, tested in one, was negative.

Conclusions:

Thymic imaging to exclude thymoma is recommended in all patients with ocular myasthenia gravis.

References: None.

Keywords: Ocular Myasthenia Gravis, Thymoma

Financial Disclosures: The author had no disclosures.

Grant Support: None.

Poster 217

The Ocular Motor Underpinnings of Rapid Number-Naming as a Sideline Performance Measure for Concussion

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Introduction:

The King-Devick (KD) test of rapid number-naming, a sensitive and rapid performance measure, adds a visual and eye movement outcome to sideline testing for concussion. We performed a laboratory-based study to characterize ocular motor behavior during the KD test in a cohort of patients with concussion to identify the features leading to KD reading time prolongation in concussion.

Methods:

We tested 12 subjects with a history of concussion (mean age 31) and compared their results to those of our normative database of non-concussed control participants (n=38, mean age 31). Participants performed a computerized version of the KD rapid number-naming task. Simultaneous infrared-based video-oculographic recordings were performed using Eyelink 1000⁺, allowing measurement of temporal and spatial characteristics of eye movements (saccade velocity and duration, inter-saccadic interval, etc.).

Results:

Average inter-saccadic intervals (a combined measure of saccade latency and fixation duration) were significantly greater among concussed subjects compared to non-concussed controls (394.43 ±140.64 msec vs. 312.45 ±52.54 msec, $p=0.036$, t -test). Digitized KD reading times were prolonged in concussed participants versus non-concussed controls (52.43 ±14.20 sec vs. 43.68 ±8.54 sec, $p=0.032$, t -test). In addition to temporal differences, concussion was also associated with larger average deviations of horizontal saccade endpoints from the centers of to-be-read numbers (0.37 ±0.35 deg vs. 0.13 ±0.31 deg, $p=0.02$, t -test). There were no differences in saccade velocity, duration, amplitude, or in vertical deviation from number centers.

Conclusions:

Results of this study demonstrate that prolonged intersaccadic intervals and larger deviations of saccade endpoints likely underlie the increased reading times for the KD test in the setting of concussion. A sensitive and rapid sideline performance measure, the KD test of rapid number-naming relies upon a diffuse network responsible for saccade target selection and planning and captures deficits in efferent visual function characteristic of concussion.

References: None.

Keywords: Trauma, Ocular Motility

Financial Disclosures: The authors had no disclosures.

Grant Support: JRR supported by NICHD 5K12 HD001097 (RMSTP program).

Poster 218**Clinical Manifestations of Extraocular Muscle Paresis in Patients with Miller Fisher Syndrome**

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Introduction:

To determine the clinical manifestations of extraocular muscles (EOM) paresis in patients with Miller Fisher syndrome.

Methods:

We retrospectively reviewed the medical records of patients with Miller Fisher syndrome who have been presented with ophthalmoplegia between 2010 and 2015. Antecedent infections, involved EOMs, strabismus pattern in primary position, involved cranial nerves, and laboratory findings were analyzed. We compared patients with complete ophthalmoplegia (CO) and patients with incomplete ophthalmoplegia (IO).

Results:

A total of 28 patients, including 15 males and 13 females, were included in the study. The mean age was 39.3 ± 19.95 years. Twenty-five had symptoms of antecedent infection. The mean time of complete recovery was 13.1 ± 7.60 weeks. Twenty-three had positivity to the serum anti-GQ1b antibody. The most involved EOM and the latest recovered EOM were lateral rectus muscle. CO was observed in 10 patients and IO was observed in 18 patients. The mean age was 59.8 ± 16.98 years in CO, and 27.9 ± 10.52 years in IO. The proportion of female was 80.0% in CO, and 27.8% in IO. The occurrence of facial palsy was 70.0% in CO, and 22.2% in IO. Patients with an elevated CSF protein were 55.6% in CO, and 17.6 % in IO (the mean checked time from initial symptoms; 3.7 days).

Conclusions:

Lateral rectus muscle was the most involved and the latest recovered EOM. Patients with complete ophthalmoplegia were much older, higher ratio of female, higher incidence of facial palsy, and more elevated CSF protein than patients with incomplete ophthalmoplegia.

References: None.

Keywords: Miller Fisher Syndrome, Extraocular Muscle Paresis, Guillain–Barré Syndrome, Complete Ophthalmoplegia, Anti-GQ1b Antibody

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 219**Differentiating Decompensated Superior Oblique Muscle Dysfunction from Ischemic Fourth Cranial Nerve Palsies**

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Introduction:

Isolated fourth cranial nerve palsies are commonly caused by decompensation of a dysfunctional superior oblique muscle ("decompensated palsies"). The differentiation of this cause from palsy caused by presumed microvascular ischemia ("ischemic palsy") at the time of initial diagnosis has value. The diagnosis a decompensated palsy is traditionally suggested by finding abnormally high vertical fusional vergences. This study was undertaken to discover if abnormally high fusional vergences are reliable in this task and whether there are other clinical characteristics that could fortify the differentiation between these two causes.

Methods:

Retrospective review of case records of adult patients diagnosed with isolated fourth cranial nerve palsies examined and followed at our clinics over the past 15 years. The gold standard of diagnosis for an ischemic palsy was full recovery within 6 months of onset. The gold standard for diagnosis of a decompensated palsy was persistence of the palsy beyond that time period and reasonable exclusion of alternative diagnoses.

Results:

Vertical fusional vergences were not always definitely increased in decompensated palsies. Nor was the presence of torsional misalignment on double Maddox rod testing. However, vertical misalignment greater in contralateral upgaze than downgaze was more common in decompensated palsies and served as a powerful discriminator.

Conclusions:

Vertical misalignment greater in contralateral upgaze than downgaze is characteristic of decompensated fourth cranial nerve palsy and is useful in differentiating this cause from an ischemic palsy. Adding this sign to increased vertical fusional vergences improves differential diagnosis.

References: None.

Keywords: 4th Cranial Nerve, Palsy, Ischemic, Decompensating

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 220**Retinal Thinning and Neurodegeneration in Xeroderma Pigmentosum: Evidence from Optical Coherence Tomography**

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Introduction:

Xeroderma pigmentosum (XP) is a rare autosomal recessive disease with cutaneous, ophthalmic and neurological manifestations. Although the etiology for XP neurodegeneration is poorly understood, it is known that complementation groups C, E and XP variants (V) are spared the severe, progressive neurological deficits that occur in more than 25% of patients from groups A, B, D, F and G. The aim of this study was to explore the association between retinal thickness measurements and neurodegeneration in patients with XP from different complementation groups.

Methods:

This was a cross-sectional study. Forty patients with XP and normal retinal examinations were divided into two groups based on their complementation group and associated likelihood of developing neurodegeneration. Group 1 contained 23 patients, with no or minimal neurological deficits, from complementation groups A, B, D, F and G. Group 2 contained 17 neurologically intact patients, from complementation groups C, E, and the variant type. A third control group was obtained from a normative database. Using spectral-domain optical coherence tomography (SD-OCT), we compared peripapillary retinal nerve fiber layer (pRNFL) and macular thickness between the groups.

Results:

We found a statistically significant reduction in both pRNFL ($p < 0.01$) and macular thickness ($p < 0.001$) for Group 1 compared with healthy controls. In contrast, there was no statistically significant difference between pRNFL and macular thickness in patients from Group 2 compared to the control group. When Group 1 and Group 2 were compared, a statistically significant reduction in total pRNFL ($p = 0.024$) and macular thickness ($p = 0.002$) was found in Group 1.

Conclusions:

Our results suggest that retinal SD-OCT scans provide a quantifiable method for assessing early neurological abnormalities in patients with XP. These findings demonstrate the potential role of retinal thickness as an anatomic biomarker and diagnostic indicator for XP neurodegeneration.

References: None.

Keywords: Xeroderma Pigmentosum, Neurodegeneration, Optical Coherence Tomography, Retinal Nerve Fiber Layer, Macular Thickness

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 221

FOTO-ED Phase III: What Happens When You Try to Teach Emergency Providers to Read Fundus Photographs?

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Introduction:

During FOTO-ED Phase II (n=355), emergency providers (EPs) reviewed 68% of patients' non-mydriatic fundus photographs, identifying 46% of the relevant abnormalities during their routine evaluations without additional ophthalmology training.¹ We undertook a quality improvement project to determine whether a web-based educational module would improve how often and how well they reviewed fundus photographs.

Methods:

Patients presenting to our Emergency Department (ED) from March-December 2014 with headache, focal neurologic defect, visual change, or diastolic blood pressure ≥ 120 had non-mydriatic fundus photography. Photographs were provided through the electronic medical record. In May 2014, a web-based educational module was released (9 training pages + pre/posttest [35 questions each]). Core EPs who worked frequently in the ED were required to complete the module by Oct 2014.

Results:

587 patients were included, 74 with abnormalities (12.6%): 24 disc pallor, 20 disc edema, 16 isolated retinal hemorrhage, 9 grade III/IV hypertensive retinopathy, and 5 retinal vascular occlusion. 61% (359/587) were evaluated by the 14/16 (88%) core EPs who completed training. EPs spent a median of 31 minutes (IQR:24-42) on training; post-test scores improved by a median of 4% (IQR:2-14%;p=0.06). Pre- vs. post-training, they reviewed 45% (80/177) vs. 43% (78/182;p=0.73) of photographs, correctly identified abnormal images in 67% (10/15) vs. 57% of cases (8/14;p=0.88), and correctly identified as normal 80% (52/65) vs. 84% (54/64;p=0.67). Untrained EPs reviewed 35% (79/228), correctly identified 50% of abnormal (6/12), and 79% of normals (53/67). Only their review frequency significantly differed from those who completed training (p=0.03).

Conclusions:

FOTO-ED demonstrated that EPs perform substantially better with fundus photography than with direct ophthalmoscopy. However, our web-based, in-service training did not result in further improvements at our institution. Developing new tools to provide relevance to fundus findings in patient management scenarios and to better integrate them into EP clinical decision-making may prove more valuable.

References:

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Keywords: Optic Neuropathy, High Intracranial Pressure/Headache, Retina, Stroke

Financial Disclosures: The authors had no disclosures.

Grant Support: Research to Prevent Blindness, K23-EY019341, R01-NS089694, P30-EY006360.

Poster 222**Visual Structure Biomarkers of Traumatic Brain Injury (TBI) in Contact Sport Athletes using Optical Coherence Tomography (OCT)**

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Introduction:

It is estimated sports-related concussion afflicts 3.8 million American athletes each year. Recent research suggests that repeated head trauma may be associated with the development of neurodegenerative changes in the brain. These changes in the brain can begin months, years, or even decades after the last brain trauma and are not visible on standard brain scans. As the eye is an extension of the brain, this study investigates the retinal structure in active and retired professional contact sport athletes with a high risk of concussion (football, boxing, hockey, soccer and rugby) compared to age-matched controls to determine if visual structure measures may be an in vivo clinical biomarker to detect nerve damage related to repeated head trauma.

Methods:

Patients with history of elite-level, high TBI risk contact sport exposure (Boxing, n=13), moderate TBI risk contact sport exposure (Football, n=20) and elite-level, non-contact sport athletes or non-athlete controls (n=25) underwent Spectral-Domain OCT optic nerve and macula scans.

Results:

Boxing athletes demonstrated significant thinning in average Retinal Nerve Fiber Layer (RNFL) compared to Controls (Boxers: 84.31±9.81µm vs. Controls: 94.72±11.10, p = 0.01, Figure 1) and Football athletes (Boxers: 84.31±9.81µm vs. Football: 93.70±9.03, p = 0.04). Average Ganglion Cell Complex (GCC) thickness was thinner in Boxing athletes compared to Football athletes (Boxers: 76.85±7.85µm vs. Football: 80.10±8.16 µm), and compared to Controls (Boxers: 76.85±7.85µm vs. Controls: 82.16±5.86 µm) however these differences were not statistically significant. No significant differences in average macular thickness or volume were observed between groups.

Conclusions:

Retinal structure thinning quantified using non-invasive OCT imaging is evident in high TBI risk contact sport athletes and may serve as an important biomarker of sport-related TBI exposure. Continued investigations will examine correlations with visual function and determine changes over time.

References: None.

Keywords: Trauma, Neuroimaging, Retina, Diagnostic Tests

Financial Disclosures: The authors had no disclosures.

Grant Support: Study supported by the Illinois Society for the Prevention of Blindness Research Grant.

Poster 223

Optic Neuritis History Does Not Influence OCT Measurement in Long-Standing Secondary Progressive Multiple Sclerosis

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Introduction:

Optical coherence tomography (OCT) is a useful outcome measure of neuronal integrity in relapsing-remitting MS (RRMS) trials.^{1,2} Its role in secondary progressive MS (SPMS) is promising but less well-understood.³⁻⁵ Retinal nerve fiber layer (RNFL) and macular thinning is generally attributed to prior optic neuritis (ON);⁴ however, OCT thinning in the absence of ON has also been demonstrated.^{6,7} This study addresses the influence of ON history on OCT readings in SPMS.

Methods:

Fifty participants in a prospective interventional clinical trial underwent baseline spectral domain OCT in each eye after pharmacologic dilation. OCT was repeated annually over a 2-year period and reviewed by an experienced OCT reader. Self-reported histories of ON were confirmed as available via medical records. Mean and quadrant RNFL thicknesses, macular volume, and mean ganglion cell layer plus inner plexiform layer (GCIPL) thicknesses were compared between patients with and without a history of ON. Two-year data was compared to baseline using paired t-tests, Tukey pairwise comparisons and multiple comparison modeling where necessary.

Results:

Scans of seventy-eight eyes from 42 subjects passed quality control. Mean (standard deviation) age was 59 (6.4) years and 60 percent of subjects were female. Mean duration of MS was 30 (9.6) years and of SPMS was 10 (5.6) years. Mean expanded disability status score was 5.4 (1.5). No significant differences between mean or quadrant RNFL thicknesses or macular volumes were observed based on ON history at baseline (corrected $p < 0.05$). A non-significant trend towards effect of ON on GCIPL thickness was initially found ($p = 0.08$). History of ON was not associated with change in any OCT measure from baseline to 2 years.

Conclusions:

History of ON did not influence RNFL or GCIPL thicknesses in an advanced SPMS cohort over 2 years, suggesting that ON is not a true confounder when using OCT outcome measures for neuroprotective trials in this population.

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Keywords: Diagnostic Tests (OCT), Secondary Progressive Multiple Sclerosis, Demyelinating Disease, Neuro-Ophth and Systemic Disease (MS)

Financial Disclosures: The authors had no disclosures.

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Poster 224

Peripapillary Subretinal Hemorrhage: Clinical and Ocular Coherence Tomography Findings in 5 Cases

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Introduction:

Peripapillary subretinal hemorrhage (PSH) has been described as a benign condition occurring in patients with myopia and tilted, dysplastic discs.¹⁻² Recognizing these hemorrhages is important as they are not always associated with drusen, ischemic optic neuropathy (ION), or peripapillary subretinal neovascular membranes, and do not require further work-up as they resolve spontaneously with no visual deficit.¹ Although their etiology is unknown, it has been shown that patients with PSH have smaller optic discs and more nerve fiber crowding than age-matched controls.² Here, we examine five cases of PSH studied with ocular coherence tomography (OCT) to better understand the anatomy of the optic disc in relation to the hemorrhage.

Methods:

A patient database extending from 1981 until August, 2015 was searched for diagnoses of “hemorrhage” and “tilted” or “hypoplastic” disc. Inclusion criteria were ophthalmoscopic evidence of PSH. Exclusion criteria included any findings other than PSH. Case histories, ocular examination, OCT, and ancillary testing were reviewed. Time of follow-up was recorded.

Results:

Five patients (six eyes) were identified with PSH. All had isolated peripapillary hemorrhages, one of which was temporal, and the rest nasal and superonasal. On OCT, the optic disc tissue extended over Bruch’s membrane into the photoreceptor layers, creating a ridge adjacent to the disc and elevating retinal layers from the RPE. There was no evidence of vitreopapillary traction causing the ridge. Upon comparing OCT with fundus photos, the hemorrhage was consistently found adjacent to the optic disc ridge.

Conclusions:

Previous papers describe PSH occurring only nasally, whereas we have found one patient with a temporal PSH and a temporally tilted nerve. We hypothesize that PSH occurs where optic disc tissue extends into the photoreceptor layers, as seen on OCT, placing stress on the border tissue of Elschnig and basement membrane causing blood to escape from adjacent choriocapillaris.

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Keywords: OCT, Peripapillary, Subretinal, Hemorrhage, Retina

Financial Disclosures: The authors had no disclosures.

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Poster 225**The Utility of a Hand-Held, Non-Mydriatic Fundus Camera for the Detection of Optic Nerve Edema**

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Introduction:

To determine the sensitivity of pictures obtained with a portable, hand-held non-mydriatic fundus camera (Pictor Plus, Volk Optical, Inc.) as an alternative to the dilated fundus examination for the detection of optic nerve edema.

Methods:

Retrospective review of non-mydriatic, fundus photographs of patients age 18 and older with normal and/or edematous optic nerves presenting to Kellogg Eye Center neuro-ophthalmology clinics. The gold standard was the treating neuro-ophthalmologist's documented optic nerve examination (dilated or undilated). The photograph reader was a fellowship trained neuro-ophthalmologist who did not examine the patient. The presence of edema was recorded as either present, absent, or uncertain. Responses of "uncertain" were grouped with "present." The mean time to evaluate the photo, overall photo quality (excellent, acceptable, ungradable), and quality of the optic nerve specifically (excellent, acceptable, ungradable) was recorded.

Results:

101 eyes were included; 52 with optic nerve edema and 49 without. None of the photographs were ungradable and it took a mean of 18.9 seconds to read each photo. Photographs interpreted by a neuro-ophthalmologist correctly identified the presence or absence of optic disc edema in 88.1% of eyes. Twelve patients were misclassified (6 false positives, 6 false negatives). In 4 of 6 false positives, the reader was uncertain about the presence of optic nerve edema.

Conclusions:

Photographs taken with a hand-held, non-mydriatic camera and interpreted by a neuro-ophthalmologist correctly identified the presence or absence of optic disc edema in 88.1% of eyes suggesting that this may be an acceptable tool for the detection and/or monitoring of optic nerve edema.

Keywords: Diagnostic Tests

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 226**Morphometric Evaluation of Asymptomatic Optic Disc Elevation and Grade I Papilledema**

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Introduction:

To demonstrate morphometric characteristics of elevation of optic disc with disc photos, orbital B scan and SD-OCT (optical coherent tomography) in asymptomatic patients and to compare them to grade I papilledema

Methods:

Among the patients who were referred to Neuro-Ophthalmology clinic regarding elevation of optic disc, asymptomatic patients were selected for group 1 and the symptomatic patients with grade I papilledema were selected for group 2. All patients underwent Humphrey visual field test, ONH (optic nerve head) OCT, orbital B scan and fundus photos and the patients in group 2 underwent lumbar puncture which showed elevated opening pressure. The data were reviewed and the average thickness of retinal nerve fiber layer (RNFL) in each quadrants on OCT was calculated. The sum of temporal and nasal quadrants was compared to inferior quadrants in each group and they were compared to the Cirrus OCT normative data.

Results:

The data were obtained from 48 cases in 24 patients including 12 asymptomatic patients (group 1) and 12 symptomatic patients of grade I papilledema (group 2). In group 1, the tests suggested optic disc drusen in 16 cases and crowded optic disc in 8 cases. The comparison of thickness of RNFL to the normative data showed no statistically significant difference in group 1 and showed statistically significant difference in group 2. (Further collection of data and final analysis of statistics are pending at the time of submission.)

Conclusions:

The comparison of the sum of RNFL in the temporal and nasal quadrants to RNFL in the inferior quadrant in OCT may be used as an adjunct method for differentiation of pseudopapilledema from grade I papilledema.

References: None.

Keywords: Diagnostic Tests, Pseudotumor Cerebri

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 227**Optical Coherence Tomography Angiography and Anterior Ischemic Optic Neuropathy**

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Introduction:

OCT angiography (OCT-A) is a new technology allowing the imaging of retinal microvascular flow without the injection of an intravenous dye. It also allows the visualization of the peripapillary vascularization. Even if the most significant contribution to the vascularization of the optic nerve comes from the choroidal blood flow, the peripapillary capillaries also contribute to the vascularization of the prelaminar portion of the optic nerve. The goal of this study is to describe the peripapillary microvasculature in nonarteritic anterior ischemic optic neuropathy (NAION) with optical coherence tomography angiography (OCT-A).

Methods:

Observational study of 6 patients at the acute phase of NAION. OCT-A was performed using a 3mm x 3mm square centered on the optic disc. A qualitative comparison was made with healthy fellow eye of each patient.

Results:

In the affected eyes, OCT-A imaging demonstrated clear modifications in the radial peripapillary network. In all these eyes, a focal disappearance of the superficial capillary radial pattern was at least demonstrated and found to be twisted and irregular. In two eyes, the alteration was more severe, with a lack of vascularization in some focal areas, appearing as dark zones.

Conclusions:

OCT-A is a safe and quick imaging tool able to demonstrate a reshaping of the peripapillary capillary network likely related to a decrease of the blood flow during the acute phase of NAION. Besides qualitative evaluation, future software upgrades will allow the quantification of the peripapillary vascular density and its blood flow.

References: None.

Keywords: Diagnostic Test, Optic Neuropathy

Financial Disclosures: Jean-F Korobelnik, Zeiss consultant

Grant Support: None.

Poster 228

Macular Thickness Measurements with Frequency Domain-OCT for Quantification of Retinal Neural Loss and its Correlation with Cognitive Impairment in Alzheimer's Disease

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Introduction:

Alzheimer's disease (AD), the most common cause of degenerative dementia, is characterized by progressive cognitive deficits, including memory disturbances, aphasia, apraxia, agnosia and visual abnormalities. The purpose of this study is to evaluate the ability of frequency domain OCT (fd-OCT) to estimate neural loss in eyes with AD. We also verified the existence of correlation between AD-related cognitive impairment and macular and peripapillary retinal nerve fiber layer (RNFL) thickness measurements.

Methods:

Macular and peripapillary fd-OCT scans were obtained from 45 eyes of 24 patients with AD and 48 control eyes. Peripapillary RNFL, macular full-thickness and inner macular thickness parameters were automatically calculated by the equipment's software. The inner macular parameters included macular retinal nerve fiber layer (mRNFL) thickness, ganglion cell layer (GCL) plus inner plexiform layer (IPL) thickness (GCL+), and RNFL plus GCL+ thickness (GCL++). The Mini-Mental State Examination (MMSE) was used to assess cognition in AD and control subjects. The two groups were compared and the existence of a correlation between MMSE scores and fd-OCT measurements was verified.

Results:

The only significantly reduced peripapillary RNFL parameters were average thickness and inferior quadrant thickness. All full-thickness macular parameters were significantly smaller in AD eyes than in controls, except for inferior outer macular segment thickness. GCL+ and GCL++ were significantly reduced in AD eyes. A significant correlation was found between most fd-OCT parameters (especially macular thickness measurements) and MMSE scores.

Conclusions:

Most fd-OCT parameters, especially those in the central macular area, were reduced in AD eyes. For the first time, the inner retinal layers of the macular area were shown to be reduced in AD as well. Moreover, neuronal loss, especially as reflected in macular parameters, correlated well with cognitive impairment in AD. Our results suggest that fd-OCT could be used as a swift and non-invasive diagnostic tool in the routine evaluation and follow-up of AD patients.

References: None.

Keywords: Alzheimer, Optical Coherence Tomography, Retinal Nerve Fiber, Macular Thickness Measurements

Financial Disclosures: The authors had no disclosures.

Grant Support: Supported by Conselho Nacional de Desenvolvimento Científico e Tecnológico (CNPq), Brasilia, Brazil (programas institucionais de bolsas de iniciação científica, XXVII BIC/UFJF - 2014/2015 and grant 307393/2014-3).

Poster 229**Optical Coherence Tomography Macular Volume Correlates with Walking Speed in Patients with Multiple Sclerosis**

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Introduction:

Optical coherence tomography (OCT) is used to measure the thickness of the layers of the retina. In multiple sclerosis (MS), it is used to quantify the effects of optic neuritis or MS, monitor disease progression, and assess for complications of disease modifying therapy. OCT is being evaluated as a potential biomarker in MS, for use in clinics and in therapeutic trials. The aim of this study was to evaluate the relationship between retinal thickness with walking speed in patients with MS/clinically isolated syndrome (CIS).

Methods:

This was a retrospective, observational study of patients diagnosed with MS or CIS. A clinic database and electronic medical record were used to identify patients with an OCT, timed 25 foot walk, and visual acuity (VA) performed within 3 months. The Spearman correlation coefficient was computed for pairs of variables, including OCT (retinal nerve fiber layer thickness, ganglion cell and inner plexiform layer thickness, and macular volume), VA, and timed 25 foot walk. Since history of optic neuritis was not available, the best eye and worst eye visual acuities were used. The OCT values were specific to the best and worst eye (referred to as correlated). A univariate analysis identified statistically significant variables. A multivariate analysis and stepwise selection were then performed.

Results:

370 patients met inclusion criteria. Univariate analysis identified a statistically significant correlation between 25 foot walk and all variables, except worst eye ganglion cell layer thickness. Correlation between best eye macular volume and walking speed was strongest ($r=0.890$, $P<0.001$). For each cubic millimeter increase in macular volume, walking speed increased by 0.876 feet/sec.

Conclusions:

OCT, a proposed biomarker for neurodegeneration, correlates with walking speed in patients with MS or CIS. Results indicate that degeneration of the retina (particularly macular volume) in MS/CIS may be a marker of global neurodegeneration outside the visual system.

References: None.

Keywords: OCT, Walking Speed, Demyelinating Disease

Financial Disclosures: Robert Bermel has served as a consultant for Biogen, Novartis, Genzyme, Genentech. Other authors have no disclosures.

Grant Support: None.

Poster 230**Effect of Upper Eyelid Surgery on Headaches**

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Introduction:

This study was undertaken to prospectively evaluate changes in headache related quality of life in patients with chronic headache and droopy eyelids, prior to and following blepharoplasty, ptosis repair, or brow ptosis repair.

Methods:

Prospective interventional study of adult patients undergoing upper eyelid or brow surgery for obscuration of superior visual field (study arm) and other oculoplastic procedures (control arm) with greater than one year of intermittent headache symptoms over an 18 month period. A validated headache quality of life survey, Headache Impact Test – 6, was administered to patients preoperatively and at a post-op visit (minimum of 7 weeks). Patients were excluded if they had known orbital, corneal, or intracranial pathology, received botulinum toxin, or had a change in headache medications.

Results:

Twenty eight patients (22 female, 6 male) met inclusion criteria for the study arm, and 19 patients (16 female, 3 male) for the control arm. Mean age was 58.7 for the study arm, and 60.7 for the control arm. There was no statistically significant difference in any of the headache locations. Mean preoperative HIT-6 score was 57.7 for the study arm and 58.1 for the control arm ($p=0.86$). Mean postoperative HIT-6 score was significantly better at 45.3 for the study arm and unchanged at 58.6 for the control arm ($p<0.05$). Mean follow up was 163 days and 157 days respectively. Subjectively, 18/28 in the study arm reported their headache symptoms were significantly better or resolved, as compared to 0/19 in the control group.

Conclusions:

Patients presenting to an oculoplastic clinic for management of visually significant upper eyelid position are more likely than patients undergoing other oculoplastic procedures to have improvement in headache related quality of life, and improvement or resolution of headache symptoms.

References: None.

Keywords: Eyelid Surgery, Headache, Quality of Life

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 231

Irrigation and Ultrasonic Drill-induced Heat Transmission.

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Introduction:

A variety of instruments are used to remove bone in orbital decompression surgery; the Sonopet™ ultrasonic bone curette has recently been described in this capacity (Sivak-Callcott 2005, Cho 2010, Vrcek 2015). Thermal injury to adjacent soft tissue (e.g. the optic nerve) remains a concern with this and other drills. Irrigation of the surgical field is used to mitigate this temperature rise. This study evaluates the influence of saline irrigation flow rate and temperature on Sonopet™ ultrasonic drill-induced heat transmission through bone.

Methods:

The Sonopet™ ultrasonic drill was used to burr unpreserved cadaveric porcine rib bone. Temperature was measured with a Thermoworks Super-Fast Thermapen™ probe. Standard recommended settings (saline at 21°C and 18ml/min) were compared to high flow (saline at 21°C and 40ml/min) and chilled saline with high flow (saline at 10°C and 40ml/min). The temperature of the emitted irrigation fluid itself was also measured under these same conditions while oscillating the tip without making contact with the bone.

Results:

The temperature of the emitted irrigation fluid after 15 seconds while allowing the Sonopet™ tip to oscillate without drilling bone was slightly lower with chilled saline with high flow rate compared to standard settings ($25.4 \pm 0.3^\circ\text{C}$ vs $24.6 \pm 0.3^\circ\text{C}$, t-test $p=0.047$). There was no significant difference between the change in bone temperature between the standard and high flow settings ($9.4 \pm 3.1^\circ\text{C}$ vs $8.4 \pm 1.9^\circ\text{C}$, t-test $p = 0.39$). A significant decrease in the change in bone temperature at 15 seconds was noted when comparing chilled high flow saline to standard settings (mean change of $9.4 \pm 3.1^\circ\text{C}$ with standard settings vs. $6.3 \pm 1.1^\circ\text{C}$ with chilled high flow saline, t-test $p = 0.003$).

Conclusions:

Varying irrigation fluid flow rate and temperature influences drill-induced temperature rise of both the bone and the emitted irrigation fluid. Chilled irrigation fluid may reduce the risk of thermal injury to adjacent tissue.

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Vrcek I, Starks V, Mancini R, Gilliland G. Use of an ultrasonic bone curette (Sonopet) in orbital and oculoplastic surgery. *Proc (Bayl Univ Med Cent).* 28(1):91-3, 2015.

Keywords: Orbital Surgery

Financial Disclosures: Brad Rabinovitz works for Stryker Corporation, which makes the Sonopet drill.

Grant Support: None.

Poster 232**Clinical Feature of Unilateral Ptosis with Positive Result in Phenylephrine Test**

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Introduction:

To investigate the etiology and clinical feature of unilateral ptosis in patients with positive result in phenylephrine test (PE).

Methods:

The medical records of 530 consecutive patients with ptosis were retrospectively reviewed. Twenty-six patients (26 eyes) had positive results among 44 patients who have received PE test. Underlying medical conditions were examined based on the previous medical history, laboratory test result and radiologic findings. The treatment method and the results were also evaluated.

Results:

In the 26 patients with positive result in PE test, 14 of 26 (53.8%) patients had associated disease. Nine (34.6%) patients were diagnosed to myasthenia gravis, 4 (15.4%) to Horner's syndrome and 1 (3.8%) to facial palsy. Among 17 patients who could be followed up more than 6 months, 12 of 17 (70.6%) patients have received lid surgery (Levator resection in 5 patients and conjunctivo-mullerectomy in 7 patients) and 5 (29.4%) patients have started medication. The palpebral fissure was 2.2mm increased after 6 months in patients with surgery and 2.0mm in patients with medication. There was no statistically significant difference in change of palpebral fissure between the patients with surgery and the patients with medication ($p=0.446$, Mann Whitney U test).

Conclusions:

It is probable the presence of comorbidities in patients who have unilateral ptosis with positive result in PE test and proper medication of associated disease and lid surgery showed comparable effectiveness. The phenylephrine test should be properly implemented and adequate examination should be taken to find underlying disease if the test showed positive result

Keywords: Ptosis, Phenylephrine Test, Unilateral

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 233**Optic Canal Temperature Change with Ultrasonic Drill Decompression of the Orbital Apex**

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Introduction:

Optic nerve injury is a feared complication of bony decompression of the optic canal and orbit apex. Thermal injury has been suggested as a mechanism. In this animal cadaveric model we assess the change in temperature within the optic canal while using the Sonopet™ ultrasonic bone drill to remove bone at the orbital apex.

Methods:

Six unpreserved porcine orbits were exenterated, with preservation of tissues within the optic canal and extreme orbital apex. Using a Sonopet™ ultrasonic drill, bony decompression was performed as follows: decompression was initiated 3cm anterior to the apex. A 2 mm deep x 10 mm (maximally) wide groove was created and advanced posteriorly to the opening of the optic canal using recommended drill settings of 18ml/min of room temperature (21°C) saline irrigation fluid and 100% power. Temperature at the optic canal was recorded at 15 second intervals using the Thermoworks Super-Fast Thermapen™ probe.

Results:

Mean increase in temperature was $4.54 \pm 3.57^\circ\text{C}$ (range 0-10.5°C). There was a significant correlation between distance from drill tip and change in temperature ($p = 0.0008$). When controlling for distance, we did not find a significant rise in temperature with drilling duration ($p = 0.74$), and while controlling for duration the correlation with distance remained significant ($p=0.0075$).

Conclusions:

In a porcine cadaveric model using the Sonopet™ to decompress the orbital apex, there was a variable change in temperature measured at the optic canal, rising up to 10.5°C above baseline. The variability may result in part from drill tip irrigation, with either obstruction from soft tissue or angle of drill tip influencing access or drainage of irrigation fluid. Our findings suggest thermal injury may be possible with decompression of the orbital apex. Further investigation is warranted.

Keywords: Orbital Surgery

Financial Disclosures: Brad Rabinovitz works for Stryker Corporation, which makes the Sonopet(TM)

Grant Support: None.

Poster 234

Brave New World: 3D Printing of the Orbit for Anatomy, Pathology, Surgical Simulation

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Introduction:

3-Dimensional printing (3D printing), or additive manufacturing, is a novel technique used for rapidly producing physical models of complex structures. These physical models can be used to teach anatomy, simulate procedures, or test surgical anatomical approaches. This project explores the application of 3D printing to ophthalmologic disease. 3D printing has become more affordable and applicable to the clinical setting. Complex orbital anatomy and pathology can be evaluated using a multifaceted approach. We propose a paradigm to create orbital models to teach anatomy and demonstrate pathologies: thyroid related eye disease, idiopathic orbital inflammatory disease, neoplasm, and maxillofacial trauma. Purpose: To demonstrate how 3D printing can be used to produce physical orbit models for the teaching of orbital anatomy, demonstrating pathology, and simulating surgical and interventional procedures.

Methods:

Under IRB approval, clinical cases of orbit anatomy and pathology were selected. Volumetric CT and MR images of the orbit were obtained on Siemens scanners (Erlangen Germany) as part of facial and orbit examinations. Using commercially available software (Mimics, USA), DICOM images were converted to 3D models. Anatomic data was segmented, simplified, and prepared for 3D printing. Models were printed in PLA (polylactic acid plastic) using 3D printer (Ultimaker 2, Ultimaker BV, Netherlands).

Results:

3D models allow trainees to learn anatomy, demonstrate pathology, and test surgical approaches and reconstruction techniques. With hands-on models, learners can grasp complex anatomy, manipulate physical representations of patient specific data, and plan surgical approaches.

Conclusions:

Creating tangible physical models of anatomy and patient specific pathology allows trainees and clinicians to better visualize the critical and complex anatomy of the orbit. This project demonstrates practical application of novel additive manufacturing to orbital anatomy and pathology.

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Keywords: 3-D printing, Teaching, Anatomy, Oculoplastics

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 235**An Incomplete Awakening: Sleep-Induced Apraxia of Eyelid Opening**

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Introduction:

Apraxia of eyelid opening has primarily been described as bilateral loss of volitional ability to open the eyes at certain times and is previously known to be associated with neurodegenerative disease. Rarely it can occur in isolation and as an idiopathic phenomenon. Recently, a few cases have been reported of unilateral apraxia of eyelid opening only upon awakening from sleep, thought to be different than daytime apraxia of eyelid opening. Here we report an additional 11 patients with apraxia of eyelid opening upon awakening.

Methods:

Retrospective, observational case series of patients collected from three separate neuro-ophthalmology practices.

Results:

All 11 patients were Caucasian women with a mean age of 59 years (range 35-80). All patients experienced complete ptosis upon awakening from sleep. Nine patients had unilateral complete ptosis, and 2 had bilateral symptoms. The duration of symptoms ranged from 3 weeks to several years. Ten of the patients reported manually elevating the eyelid to open it, while 1 patient waited for the eyelid to naturally open. After initial manual elevation, all patients reported normal function and position of the eyelids for the remainder of the day. Seven patients had a history of autoimmune disease. Slit lamp and fundus exams were negative for ocular pathology to explain the patients' symptoms. Nine of the patients had brain imaging, all negative for acute pathology.

Conclusions:

Apraxia of eyelid opening occurring only upon awakening from sleep is a rare and relatively newly described entity of which neuro-ophthalmologists should be aware. Imaging and extensive laboratory testing are unlikely warranted without associated neurologic or ocular findings. While our case series suggest that there may be a Caucasian female preponderance as well as an autoimmune relationship to this phenomenon, further reports and investigation are warranted.

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Keywords: Apraxia

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 236**Dynamic Pupillary Responses in Patients with Deep Sedation from Titrating Doses of Continuous Anesthetics**

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Introduction:

Deep anesthetic sedation is the hallmark treatment of refractory status epilepticus (RSE). Use of short half-life anesthetics promotes recovery of consciousness following discontinuation. However, some patients have an unpredictable delay in recovery, adversely affecting their prognosis. Serial neurological examinations are the key measure of recovery with pupillary light reflex (PLR) as a fundamental component of the neurological examination. Automated pupillometry in intensive care units is replacing manual pupil assessment with accurate and objective assessment, which can be monitored over time. Dynamic changes in PLR can be correlated with neurological recovery in a comatose patient.

Methods:

With approval of our institution's review board, we prospectively followed 25 patients treated for RSE with anesthetics, including propofol, midazolam and ketamine. We used portable automated pupillometers to serially monitor PLR on and during titration of anesthesia. This was correlated with serial neurological assessments.

Results:

Upon completion of enrollment we will review data, including medications, treatment response of RSE, serial pupillary assessments, and neurological exam findings. Dynamic pupillary responses will be quantified as NPI™ (Neurological Pupillary Index), comparing recorded values to normative models. This index will be correlated with doses of anesthetics and recovery of level of consciousness.

Conclusions:

This data is valuable in assessing the depth of individual or combination anesthetics and their long term neurological effects. Prognostication of patients treated with anesthetics for RSE is complicated by the recovery of level of consciousness being difficult to predict. We propose that this objective measure would be valuable in quantifying progress in patients recovering from RSE and predicting their neurological recovery.

References: None.

Keywords: Pupils, Pupillary, Reflex

Financial Disclosures: The authors had no disclosures.

Grant Support: None.

Poster 237

The Effect of Entrance Pupil Size and Iris Pigmentation on the Melanopsin Mediated Post-Illumination Pupil Response

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Introduction:

The influence of pupil size and iris color on afferent pupillomotor input and efferent pupillomotor output to red and blue light evoked pupil contraction was studied, using brimonidine induced anisocoria.

Methods:

Pupil responses were recorded using a binocular infrared computerized pupillometer to a series of 1 second duration Ganzfeld red and blue light stimuli (1.0, 1.5, 2.0 and 2.6 log cd/m²) given to each eye. Post-illumination pupil response (PIPR) was the difference in percent pupil contraction to blue vs. red light stimulation at six seconds following stimulus offset. One eye of nine normal subjects was treated with 0.2% brimonidine tartrate ophthalmic solution to achieve pupil size reduction. The consensual pupil response from the eye not treated with brimonidine was compared before and after treatment to study the impact of a smaller pupil to the pupillomotor input. Iris color was graded by inspection.

Results:

Brimonidine treatment produced significant reduction in pupil size in normal subjects (mean reduction in pupil size: 1.72±0.35 mm, p<0.05). The consensual PIPR response was decreased when the brimonidine-treated, miotic pupil was stimulated compared to stimulation of the untreated eye with larger entrance pupil for 2.6 log cd/m² (mean±SD: 28.5±11.6 vs. 35.9±11.2, p<0.05). Brimonidine induced miosis also significantly reduced the PIPR in the treated, miotic pupil compared to the consensual pupil (mean±SD: 9.8±9.8 vs. 16.4±7.9 for 2.0 log cd/m² and 17.1±9.2 vs. 28.5±11.6 for 2.6 log cd/m², p<0.05), indicating an additional reduction in pupil response due to efferent mechanical effects of miosis. This brimonidine induced reduction in afferent pupillomotor input and efferent pupillomotor output did not differ between subjects with dark vs. light irides.

Conclusions:

Smaller entrance pupil size causes significant reduction of afferent pupillomotor inputs and efferent pupillomotor outputs of the PIPR response and was not influenced in this study by iris pigmentation.

Keywords: Post-Illumination Pupil Response, Melanopsin Retinal Ganglion Cell, Entrance Pupil Size, Iris Pigmentation

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