North American Neuro-Ophthalmology Society

Poster Presentations

Tucson Salons A-E

Tuesday, March 9, 2010

6:00 p.m. – 10:00 p.m.

Authors will be standing by their posters at the hours indicated below:

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

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

**Tour of Posters: 8:30 p.m. – 10:00 p.m. (Arizona Salons 1-6)*

Tour Moderators: Kathleen Digre, MD and Lanning Kline, MD

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2	A Case of Unilateral Optic Neuritis Due to Sinus Polyposis	Lisa Law
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81	Thyroid Eye Disease in the Elderly	Hana Leiba
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84	Visual Neuropraxia and Progressive Vision Loss fromThyroid-Associated Stretch Optic Neuropathy	Lenworth Johnson
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93*	Effects of Target Motion Perturbation during Initiation and Steady-State Pursuit in Humans with Infantile Esotropia	Zahra A. Hirji
94*	Anti-Aquaporin-4 Antibody in Chinese Severe Optic Neuritis	Chuntao Lai
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Poster 1

Recurrent Leber's Idiopathic Stellate Neuroretinitis with Diffuse Optic Nerve Enhancement on Orbital Fat-suppressed Cranial MRI

Michael Vaphiades, Eric Wigton

University of Alabama at Birmingham, Birmingham, Alabama, United States

Introduction:
Leber's idiopathic stellate neuroretinitis is not usually associated with optic neuritis and therefore not with multiple sclerosis. The macula star implies disc vasculopathy and secondary leakage of lipoproteinaceous into the macula and optic neuritis would not be expected to produce a secondary macular exudate. We present a case of concurrent Leber's idiopathic stellate neuroretinitis and optic neuritis in the same eye as evidenced clinically and on MRI.

Methods:
Case report.

Results:
6-year-old girl presented with a 4 day history of painless abrupt visual loss OS. She suffered a decrease in vision OD three years prior from Leber's idiopathic stellate neuroretinitis and was treated with antibiotics without improvement of vision.
Examination showed a visual acuity of NLP OD and 20/400 OS, Color vision was 0/7 OD and 1/7 OS Ishihara pseudoisochromatic plates. The pupils were 7 mm OU with no reactivity OD and 2+ reactivity OS and a large RAPD OD. Dilated fundoscopic examination showed an atrophic optic nerve with macular pigmentary change OD and optic nerve edema with a macular star figure OS. A fat-suppressed contrasted enhanced orbital MRI showed diffuse marked enhancement of the left optic nerve and otherwise normal scan. She was treated with oral azithromycin and high dose IV methylprednisolone. Extensive laboratory studies and LP were unremarkable. Eight weeks after treatment, her vision normalized to 20/20 OS.

Conclusion:
Our patient had acute Leber's idiopathic stellate neuroretinitis and optic neuritis OS. We suspect prompt treatment of the optic neuritis component of her visual loss with IV methylprednisolone likely preserved vision OS, unlike the right eye three years prior when treated with antibiotics alone for neuroretinitis. Generally the presence of Leber's idiopathic stellate neuroretinitis precludes the diagnosis of optic neuritis and multiple sclerosis, our case is an exception and therefore has profound prognostic and therapeutic implications.

References:
1. Parmley VC, Schiffman JS, Maitland CG, et al. Does neuroretinitis rule out multiple sclerosis? Arch Neurol 44:1045-8, 1987.
2. Purvin VA, Chioran G. Recurrent neuroretinitis. Arch Ophthalmol 112:365-71, 1994.
3. Schmalfuss IM, Dean CW, Sistrom C et al. Optic neuropathy secondary to cat-scratch disease: Distinguishing MR imaging features from other types of optic neuropathies. AJNR 26: 1310–16, 2005.
4. The optic neuritis study group. Multiple sclerosis risk after optic neuritis: Final optic neuritis treatment trial follow-up. Arch Neurol 65:727-732, 2008.
5. Wals KT, Ansari H, Kiss S, et al. Simultaneous occurrence of neuroretinitis and optic perineuritis in a single eye. J Neuro-Ophthalmol 23:24-7, 2003.

Key Words: Leber's idiopathic stellate neuroretinitis, Neuroretinitis, Optic neuritis, Macular star, Multiple sclerosis

Financial Disclosure: NONE

Poster 2

A Case Of Unilateral Optic Neuritis Due To Sinus Polyposis

Lisa Law, Rachel Robbins, Michaela Mathews

University of Maryland, Baltimore, MD, United States

Introduction:
Aggressive sinus polyposis, while a pathologically benign process, can involve the eye in rare cases. Complications include proptosis, ophthalmoplegia, and optic neuritis. We present a case of subacute optic neuropathy and proptosis, mimicking a sinonasal malignancy, which was ultimately caused by sinus polyposis.

Methods:
A 20-year-old male convict presented to the Eye clinic with progressive proptosis and painless loss of vision in his left eye for 3 months. His previous treatment at the prison infirmary included topical and oral medication for allergic / bacterial conjunctivitis. His general health was excellent, except for seasonal allergies. Examination findings were: Best-corrected vision in the right eye of 20/20 and light perception in the left eye, mildly limited adduction in both eyes, and a left eye with a 4+ APD, 5mm of proptosis, and a pale optic nerve. The remainder of the eye examination was unremarkable.

Results:
MRI imaging revealed marked expansion of the paranasal sinuses suspicious for polyposis or inflammatory changes. There was bilateral enhancement of the optic nerves likely due to neuritis from mass effect. CT demonstrated diffuse osseous expansion and remodeling of the paranasal sinuses. Biopsy confirmed the diagnosis of nasal polyposis.

Conclusion:
Less than 30 cases of optic neuritis due to sinus polyposis or mucoceles have been reported in the literature. Patients usually present with a history of chronic sinusitis slowly progressive visual field defects, proptosis, or diplopia. Our patient presented with subacute loss of vision and proptosis, which was highly suspicious of a malignant process. However, imaging and biopsy revealed the benign nature of the underlying disease. The treatments of choice are surgical debulking and oral steroids. Oculoplastic reconstruction of the orbit may also be necessary to restore normal anatomy and relieve apical crowding.

References: NONE

Key Words: Sinus polyposis, Optic Neuritis

Financial Disclosure: NONE

Poster 3

The Prognosis of Non-Arteritic Ischemic Optic Neuropathy According to the Age of Onset

Ji Woong Chang, Jin Choi, Ho Kyung Choung, Jeong Hun Kim, Young Suk Yu, Seong-Joon Kim

[1]Department of Ophthalmology, Seoul National University Hospital, Seoul, Korea, Republic of, [2]Seoul Artificial Eye Center, Seoul National University Hospital Clinical Research Institute, Seoul, Korea, Republic of

Introduction:
We divided the patients with NAION into two groups as NAION was developed younger or older than 50 years old and compared those two groups to find out if there was any difference of visual prognosis for vision and visual field between two groups.

Methods:
Medical records of NAION patients from 1989 to 2009 were reviewed. Two groups were separated with a standard of age 50. Visual acuity, pattern of visual field defect and the change of them were compared between two groups.

Results:
Total 52 eyes of 48 patients of NAION were enrolled. Younger age group (<50) was comprised of 14 eyes of 12 patients (26.9%). Older age group (≥50) included the rest 38 eyes of 32 patients. Mean age of each group were 43.9 and 64.8 respectively. A half of eyes in each group had <20/200 of visual acuity at initial examination. 28.6% in younger age group and 26.3% in older age group showed visual acuity improvement ($p>0.05$). Superior or inferior altitudinal visual field defect was the most common pattern in general status of visual field defect. 42.9% and 39.5% of eyes in each group demonstrated improved general status of visual field defect ($p>0.05$). 21.4% and 50.0% of eyes in each group showed improved visual field defect in central 5 degree ($p>0.05$).

Conclusion:
The visual prognosis of visual acuity and visual field defect was not significantly different between younger and older age group.

References: NONE

Key Words: non arteritic ischemic optic neuropathy, visual acutiy, visual field, 50 years old, prognosis

Financial Disclosure: NONE

Poster 4

Clinical Impact and Management of the Pathologic Diagnosis of "Healed Arteritis"

Don Bienfang

BWH, Boston, MA, United States

Introduction:
What is the implication of this diagnosis?

Methods:
We looked at 43 biopsies with this diagnosis and reviewed the records on what happened to the patients.

Results:
Despite varying responses by the clinicians to this information only three patients developed serious complications. But none of the complications were in patients who were untreated.

- **Conclusion:**
  At this academic institution, over 10% of temporal artery biopsies received a pathologic diagnosis of "healed arteritis."
- In most cases, a pathologic diagnosis of healed arteritis did not change therapy.
- Over the available duration of follow-up (0 – 9.9 years), poor outcomes were rare.
- It remains unclear whether a diagnosis of "healed arteritis" is clinically meaningful.
- Future studies are necessary to determine whether more stringent pathologic criteria can identify a subset of patients with worse outcomes.

References: NONE

Key Words: Poster, Healed Arteritis

Financial Disclosure: NONE

Poster 5

Radiation Induced Optic Neuropathy: A Retrospective Review of Treatments and Outcomes

Niranjan Bhandare[1], William Mendenhall[1], M. Tariq Bhatti[2]

[1]University of Florida, Gainesville, Florida, United States, [2]Duke University Medical Center, Durham, North Carolina, United States

Introduction:
Radiation induced optic neuropathy (RION),including both anterior radiation induced optic neuropathy (A-RION) and posterior radiation induced optic neuropathy (P-RION), have been recognized as a delayed complication after high doses of radiation therapy (RT) for head and neck cancers often leading to permanent vision loss. The purpose of the study is to analyze the treatments and outcomes in patients with RION at one major academic center.

Methods:
A retrospective chart review of pre- and post- RT ophthalmologic records of 17 patients who were subjected to treatment for RION between 965-2008.

Results:
Of the six patients with A-RION, one treated with hyperbaric oxygen (HBO) immediately at onset of symptoms showed improvement in vision. None of the 5 patients with A-RION treated with corticosteroids alone or with corticosteroids and HBO showed stabilization or improvement in vision.

Of the 11 patients with P-RION, Three patients treated immediately at onset of symptoms with combination therapy (two with corticosteroids and HBO and one with warfarin and HBO) showed improvement in vision. None of the 7 patients (one treated with HBO, three treated with corticosteroids alone, one with corticosteroid and HBO, and two with warfarin alone) had stabilization or an improvement in vision. One patient treated with Bevacizumab showed stabilization in vision but no improvement, though the follow up on this patient remains short.

Conclusion:
Although there was vision stabilization and improvement in some of cases of RION, none of the treatment options delivered consistent results. Anti-vascular growth factor agents may be a potential treatment option for patients with RION but it requires further investigation.

References: NONE

Key Words: radiation optic neuropathy, anti-vascular growth factor agents, Radiation treatment

Financial Disclosure: NONE

Poster 6

Leber's Hereditary Optic Neuropathy (LHON) Mimicking Neuromyelitis Optica (NMO)

Gregory Van Stavern, Collin McClelland

Washington University School of Medicine, St. Louis, Missouri, United States

Introduction:
Leber's Hereditary Optic Neuropathy (LHON) is a mitochondrial disease marked by subacute onset optic neuropathy in one eye frequently followed by subsequent involvement of the contralateral eye. Rarely patients with LHON have an associated multiple sclerosis-like disease. We report a case of subacute, bilateral simultaneous-onset optic neuropathy in a patient with a prior history of cervical myelopathy found to have a pathogenic LHON mutation.

Methods:
Single Case Report

Results:
A 65-year-old African American female with a one year history of cervical myelopathy presented after sequential, subacute onset of simultaneous, painless vision loss in both eyes. There was no family history of autoimmune diseases or unexplained vision loss. The exam was significant for best corrected visual acuity of 6/200 OU, normal anterior segments, no significant retinal pathology, and optic nerves with bilateral cup:disc ratio of 0.75 and moderate temporal pallor. An extensive work-up had demonstrated no signs of systemic malignancy or vasculitis, normal cerebrospinal fluid, no nutritional deficiencies, and absence of serum anti-aquaporin4 antibodies. Brain and orbital MRI were normal, while spinal cord MRI showed diffuse cervical spine T2 hyperintensity of the posterior columns without contrast enhancement. Genetic testing revealed the 14484 LHON mitochondrial DNA mutation.

Conclusion:
Leber's hereditary optic neuropathy (LHON) is rarely associated with multiple sclerosis-like features which can confound the diagnosis. We present a case of LHON masquerading as neuromyelitis optica (NMO). We highlight features of the clinical examination and MRI which were suggestive of an alternative diagnosis, and review the literature regarding LHON and MS. The diagnosis of LHON should be considered in all cases of acute or subacute bilateral optic neuropathy, including presumed sero-negative NMO.

References: NONE

Key Words: Leber's Hereditary Optic Neuropathy , Neuromyelitis Optica

Financial Disclosure: NONE

Poster 7

Cognitive Impairment In Optic Neuritis And Early Stage Of Multiple Sclerosis

Xiaojun Zhang, Jingting Peng, Yuhong Gao, Houliang Sun, Yanbin Sun, Yang Lu, Kemei Chi

Capital Medical University Beijing Tongren Hospital, Beijing, China

Introduction:
To evaluate cognitive impairment in idiopathic demyelinating optic neuritis (IDON) and early stage of multiple sclerosis (MS)

Methods:
A Consecutive group of IDON patients were divided into 2 groups according to with (IDON-A) or without (IDON-N) abnormal brain MRI. MS patients with course shorter than 5 years and minor physical disabilities (EDSS≤4 scores) were included as early MS group. A battery of neuropsychological tests and three-tone paradigm auditory P_{300} were performed.

Results:
Information processing speed, attention, verbal and spatial memory function in the IDON-A and MS groups were found worse than IDON-N and control groups (P<0.01). Executive function were statistically worse (P<0.01, 0.05) in IDON-A and MS groups than control group, but not significantly different with IDON-N group. No significant differences were found between the IDON-N group and control group in almost all neuropsychological tests. P_{300} latency was similar between IDON-A and MS groups, but statistically longer than IDON-N group. Cognitive decline was weakly correlated with disease duration, EDSS scores and depression, but it was independent of numbers of attack.

Conclusion:
Cognitive capacity damage, especially information process speed, attention, verbal and spatial memory, executive function, can appear in very early stage of MS and even in clinically IDON stage.

References: NONE

Key Words: Optic neuritis, Multiple sclerosis, Cognitive dysfunction, Neuro-psychological tests, Event-related potential

Financial Disclosure: NONE

Poster 8

Case-Crossover Study of PDE5 Inhibitor Exposure as a Potential "Trigger Factor" For Acute NAION

Alexander Walker[2], Ulka Campbell[1], Kenneth Petronis[1], Brian Klee[1], Francesca Kolitsopoulos[1], Jingping Mo[1]

[1]Pfizer Inc., New York, NY, United States, [2]World Health Information Science Consultants, LLC, Wellesley, MA, United States

Introduction:
Nonarteritic anterior ischemic optic neuropathy (NAION), a condition characterized by sudden partial vision loss, has been rarely reported in patients taking phosphodiesterase type 5 inhibitors (PDE5i). Prior studies have not definitively addressed whether PDE5i exposure is associated with increased risk of NAION. The objective of this study is to examine whether use of PDE5i, as a class, triggers the onset of acute NAION within a pharmacokinetically-defined time period following drug ingestion.

Methods:
A case-crossover study has been initiated. Potential cases of acute NAION are prospectively identified from over 100 ophthalmology centers in the United States and Europe. Detailed information on PDE5i exposure prior to NAION symptom onset is obtained via telephone interviews from a centralized Study Call Center. An Adjudication Committee classifies subjects as acute NAION cases or non-cases on the basis of findings from the ophthalmologic exam, fundus photography and other clinical assessments. For the primary analysis, the case window will be defined as the one-day period immediately preceding symptom onset, and there will be 29 control windows corresponding to the 29 days preceding the case window. A stratified (by subject) Mantel-Haenszel analysis will be used to estimate the odds ratio. Sample size estimates, based on a detectable odds ratio of 3.0, require 40 acute NAION cases who used a PDE5i in the month prior to symptom onset; usage data indicate that this number of exposed men requires identification of approximately 800 NAION cases.

Results:
Subject enrollment was initiated in October 2008 and is being monitored by a Scientific Steering Committee

Conclusion:
Using detailed information on PDE5i exposure and standardized NAION case classification, this case-crossover study will provide real-world evidence of whether exposure to PDE5i triggers the onset of acute NAION.

References: NONE

Key Words: NAION, PDE5 inhibitors, Pharmacoepidemiology, Drug safety

Financial Disclosure: U Campbell, K Petronis, B Klee, F Kolitsopoulos, and J Mo are all employees of Pfizer.

Poster 9

Interferon Alpha-Associated Optic Neuropathy

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Introduction:
Peginterferon alpha is approved for treatment of chronic hepatitis C (CHC). Interferon alpha retinopathy is common. We describe a patient who developed NAION while taking peginterferon alpha and review the literature.

Methods:
We report a single case and review the literature. A PUBMED search was conducted using the search terms: *ischemic, optic neuropathy, interferon, ribavirin.* Additional studies were identified from bibliographies of retrieved articles.

Results:
A 52-year-old man with CHC presented with painless, bilateral, simultaneous NAION and peripheral neuropathy. Symptoms began 19 weeks after starting peginterferon alpha-2a and ribavirin. The peripheral neuropathy and vision OD improved, but the vision OS worsened after stopping interferon.

We identified 23 cases of NAION during interferon alpha therapy. Patients lost vision 1-40 weeks after initiating therapy. Of 13 patients receiving ribavarin and interferon alpha therapy, 6 experienced bilateral NAION. Of 21 eyes had documented initial and followup acuities, 8 improved, 1 worsened, and the rest remained stable. One patient had a painful peripheral neuropathy.

Conclusion:
Interferon alpha treatment may result in NAION. Clinicians should consider this diagnosis and discontinue interferon alpha among patients presenting with bilateral, simultaneous NAION.

References: NONE

Key Words: interferon alpha, anterior ischemic optic neuropathy

Financial Disclosure: NONE

Poster 10

NON-ARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY (NAION) AND RISK OF CEREBROVASCULAR AND CARDIOVASCULAR DISEASE

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Introduction:
Previous reviews have not found NAION to be a risk factor for stroke, but patients with NAION do appear to be at increased risk of cardiovascular disease. The objective of this investigation was to ascertain the risk of cerebrovascular and cardiovascular disease in our patients with NAION. We used cardiac CT as an independent means of ascertaining the prevalence of cardiovascular disease in NAION patients.

Methods:
University & Eye Center records were searched for NAION patients (ICD 377.41) during the period 1992-2008. Patient characteristics were compared between the groups of subjects with and without cerebrovascular disease. The Framingham stroke risk profile was used to determine the number of expected strokes. Cardiac CT was used in a subset of patients to estimate the prevalence of cardiovascular disease.

Results:
176 NAION cases were confirmed. Six subjects had suffered stroke prior to presentation. With a mean follow-up period of 279 days, 2 subjects suffered stroke after being diagnosed with NAION. 1.8 strokes would have been expected during a 2-year follow-up. The groups with and without stroke did not differ significantly with regard to the presence of diabetes, hypertension, atrial fibrillation, or myocardial infarction. 38% of subjects with stroke had cardiovascular disease and 8% of patients without stroke had cardiovascular disease (p-value 0.03). Calcium scoring using cardiac CT is ongoing and these data will be presented at the Annual Meeting.

Conclusion:
Our data are consistent with previous reports indicating that NAION does not appear to be an independent risk factor for stroke, but that NAION patients are more likely to manifest cardiovascular disease. We will have a more accurate determination of the prevalence of cardiovascular disease once calcium scoring of cardiac CT examinations is complete. We expect that the number of patients with cardiovascular disease may be underestimated by physicians treating patients with NAION

References:
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Key Words: NAION, Cerebrovascular disease, Stroke, Coronary Calcium Score, Coronary artery disease

Financial Disclosure: NONE

Poster 11

Toluene Optic Neuropathy: MRI and Pathological Features

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Introduction:
Toluene (methyl benzene) is a colorless liquid found in glues, paints, and industrial products. Vapour exposure occurs through occupation or purposeful inhalation. Toluene is lipid soluble and rapidly absorbed by the lipid-rich CNS. Clinical effects are mediated through stimulation of the γ-aminobutyric acid (GABA) receptor complex. Prolonged exposure leads to neurologic abnormalities.

Methods:
Two men presented with multifocal CNS defects and bilateral optic neuropathy of unclear etiology. After numerous diagnostic tests, including brain MRI, lumbar puncture, serum studies, and in one patient a brain biopsy, chronic inhalational of toluene was found to be the cause.

Results:
Both patients reported progressive visual loss accompanied by decline in neurologic function. Examination of the first patient revealed count fingers OD, 20/400 OS, absent color vision, and optic nerve pallor OU. The second patient was light perception OU with optic nerve pallor OU.

Brain MRI in our first patient showed hyperintensity of cerebral white matter, loss of gray/white matter differentiation, and thinning of the corpus callosum. MRI of the second patient revealed diffuse symmetric leukoencephalopathy.

Neuropathologic examination of the frontal deep white matter revealed myelin pallor and rarefaction with chronic astrogliosis and microgliosis. Rare perivascular chronic inflammation was seen.

Conclusion:
Our findings are consistent with previously reported cases, but etiology of the optic neuropathy was particularly mystifying because initially neither patient admitted to solvent abuse. Therefore, both patients underwent extensive evaluation, including brain biopsy in one patient, which delayed final diagnosis.

To our knowledge, this is the only reported case of diagnostic brain biopsy premortem. Our second case is interesting because the characteristic pattern of bilateral symmetric signal abnormality in the cerebral white matter on brain MRI lead to diagnosis.

Timely diagnosis is important because patients may experience improvement in neurologic and ocular manifestations with abstinence, whereas continued abuse can result in permanent loss of neurologic function.

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Key Words: Toluene, Optic Neuropathy, Solvent Abuse, Leukoencephalopathy, Huffing

Financial Disclosure: NONE

Poster 12

Low IOP as an Indicator of MS in Patients Suspected of Suffering from Optic Neuritis, Nystagmus or Diplopia

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Introduction:
IOP is checked routinely in all patients presenting to a Neuro-ophthalmologist for different causes of visual disturbance. Our neuro-ophthalmologist noted that patients who carried a diagnosis of MS, or who later were diagnosed with this disease, often had lower than normal IOP's on examination in both eyes in the range of 10. A retrospective study was performed to verify the observation, and confirmed this suspicion.

Methods:
Between 2004 and 2008 a retrospective chart review was performed. Thirty patients (60 eyes) with Multiple Sclerosis were included in the study. All patients had been followed by one examiner. Goldmann Applanation Tonometry was used on all patients. Thirty age matched controls were also studied. Patients with a history for glaucoma were excluded from the study. Patients who received IV steroids within 1 month of the evaluation were also excluded. Corneal thickness was measured in most cases. The data was analyzed by Student t test with statistical significance set at $p < .05$.

Results:
The IOP in the MS group ranged from 7-18mm Hg with a mean of 12.3mm Hg and a median of 13mm Hg, while in the control group it was higher with a mean of 14-23mm Hg and a median of 17mm Hg. When the IOP was corrected by CCT the mean IOP in the MS group was 11.6 compared to 15.8 in the control group. No significant difference ($p=.25$) was found between the CCT of the MS group (mean 556 um) versus the control group (mean 560 um).

Conclusion:
Patients with MS often have lower IOP than the normal population, or patients with other forms of optic neuropathy. Though OCT is very helpful in catching subclinical RNFL loss in patients with MS, earlier suspicion of the disease can be entertained when a lower IOP is noted on initial screening exam.

References:
1. Colton, Ederer. The Distribution of IOP in the General Poulation. Survey of Ophthalmology; 25(3):123-128, 1980.
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Key Words: Intraocular pressure, multiple sclerosis, optic neuropathy, corneal thickness, diagnostic tool

Financial Disclosure: NONE

Poster 13

Intravitreal Bevacizumab for Treatment of Non-Arteritic Anterior Ischemic Optic Neuropathy

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Introduction:
Currently there is no proven treatment for non-arteritic anterior ischemic optic neuropathy (NAION). Recent studies have shown promising visual results with the injection of intravitreal triamcinolone [1-4] and bevacizumab [5] in NAION. The proposed mechanism of this improvement is the reduction of initial post ischemic inflammation leading to reduce long term tissue damage.

Methods:
In this non-randomized controlled clinical feasibility trial, criteria for inclusion were diagnosis of NAION and presentation within 3 days of NAION event. Patients were excluded if they refused participation. Participants consenting to the treatment arm were given intravitreal injection of bevacizumab (1.25mg/mL) within 7 days of NAION event; controls were followed with no intervention. Clinical assessment of optic nerve swelling, visual acuity measurement, visual field assessment and monitoring for complications were performed on day 0, post injection day 1 and at 1, 3, 6 and 12 months.

Results:
At the time of writing recruitment is 40% complete. Age at baseline was 56 and 71 years for control and treatment respectively. Baseline visual acuity and mean deviation was 20/200 and 20/100 and -14.1 and -6.4 for control and treatment. Disk swelling was 2+ for both groups. Three and six month data is forthcoming.

Conclusion:
The use of intravitreal bevacizumab for the treatment of acute NAION may reduce optic nerve swelling, leading to improved visual acuity and visual field outcomes. Interim results will be discussed.

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Key Words: NAION, bevacizumab, treatment

Financial Disclosure: NONE

Poster 14
Retrospective Review of Monocular Hemianopia: Patterns of Visual Field Loss and Etiologies

Scott Haines, Timothy Martin

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Introduction:
Monocular hemianopia is an unusual pattern of visual field loss that has been reported to be due to both non-organic and organic causes[1,2]. *Temporal* monocular hemianopia is well-established as a potential result of optic nerve or chiasm lesions [1], but the clinical implications of *nasal* monocular hemifield defects are less clear. This study explores the characteristics of Humphrey threshold perimetry and clinical diagnosis in patients with monocular *temporal or nasal* hemifield visual field defects that respect the vertical meridian.

Methods:
Forty-four consecutive patients with monocular hemianopia were categorized by diagnosis (including non-organic) and visual fields characteristics: *nasal* or *tempora*l, *perfect* (complete, with absolute respect of the midline) or *imperfect* (incomplete, some extension across the midline, or minor visual field abnormalities in the fellow eye). Patients with multiple visual fields that changed over time offered additional insight.

Results:
The 23 patients with organic etiologies included: tumors involving the chiasm (7), demyelinating disease (6), traumatic optic neuropathy (3), other optic nerve disease (5) and retinal disorders (2) . Twenty-one patients were categorized as non-organic. *Nasal* hemianopia was more common in the non-organic group (8 of 13), as was *perfect* hemianopia (12 of twenty). Changes in repeat perimetry were instructive: a bitemporal visual field defect became a monocular *temporal* defect after resection of a pituitary tumor; a patient with non-organic monocular hemianopia switched from *temporal* to *nasal*.

Conclusion:
Both *nasal* and *temporal* monocular hemianopia can be the result of disease. *Perfect* and *nasal* hemianopias are more likely to be non-organic.

References:
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2. Keane JR. Patterns of hysterical hemianopia. J Neurol 51(4):1230-31; 1998.

Key Words: hemianopia, monocular, functional, non-organic, etiology

Financial Disclosure: NONE

Poster 15

Low Contrast Visual Acuity is Decreased in Patients with Parkinsonism

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Introduction:
Studies evaluating visual/visual spatial and motor symptoms in [non-demented Parkinsonian patients (PD)] report problems in tasks dependent on visual/visual spatial contrast sensitivity[1] including difficulties with navigation, driving[2], etc. Human retina contains dopaminergic amacrine cells, and retinal dopamine content is lowered in PD patients.[3] Identified defects in low spatiotemporal contrast sensitivity in PD patients are presumed the consequence of loss of dopamine. Further, the inner-retinal layer is demonstrably thin in patients with relatively early Parkinsonism.[4] We studied patients in various stages of Parkinsonism with low contrast sensitivity testing.

Methods:
A pilot study of 61 patients, 31 with PD and 30 controls (average age: 74 y/o PD; 71 y/o control). Low contrast visual acuity was tested using front illuminated Low-Contrast Sloan Letter Charts (LCSLC- Precision Vision, LaSalle, IL) at 2 meters; room lighting 80-100 cd/m^2. Patients wore distance correction (>20/40) and read charts binocularly. 100%, 2.5%, and 1.25% charts used. Parkinsonism was scored using the United Rating Scale for Parkinsonism (UPDRS). Retinal nerve fiber and macular thickness evaluated using Optical Coherence Tomography (spectral-domain OCT Analysis). We excluded patients with known eye pathologies including retinopathies, significant macular degeneration, glaucoma, optic neuropathies, Alzheimer's disease, and strokes involving the visual pathways. Wilcoxon (Mann-Whitney) distribution free rank sum tests were applied to each of the data level subsets factored by Parkinson's (P) or Control (C).

Results:
There is evidence to reject the null hypothesis and conclude the location of distribution of Parkinson's scores was significantly less than controls at levels 2.5% (significant at 0.0075) and 1.25% (significant at 0.0002). The evidence is especially strong at the 1.25% level.

Conclusion:
PD patients have substantially poorer low contrast visual acuity than controls. Low contrast sensitivity testing may prove valuable for nonmotoric screening in suspected Parkinsonism and during neuroprotective treatment of PD. Test correlations with OCT and UPDRS will be discussed.

References:
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Key Words: Parkinsonism, low contrast sensitivity, OCT, UPDRS

Financial Disclosure: NONE

Poster 16

Rarebit Perimetry: Number Of Test Passes Needed To Identify Visual Field Defects

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Introduction:
Rarebit perimetry has been demonstrated to effectively detect a wide range of visual field defects. The current protocol includes 5 test passes for data collection. In a recent study we demonstrated the usefulness of Rarebit to detect visual field defects in hospitalized patients. Since attention span is limited we were interested in whether the test could be shortened in duration without losing the ability to recognize pattern defects.

Methods:
10 individuals with distinct visual field abnormalities (initially detected by standard automated perimetry) underwent Rarebit testing on each eye utilizing the protocol described by author Lars Frisen, first completing 5 passes followed by 2 passes. A neuro-ophthalmologist blinded to the patient data verbally described the defect pattern of visual fields from each patient, randomly selected as either 5-pass or 2-pass fields, and then attempted to match these fields with their correlates (e.g. 5-pass with 2-pass fields) based on pattern recognition and verbal description. In addition, mean test time (MTT) for each eye was compared between the 5-pass and 2-pass tests.

Results:
10 patients (3 men, 7 woman) ages 38-88 (average 60.6, SD $\pm$ 15.0) participated. In the matching exercise, each field was successfully paired with its correlate, and verbal descriptions of correlate fields were consistent. MTT for each eye was 6.95 min (SD $\pm$ 0.71 min) on 5-pass tests and 2.37 min (SD $\pm$ 0.29 min) on 2-pass tests.

Conclusion:
Rarebit perimetry can reliably demonstrate visual field defects when performed using 2 passes. Additionally, 2-pass testing is 65.9% shorter in duration compared to 5-pass testing and is completed in approximately 7 minutes for both eyes, including time allowed for patient instruction. A visual field test that minimizes testing time while maintaining reliable detection of defects may benefit patients that are unable to complete time-intensive exams.

References: NONE

Key Words: perimetry, visual fields, Rarebit

Financial Disclosure: NONE

Poster 17

Homonymous Hemianopia from Optic Tract Infarction in Deep Cerebral Venous Thrombosis

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Introduction:
Thrombosis of the deep cerebral system is rare. Frequently reported risk factors include oral contraceptives, pregnancy and the pueriperium. The most commonly reported parenchymal abnormality on imaging is thalamic edema and the most frequently described clinical presentation has been headache and altered level of consciousness. Homonymous hemianopia has been described in thrombosis of the deep cerebral venous system, but the visual field deficit and corresponding anatomic injury have not been described in detail. Damage to the optic tract has not been previously identified as the cause of the homonymous hemianopia.

Methods:
Case study and literature review.

Results:
We report the case of a 22 year old man who developed deep cerebral venous and sinus thrombosis following a stay at high altitude. He exhibited a complete right homonymous hemianopia, hemisensory deficit and hemiplegia. Brain imaging demonstrated deep cerebral venous system thrombosis and asymmetric signal abnormalities in the region of the thalamus, optic tracts, and internal capsule.

Conclusion:
This is the first report to localize homonymous hemianopia in this setting to the optic tract and to provide full clinical and imaging details. Homonymous hemianopia was the patient's most debilitating deficit. Its persistence is explainable by the degree of damage to the optic tract evident on imaging.

References: NONE

Key Words: Optic Tract Infarction, Deep Cerebral Venous Thrombosis, Homonymous Hemianopia

Financial Disclosure: NONE

Poster 18

Homonymous Hemianopia as a Clinically Isolated Syndrome Heralding Multiple Sclerosis

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Introduction:
Retrogeniculate involvement of the visual pathway manifesting as homonymous hemianopic visual field loss is rare in the course of multiple sclerosis (MS). Optic nerve involvement overwhelmingly predominates. We report sectoral, quadrantic or complete hemianopia as presenting manifestations in 3 patients.

Methods:
Cases studied with MRI, perimetry, CSF analyses and evoked potentials.

Results:
1). A 43 year old woman presented with left sectoranopia with imaging and C.S.F findings consistent with MS. MRI showed plaques in the right lateral geniculate body, and optic radiation. Follow up showed incomplete improvement in the visual field over 2 years.
2). A 38 year old man developed sudden blurry vision in the left peripheral field. Perimetry revealed bilateral homonymous hemianopia and MRI showed multiple white matter lesions in both optic radiations and elsewhere consistent with MS.
3). A 22 year old woman complained of blurred vision. Goldmann and Humphey perimetry revealed complete left homonymous loss and several months later a left inferior quadrantanopic defect. MRI showed demyelination in the parieto-occipital lobes. Possible MS was diagnosed. She had three relapses with ataxia after two years. MRI showed regression of some large lesions and appearance of new demyelination lesions but she has been stable during follow up visits with little improvement in the visual field over 4 years.

Conclusion:
MS can present with homonymous hemianopia. One possible explanation for the rarity of recognized visual loss from retrochiasmal demyelination is that the optic radiation anatomically fans out to a large area and a plaque affects fewer fibers than in the optic nerve where fibers are compactly arranged. Also, physiologic conduction properties of demyelinated optic radiation fibers may differ from retinal axons. Diffuse involvement of the posterior visual pathway (in one patient) and this being the only initial manifestation may be reasons for this rarely recognized presentation with hemianopia.

References: NONE

Key Words: Multiple Sclerosis, Homonymous Hemianopia, Clinically Isolated Syndrome, Visual Field, Demyelination

Financial Disclosure: NONE

Poster 19

Are Red-Green Color Tests the Best Option Out There?

Naomie Kraus, Nilufer Kale, Eric Eggenberger

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Introduction:
Various studies have shown that optic neuropathies might present with dyschromatopsia and current clinical measures such as Ishihara color plates or Hardy-Rand –Rittler color plates have been utilized to assess color desaturation. However, the degree of color desaturation is still evaluated subjectively and evaluating the types of color loss presented with certain types of optic neuropathies including optic neuritis is warranted. In this study, we aim to introduce a computer program as an alternative method to quantify color desaturation and discuss the current red-green tests utilized for dyschromatopsia.

Methods:
Patients with unilateral optic neuropathy are identified from electronic medical records from the Michigan State University database. A Powerpoint presentation has been designed wherein a colored square (i.e. red, blue, yellow, or green) is presented on one-half of the screen, and a similar square of color desaturation is presented on the other half. The degree of desaturation increases from one slide to the next. A black curtain in between the subject's eyes runs from the subject's face to the computer screen so that the subject's eyes see a different side of the screen. The patient uses the keyboard arrows to forward the slides, increasing the desaturation of the color square presented to the normal eye until it equals in saturation the color square of the affected eye.

Discussion:
The results of this study will be beneficial to determine the accuracy an additional method to assess dyschromatopsia. Quantification of color desaturation can also be used as an additional parameter in guiding clinicians in diagnosing, monitoring and treating patients with optic neuropathy.

References: NONE

Key Words: optic neuropathies, dyschromatopsia

Financial Disclosure: NONE

Poster 20

Retinal Thickness in Patients with Mild Cognitive Impairment and Alzheimer's Disease

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Introduction:
Mild cognitive impairment (MCI) may in some cases represent a transition to early Alzheimer's disease (AD). The retinal nerve fiber layer (RNFL) is composed of axons originating in retinal ganglion cells that eventually form the optic nerves. Previous studies have shown that the RNFL is involved in patients with memory complaints, particularly AD. The objective of this study was to assess the relationship between MCI, AD and loss of RNFL.

Methods:
In this study, patients fulfilling diagnostic criteria for MCI (n= 24) and AD (n=31), and cognitively normal age-matched controls (n=18) have undergone neuro-ophthalmologic and optical coherence tomography (OCT) examinations, to measure RNFL thickness.

Results:
There was a decrease in RNFL thickness in both study groups (AD and MCI) compared to the control group, particularly in the superior and inferior quadrants of the optic nerve. Although AD patients showed more severe changes than MCI cases, the differences were statistically not significant. Furthermore among AD patients, there was no relation to the severity of the dementia.

Conclusion:
Our data confirm the retinal involvement in AD, as reflected by loss of axons in the optic nerves. The relationships of these findings to the cognitive changes need to be studied further in a larger cohort of patients. Furthermore, prospective studies should clarify whether MCI patients with reduced RNFL thickness are at an increased risk to develop dementia.

References: NONE

Key Words: retinal nerve fiber layer, Mild cognitive impairment , Alzheimer's disease

Financial Disclosure: NONE

Poster 21

Ultrahigh Resolution Optical Coherence Tomography Findings In Patients With Microscotomas and Metamorphopsia

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[1]New England Eye Center, TUFTS Medical Center, Boston, United States, [2]Department of Electrical Engineering and Computer Science, MIT, Cambridge, United States

Introduction:
Patients with small idiopathic central or paracentral microscotomas may have a normal clinical ophthalmic exam. Ultrahigh resolution optical coherence tomography (UHROCT) may be useful to objectively detect and define visually significant structural retinal defects in these patients.

Methods:
Time domain and spectral domain based UHROCT was used to obtain macular scans in patients with idiopathic central or paracentral microscotomas or focal (less than 1 degree) areas of metamorphopsia detected on Amsler grid. A full ophthalmic exam was performed including ancillary testing such as visual fields, multifocal electroretinograms and fluorescein angiograms.
Patients with solar maculopathy or epiretinal membranes were excluded from the study. 29 patients were scanned between 2005 and 2008. 5 patients were selected showing distinct macular defects on UHROCT.

Results:
Five out of 29 patients showed distinct macular defects visualized by UHROCT. The defects remained stable over time. Best corrected visual acuity ranged from 20/20 to 20/30 in the affected eyes. Ancillary tests were normal.

Conclusion:
Higher image resolution and acquisition speed of UHROCT allows detection and visualization of distinct outer retinal macular defects in patients presenting with central or paracentral microscotomas and metamorphopsia on Amsler grid. The defects remained stable over time.

References:
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Key Words: Ultrahigh resolution optical coherence tomography, Microscotomas, Metamorphopsia

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Poster 22

Evaluation of Retinal Nerve Fiber Layer Thickness Measurements in Longitudinally Extensive Tranverse Myelitis

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Introduction:
Acute transversal myelitis (ATM) is a disease characterized clinically by acute neurologic dysfunction in motor, sensory and autonomic nerves and tracts of spinal cord. When the spinal cord involvement occurs in less than two vertebral segments, there is high risk of developing multiple sclerosis (MS). However, when ATM is associated with longitudinally extensive lesions on magnetic resonance image, spanning three or more vertebral segments, there is a low risk of developing MS and the condition is known as longitudinally extensive transverse myelitis (LETM).
The purpose of this study was to measure the RNFL thickness using OCT in patients with idiopathic LETM without previous episodes of optic neuritis and evaluate the possible occurrence of subclinical involvement of the optic nerve in these patients.

Methods:
Twenty six eyes from 26 patients with LETM and 26 eyes from 26 controls subjects underwent time-domain OCT and standard automatic perimetry (SAP). The measurements in both groups were compared. Areas under receiver operating characteristics curve (AUC) were calculated. Correlations between RNFL thickness measurements and SAP mean deviation (MD) values were performed using Pearson's correlation coefficient (R).

Results:
No significantly difference in measurements of RNFL thickness between LETM group and control group was found, except for the nasal quadrant and the 30-degree segment located at the 3 o'clock position (P = 0.04 and 0.006, respectively). The higher AUC`s were found for the same parameters (AUC = 0.65 and AUC = 0.72, respectively). A significant correlation was observed between the 3 o'clock segment measurement and the visual field assessed using the mean deviation.

Conclusion:
Patients with LETM can present localized RNFL loss on OCT particularly in the nasal disc area associated with discrete visual field defects even in absence of previous episodes of optic neuritis. These findings are consistent with optic nerve damage and emphasizes that patients with LETM may experience attacks of subclinical optic neuritis.

References: NONE

Key Words: Optical Coherence Tomography, Retinal Nerve Fiber Layer, Transverse Myelitis

Financial Disclosure: NONE

Poster 23

Contribution of Structural and Functional Visual Outcomes to Work Capacity and Employment Status in Multiple Sclerosis

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Introduction:
Work capacity and employment status are increasingly recognized as important outcomes in multiple sclerosis (MS). The extent to which visual symptoms, reduced function, and visual pathway axonal loss contribute to work disability has not been examined. We determined how visual outcome measures, retinal nerve fiber layer (RNFL) thickness, and vision-specific quality of life reflect work capacity and employment status in an MS cohort.

Methods:
Patients with MS completed a vocational status questionnaire. Work status was categorized as disabled (receiving disability pension), reduced-capacity, or full-time. Those unemployed for non-MS reasons or retired due to age were not included in primary analyses (n=10). Low-contrast letter acuity (LCA) and high-contrast visual acuity (HCA) testing were performed. RNFL thickness was measured by time-domain OCT and spectral-domain OCT high-resolution optical coherence tomography (OCT). HRQOL was assessed using the NEI-VFQ-25 with Neuro-Ophthalmic Supplement, Impact of Visual Impairment Scale (IVIS), and SF-36.

Results:
MS patients (n=47, 94 eyes), aged 48±9 years, were classified into one of the three work status categories. Visual symptoms were most common among patients who were disabled (71%) compared to reduced capacity or full-time (40%). Disabled patients had the worst scores for NEI-VFQ-25 composite ($p=0.008$, linear regression, accounting for age), Neuro-Ophthalmic Supplement ($p=0.002$), IVIS ($p=0.003$), and SF-36 Physical Components Score ($p<0.001$). RNFL thickness was lowest in disabled patients ($p=0.004$ [time-domain OCT], $p=0.001$ [spectral-domain OCT]), even accounting for visual symptoms and history of ON ($p=0.004$). Worse visual function scores were associated with greater probability of visual symptoms but did not correlate independently with work status.

Conclusion:
Measures of vision-specific HRQOL and RNFL thickness reflect work capacity and employment status in MS, even among a heterogeneous cohort of patients not selected for visual symptoms. Anterior visual pathway axonal loss and visual dysfunction are likely contributors to disability in MS, and may be markers for potential economic impacts of this disorder.

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Key Words: Multiple Sclerosis (MS), Low-Contrast Letter Acuity, Optical Coherence Tomography (OCT), Quality Of Life, Disability

Financial Disclosure: NONE

Poster 24

OCT-3 Quality Control in Two Multi-Center Multiple Sclerosis (MS) Therapeutic Trials

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Introduction:
To report the influence of Optical Coherence Tomography (OCT) quality control (QC) procedures on rates of test parameter and shipping errors in two multi-center MS therapeutic trials.

Methods:
The UC Davis OCT Reading Center evaluated 10,910 OCT scans from 534 patients enrolled in two MS therapeutic trials. A total of 2,178 scans from 40 patients were evaluated in Trial 1, and a total of 8,732 scans from 494 subjects were evaluated in Trial 2. Trial 1 involved secondary progressive MS patients while Trial 2 involved relapsing-remitting MS patients. Technicians were certified for OCT in Trial 1 and were required to submit OCT data in accordance to the Reading Center protocol. There was no technician certification or ongoing monitoring in Trial 2. The Reading Center's QC system addressed two areas of clinic performance: test parameter errors and shipping errors. Test parameter errors included, (1) signal strength < 7, (2) exported data missing, (3) missing scans, (4) scans requiring redraws, and (5) scans performed on the wrong OCT instrument.

Results:
A total of 22% (1,926/8,732) of the OCT scans had signal strength < 7 in Trial 2, while only 3.7% (81/2,178) of the scans had a signal strength < 7 in Trial 1. Trial 2 had 10.3% (897/8,732) of the scans missing exported data which required manual entry. Trial 1 had 0 scans with the same error. Trial 2 had 67 scans (1%) performed on the wrong instrument and 64 scans (1%) required a redraw of the analysis line, while Trial 1 had neither error.

Conclusion:
Technicians who are trained and certified in standard fashion help reduce poor quality OCT data in MS trials. Prompt transmission of OCT data to the Reading Center for ongoing and intensive QC monitoring and rapid feedback to technicians helps to reduce the frequency of incorrect testing parameter and shipping errors.

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Key Words: Multiple Sclerosis, Quality Control, Optical Coherence Tomography

Financial Disclosure: NONE

Poster 25

Optical Coherence Tomography (OCT) Measurements and Visual Function Tests in a Secondary Progressive MS Trial of the Compound Dirucotide

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Introduction:
Axonal loss in multiple sclerosis (MS) is an important cause of visual loss and disability, particularly in secondary progressive MS (SPMS). Optical coherence tomography (OCT) measurements of retinal nerve fiber layer (RNFL) thickness are included in recent MS trials as an exploratory outcome, allowing correlations of structure with visual function. We performed a visual outcomes and OCT sub-study for patients in Maestro-03 to examine the feasibility and potential usefulness of these measures in MS trials.

Methods:
Maestro-03 was a randomized, placebo-controlled study of a synthetic myelin based protein, dirucotide, in SPMS.* Thirty-five patients from 10 U.S. sites underwent OCT-3 RNFL thickness measurements at baseline, 3, 6, and 12 months. Low-contrast letter acuity (2.5%, 1.25% levels) and high-contrast visual acuity were assessed. Administration of study drug occurred at 6 month intervals; the trial was ended at 12 months due to lack of benefit in a similar European study.

Results:
Among 70 eyes, baseline RNFL thickness (84.3±14.2 μm) was similar to reported values for SPMS and lower than that of relapsing MS cohorts (92.5±16.7μm). Changes in RNFL thickness in this brief trial (follow-up 8.2±2.8 months) were minimal for treatment (n=40 eyes, RNFL change -0.21μm) and placebo (n=30 eyes, +1.12 μm, p=0.41). Visual function scores demonstrated changes in the direction of improvement for treatment with worsening in the placebo group; differences were significant for low-contrast acuity at 1.25% (treatment: +4.1 letters; placebo: -1.6 letters, p=0.02).

Conclusion:
This sub-study provides a first analysis of OCT measurements as a structural correlate to visual function tests in an MS clinical trial. Treatment studies of longer duration that target relapsing MS patients (greater RNFL thickness to start at baseline) will complement these feasibility data and provide larger-scale observations on changes over time in OCT and low-contrast acuity in the MS trial setting.

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Key Words: multiple sclerosis (MS), visual function, optical coherence tomography (OCT), clinical trials

Financial Disclosure: NONE

*non-FDA approved treatment for MS

Poster 26

Retinal Nerve Fiber Layer (RNFL) Thickness by Optical Coherence Tomography (OCT) in Multiple Sclerosis (MS): Relation to Visual Function Using Current vs. High-Resolution Technologies

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Introduction:
Retinal nerve fiber layer (RNFL) thickness by optical coherence tomography (OCT) correlates with new and established measures of visual function, including low-contrast letter acuity. OCT Fourier domain technology has afforded higher resolution (5-7 μm) and faster processing/scanning speeds, but how well these new methods provide measures that correlate with visual function remains to be determined. We compared RNFL thickness measurements by time-domain OCT to those of high-resolution spectral domain OCT with respect to their capacity to reflect visual dysfunction.

Methods:
Patients with MS participated in this study as part of an ongoing investigation of visual function and OCT in MS. RNFL thickness was measured for the average (360º around optic disc) using time domain and high-resolution spectral domain OCT. High-contrast visual acuity (VA) and low-contrast letter acuity at 2.5% and 1.25% contrast were assessed.

Results:
Data were analyzed for 121 patients with MS (242 eyes, age 45±12 years). RNFL thickness was 90.3±16.6 μm for time domain OCT and 83.5±14.0 μm for spectral domain OCT. Two-line (10-letter) differences in low-contrast acuity scores were associated with 6.0-6.2 μm decrements in RNFL by time domain OCT and 4.7-4.8 μm losses by spectral domain OCT ($p<0.001$ for both methods, accounting for age and within-patient inter-eye correlations). When RNFL thickness was converted to z-scores to account for differences in scaling between the two methods, patterns suggesting greater correlations for OCT-3 were again observed. Time domain and spectral domain OCT distinguished MS vs. control eyes equally well on the basis of RNFL thickness.

Conclusion:
High-resolution OCT and time domain OCT RNFL thickness differ with respect to correlations with visual function, yet demonstrate equal capacity to identify RNFL thinning in MS eyes (distinguish MS vs. controls). These results suggest that future clinical trials will need to account for differences in OCT technologies when interpreting structure-function correlations. Longitudinal studies and trials will further define the advantages of high-resolution/ Fourier domain OCT.

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Key Words: Multiple Sclerosis (MS), Optical Coherence Tomography (OCT), Visual Function

Financial Disclosure: NONE

Poster 27

Autosomal Dominant Optic Atrophy Shows A Significantly Thinner Macula OCT Compared To Other Optic Neuropathies

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Introduction:
We noticed that patients with a diagnosis of ADOA appeared to have thinned sectors on macula OCT compared to other optic neuropathies. We compared patients with ADOA, non-arteritic anterior ischemic optic neuropathy (NAION) and optic nerve hypoplasia (ONH) in order to study this in more detail.

Methods:
One eye of patients with a diagnosis of ADOA (n=5), NAION (n=8) and ONH (n=15) had a macula OCT performed. The 8 macula sectors from each eye were compared to the normative average thickness (microns). The average thickness of the 8 sectors was also calculated and compared between diagnostic groups by Analysis of Variance (ANOVA).

Results:
A significant thinning of the average macula thickness was found for ADOA (mean difference from normal=-45 microns) compared to NAION (difference=-24 microns) and ONH (difference=-16 microns) using ANOVA (p=0.034). Of the total number of macula sectors analyzed in each diagnostic group, the proportion of sectors that showed thinning at the 5th or 1st percentile was 72% of sectors for ADOA, 55% of sectors for NAION, and 30% of sectors for ONH eyes. We are currently segmenting the macula layers using a 3D software algorithm that we have developed to ascertain which layers of the retina are thinned in the eyes showing thinning of the total retinal thickness in the macula.

Conclusion:
ADOA shows greater total retinal thinning compared to NAION and ONH eyes and may provide yet another useful piece of information that can aid in the differential diagnosis in cases with uncertainty. This may also point towards a differential effect of this mitochondrial eye disease on the macula and appears to involve more than just the retinal nerve fiber layer. We hope to determine if any other retinal layers besides the RNFL and ganglion cell layers may be affected by ADOA.

References: NONE

Key Words: Autosomal Dominant Optic Atrophy (ADOA), Non-arteritic Anterior Ischemic Optic Neuropathy (NAION), Optic Nerve Hypoplasia (ONH), Macula OCT, Retinal Layers

Financial Disclosure: NONE

Poster 28

Maximizing the Reliability of Serial Time-Domain OCT Testing in Multiple Sclerosis Patients

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Introduction:
Optical coherence tomography (OCT) demonstrates thinning of the retinal nerve fiber layer (RNFL) in patients with multiple sclerosis (MS). I recently reported the results of serial testing of 142 eyes from a large group of patients with MS, identifying cutoffs for normally-distributed noise to distinguish pathological RNFL loss. Despite quality controls that eliminated over 40% of eligible eyes, cutoff values were up to about 30 microns for individual quadrant data. In the current study, I analyzed a subgroup with stricter criteria for image centering to minimize spurious intertest variation.

Methods:
I performed retrospective analysis of serial tests done an average of 13 months apart in patients from a large MS clinic, using a Zeiss-Meditec Stratus 3000 unit. Of 251 eligible eyes, 109 eyes were rejected by initial criteria for signal strength, artifact and scan centering. Intertest changes in RNFL thickness showed approximately normally distributed, zero-centered values consistent with noise, and strictly-negative outliers that I attributed to pathological RNFL loss. For the current study, I accepted only scan images with near-perfect optic disc centering and serial data traces that were superimposable. I also eliminated eyes showing significant RNFL loss between tests. These criteria yielded a subgroup of 69 eyes from 58 patients.

Results:
Strict centering criteria significantly reduced normally-distributed intertest changes from presumed noise for individual quadrant data. Initial and subgroup (in brackets) values in microns were as follows: mean RNFL thickness 8.5 (7.2), temporal quadrant 11 (11), nasal quadrant 30 (11), superior quadrant 25 (15) and inferior quadrant 29 (14).

Conclusion:
Meticulous attention to time-domain OCT scan quality, including image centering, significantly improves test-retest precision. With carefully monitored data, resolution may approach the putative limit of 7-10 microns for this technology, even for individual quadrant means. This level of accuracy may allow identification of subtle defects in individual patients over time.

References: NONE

Key Words: Optical coherence tomography, optic nerve, axonal loss, multiple sclerosis

Financial Disclosure: NONE

Poster 29

B-scan Ultrasound Estimates of Intracranial Pressure: Pediatric Emergency Room and ICU Method at 3mm behind the tip of the Optic Nerve Compared to the Neuro-Ophthalmology Method measured at 10mm behind the Sclero-dural Junction

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Introduction:
For 2 years pediatric emergency rooms and intensive care units have used B-scan ultrasound to estimate intracranial pressure (ICP). Articles by these physicians have proposed norms for optic nerve diameter vs. age. For over 25 years, Neuro-Ophthalmologists have also used B-scan ultrasound to diagnose and follow nerve and orbit pathology. In patients with headache, it is a non-invasive screening technique helpful to see which patients without obvious papilledema need lumbar puncture. This study compares the two techniques for consistency in predicting ICP.

Methods:
Over 1000 consecutive eyes were scanned using both techniques: (1) the pediatric technique measuring the dura to dura optic nerve diameter 3mm behind the tip of the nerve, and (2) the neuro-ophthalmology technique measuring the diameter 10mm behind the sclero-dural junction. Their conclusions of “normal” or “abnormal” were compared with age, ICP measurements, surgical management, and other pathology (e.g. drusen).

Results:
There was 78% agreement between the two methods in predicting normal or elevated ICP. The most common pattern of disagreement was for the anterior pediatric measurement to be smaller (predicting normal ICP) than the posterior neuro-ophthalmology measurement (predicting abnormal high ICP). This was seen in 19% of the eyes. The remaining 3% of the eyes scanned had wider anterior measurements than posterior measurements.

Conclusion:
There is a 78% agreement rate between the two B-scan ultrasound measurement techniques in predicting normal or raised ICP. The pediatric anterior measurement was falsely “normal” in people with prior optic nerve sheath decompression and some people with no visible papilledema. The 10mm posterior measurements capture those 19% false-negatives in people with prior optic nerve sheath decompression surgery, no visible papilledema, but in whom there is still raised ICP. This also helpful in assessing ICP after neurosurgical shunt surgeries when there is no anterior swelling on clinical exam.

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Key Words: NONE

Financial Disclosure: NONE

Poster 30

Feasibility of Non-Mydriatic Fundus Photography in the Emergency Department (ED)

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Introduction:
Visualization of the ocular fundus remains a critical part of the screening physical examination for many patients in the ED. Non-mydriatic fundus photography offers a promising alternative to direct ophthalmoscopy in the ED setting. Our objective was to determine the practical feasibility of non-mydriatic fundus photography in the ED.

Methods:
Interim analysis of 205/350 planned, consecutive patients seen in a university hospital ED who had non-mydriatic photography of both eyes obtained by nurse practitioners (NPs) during their usual ED shift. Nurses rated their impression of the ease and speed of obtaining photographs on each patient on a 10 point Likert scale (10 best). Patients similarly rated their impression of the comfort, ease, and speed of having photography taken. Time of arrival, clinical evaluation, photographs, and discharge were recorded.

Results:
The mean rating was 8.7/10 for nurse ease and 8.7/10 for speed. The nurses rated 58% of encounters 10/10 for ease and 58% 10/10 for speed. The mean rating of patients was 8.7/10 for comfort, 9.3/10 for ease, and 8.9/10 for speed. 53% of patients rated their photo session 10/10 for comfort, 73% for ease, and 65% for speed. Median ED length of stay was 7.1 hours (interquartile range[IQR]: 4.3-13.9 hours). Median photography session length was 2.3 minutes (IQR: 1.5-3.5 minutes), typically accounting for 0.5% of the patient's ED total visit. 29% of photographs were taken during the first quarter of a patient's ED visit and 12% were taken before the patient was evaluated by a clinician.

Conclusion:
NPs and patients are generally very pleased with the ease, comfort, and speed of non-mydriatic fundus photography ED, and typical photography sessions represent a trivial portion of a patient's overall ED visit. If proven clinically valuable, non-mydriatic fundus photography could be implemented within the ED workflow without adding a significant burden to nurses or patients.

References: NONE

Key Words: non-mydriatic photography, emergency department, feasibility, telemedicine

Financial Disclosure: NONE

Poster 31

Quality of Non-Mydriatic Fundus Photos Taken by Nurse Practitioners (NPs) in the Emergency Department (ED)

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Introduction:
There is ample evidence that fundus photography with pupillary dilation is a useful screening tool in ophthalmology. However, pupillary dilation is impractical in the ED. Non-mydriatic photography offers an alternative approach but has not been applied to the ED setting and is more challenging. Our objective was to determine if the quality of non-mydriatic fundus photography taken by NPs is sufficient for screening of neuro-ophthalmic emergencies in the ED.

Methods:
Interim analysis of 193/350 consecutive planned patients photographed by NPs during their usual ED shift. Nurses received less than 30 minutes of fundus photography training. Photographs were taken of both eyes of each patient in a triage area in ambient or partially obscured lighting conditions. The photographs were reviewed by two neuro-ophthalmologists. Quality was graded from 1 to 5 (1: inadequate for any diagnostic purpose, 2: unable to exclude all emergent findings, 3: only able to exclude emergent findings, 4: not ideal but still able to exclude subtle findings, 5: ideal quality). Four regions were independently evaluated for quality: (1) optic disc, (2) macula, (3) superior and inferior (4) vascular arcades. Primary outcome measures were the frequency of high quality images (grade 4 or 5) and inter-observer agreement.

Results:
165/193 (85%) had at least one eye with a high quality photograph, 120 (62%) had high quality pictures of both eyes, and 3 patients (2%) had no photograph of diagnostic value. The inter-observer agreement among the 2 neuro-ophthalmologists was very good (kappa 0.89 [95%CI:0.81-0.98]). Agreement between the NPs and the neuro-ophthalmologists was moderate (kappa 0.43 [0.32-0.55]). Quality of optic disc region (1) was better than the quality of the other retina regions (2, 3 and 4).

Conclusion:
The quality of non-mydriatic fundus photography obtained by NPs in a clinical ED setting is adequate to assess neuro-ophthalmic emergencies, particularly changes of the optic disc.

References: NONE

Key Words: non-mydriatic photography, emergency department, quality, telemedicine

Financial Disclosure: NONE

Poster 32

Intraobserver Reliability of Non-Mydriatic Fundus Photography Quality Rated on a Desktop Computer vs. Smartphone

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Introduction:
The increased usage of smartphones (mobile phones with advanced capabilities) has made possible the use of these devices for telemedicine. While personal digital assistants and mobile phones have been already studied as a remote interpretation medium in radiology, dermatology, and neurology, this technique has not yet been applied to ophthalmology. Remote interpretation of funduscopic images for emergency department (ED) consultation regarding patients presenting with neuro-ophthalmic complaints is a potential application of these devices. Our objective was to determine whether the quality of non-mydriatic fundus photography displayed on a smartphone is similar to that on a desktop computer monitor.

Methods:
100 non-mydriatic fundus photographs were randomly selected from the 970 pictures taken from 193 consecutive patients photographed during their ED evaluation. All photographs were reviewed by a neuro-ophthalmologist using a standard computer screen and again, two months later, using a smartphone (Apple iPhone). Quality was graded from 1 to 5 (1: inadequate for any diagnostic purpose, 2: unable to exclude all emergent findings, 3: only able to exclude emergent findings, 4: not ideal but still able to exclude subtle findings, 5: ideal quality). Primary outcome measure was the intra-observer agreement.

Results:
Among the 100 photos used for this study when rated on the desktop monitor screen, 29 were grade 1; 13, grade 2; 13, grade 3; 19, grade 4; and 26, grade 5. On the smartphone screen; 19 were grade 1; 16, grade 2; 11, grade 3; 19, grade 4; and 35, grade 5. The intra-observer agreement was very good (kappa 0.86 [95%CI: 0.60-1.00]).

Conclusion:
The quality ratings of non-mydriatic fundus photography obtained in a clinical ED setting are similar regardless of whether they are viewed on a standard computer monitor or on a smartphone screen.

References: NONE

Key Words: non-mydriatic photography, emergency department, smartphone, quality, telemedicine

Financial Disclosure: NONE

Poster 33

Occult Macular Dystrophy As A Cause Of Unexplained Vision Loss

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Introduction:
Reduced visual acuity or visual field abnormalities in the setting of a normal papillary and funduscopic examination frequently leads to a diagnosis of functional vision loss. However, with mfERG and OCT testing showing foveal photoreceptor loss or dysfunction, occult macular dystrophy (OMD) has been identified as an organic cause of visual loss. We present 8 cases of OMD and describe the clinical, electrophysiological and OCT characteristics of this rare disorder.

Methods:
Ten patients (19 eyes) ages 16 to 62 years with complaints of visual loss were examined. Visual acuity, color vision, Goldmann and Humphrey visual fields, mfERG, full-field ERG, fundus examination, fundus photos, and OCT were obtained. mfERG was recorded as a scaled array of 103 hexagons. Amplitude and implicit time of six concentric rings were compared to normal controls (n=15). Linear regression was used to determine correlation between foveal thinning or mfERG P1 amplitude and visual acuity.

Results:
Visual acuity ranged from 20/20 to 20/400. Color vision was normal to severely affected. GVFs were normal in 4 patients and 4 patients had central or paracentral scotomas. Full field ERGs were normal in all patients. Fundus examinations revealed normal optic nerve appearance without macular or peripheral retinal changes in all eyes. In some patients, OCTs showed thinning of the fovea. mfERGs showed significant amplitude reduction centrally ($P<0.0001$) and prolongation of implicit times ($p<0.0001$). There was no correlation between foveal thinning or mfERG P1 amplitude and visual acuity.

Conclusion:
OMD is a condition characterized by subnormal acuity, central scotoma, variable decrease in color vision, normal funduscopic appearance, and variable progression over time. Full field ERG is normal, but mfERG is abnormal in all patients. OCT may show thinning of the fovea. The severity of foveal thinning and mfERG amplitude loss does not correlate with visual acuity. Foveal dysfunction and/or photoreceptor loss are likely causes of vision loss in OMD.

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Key Words: occult macular dystrophy, optical coherence tomography, multifocal electroretinography, vision loss

Financial Disclosure: NONE

Poster 34

Vascular Risk Factors for Central Retinal Artery Occlusion

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Introduction:
The aim of this study is to determine the proportion of patients presenting with thromboembolic central retinal artery occlusion (CRAO) who had undiagnosed vascular risk factors amenable to modification.

Methods:
A retrospective audit of consecutive patients with non-arteritic/thromboembolic CRAO presenting between 1997 and 2008 in a single tertiary teaching hospital.

Results:
Thirty-three patients with non-arteritic CRAO were identified. Twenty-one patients (64%) had at least one new vascular risk factor found after the retinal occlusive event, with hyperlipidemia being the most common (36%). To better control their vascular risk factors 18 patients (54%) were given a new or altered medication. Nine patients had more than 50% of ipsilateral carotid stenosis; six of these proceeded with carotid endarterectomy or stenting. One patient had significant new echocardiogram finding. Systemic ischaemic event post CRAO occurred in two patients with stroke and acute coronary syndrome.

Conclusion:
Patients presenting with CRAO often have a previously undiagnosed vascular risk factor that may be amenable to medical or surgical treatment. As this population is at a high risk of secondary ischaemic events, risk factor modification is prudent.

References: NONE

Key Words: Central retinal artery occlusion, Vascular risk factors, Morbidity and Mortality

Financial Disclosure: NONE

Poster 35

A Prospective Case-Control Study Of Retinal Hemorrhages In Infancy

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Introduction:
Retinal hemorrhages (RH) are a common finding in pediatric abusive head trauma (AHT), and severe forms of RH are generally thought to be indicative of an acceleration-deceleration injury. However, few studies have compared the retinal findings in AHT with other conditions considered high risk for RH. Further, the conditions under which RH occur and the underlying mechanisms have not been entirely delineated.

Methods:
This is a prospective, consecutive, multicentre case-control study. Infants and children (4 weeks – 3 years) admitted with head injury, major trauma, acutely raised intracranial pressure (ICP), acutely raised intrathoracic pressure (CPR, trauma), and hypoxia (choking, apnoea) are eligible for inclusion.

During their admission, patients receive a dilated eye examination, recorded with the RetCam. Retinal images are later reported by two experts, masked to the mode of injury.

Results:
Of 58 head injuries, 6 had no clinical or radiological evidence of direct impact. These cases had a significantly higher rate of RH than head injury with impact (66% v.s. 4%, p = 0.001). Of the cases of non-impact head injury 5 were due to AHT and 80% of these (4/5) had RH. These were all bilateral, affecting multiple layers and extending beyond the posterior pole. RH due to impact head injury were mild and unilateral.

Of the patients with raised ICP, 27% (3/11 cases) had RH. In the raised intrathoracic pressure cohort 6% (1/18 cases) had RH. No cases in the hypoxia cohort (8 cases) demonstrated RH.

Conclusion:
Severe, bilateral RH appear to be a marker for AHT in the absence of a recognized medical condition. Raised intrathoracic pressure and hypoxia do not appear to be major contributing factors to RH. Raised ICP has demonstrated some degree of retinal involvement.

References: NONE

Key Words: retinal hemorrhages, abusive head trauma, raised intracranial pressure, raised intrathoracic pressure, hypoxia

Financial Disclosure: NONE

Poster 36

Retinal Arteriolar Occlusions and Multifocal Areas of MRI Restricted Diffusion Indicate Infarction in Susac's Syndrome

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Introduction:
The differential diagnosis of multifocal high signal abnormalities on MRI in a patient with acute encephalopathy can be challenging. The finding of restricted diffusion suggests but does not prove infarction rather than demyelination. The finding of multifocal retinal arteriolar occlusions adds support to infarction as the underlying mechanism. We describe a patient in whom the combination of MRI and retinal findings allowed Susac's syndrome to be distinguished from multiple sclerosis. Although our findings have been previously described, we provide an elegant correlation.

Methods:
Single case report

Results:
A 22-year-old woman presented with headache, drowsiness, confusion, ataxia, tinnitus, and hearing loss. Brain MRI revealed multiple T2 hyperintensities involving the corpus callosum, basal ganglia, and thalamus with restricted diffusion. Lumbar puncture showed only elevated protein. Atypical multiple sclerosis was the presumed diagnosis. She was treated with IV methylprednisolone with temporary clinical improvement. When symptoms worsened, repeat MRI revealed additional lesions, many with restricted diffusion. The patient had no visual complaints, but ophthalmoscopy revealed cotton wool spots, small intraretinal hemorrhages, and areas of retinal whitening. Audiometry revealed sensorineural hearing loss binaurally. These findings were strongly consistent with Susac's syndrome. Plasmapheresis, aspirin and azathioprine were added to the corticosteroid regimen. The patient's clinical status gradually improved but she remained partially deaf and cognitively impaired.

Conclusion:
The retinal findings in this patient contributed to the recognition that she did not have multiple sclerosis, but rather Susac's syndrome, a microangiopathy of the brain, retina and inner ear. This distinction is important as the two conditions are treated differently. Although the efficacy of treatment in Susac's syndrome has not been rigorously verified, the standard approach includes a regimen of immunosuppressive agents combined with immunomodulatory and/or antithrombotic therapies.

References: NONE

Key Words: Susac's syndrome, branch retinal artery occlusion, multiple sclerosis, restricted diffusion

Financial Disclosure: NONE

Poster 37

Ocular Neo-Vascularization Following Central Retinal Artery Occlusion

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Introduction:
Considerable debate exists within the literature as to the incidence and etiology of neo-vascularization following central retinal artery occlusion (CRAO). A large retrospective study described neo-vascularization of the iris (NVI) in 2.5% of patients with CRAO, whereas several smaller prospective and retrospective studies have reported an incidence of 16.6-18.2%.[1-4]

We conducted a retrospective study to determine the incidence of ocular neo-vascularization following acute CRAO in our institution.

Methods:
A retrospective audit of consecutive patients with non-arteritic/thromboembolic CRAO presenting between 2000 and 2008 in a single tertiary teaching hospital.

Results:
Thirty patients were identified as having non-arteritic CRAO, and of this cohort 6 patients (20%) developed ocular vascularization. Neo-vascularization of the iris (NVI) occurred in 5 cases; rubeotic glaucoma occurred in 4 cases (mean peak intra-ocular pressure = 46mmHg). Neo-vascularization at the disc (NVD) occurred in 4 cases. Mean time from retinal occlusive event to observed neo-vascularization was 58 (± 32) days. In 2 cases of neo-vascularization there was haemodynamically significant (>70%) ipsilateral carotid stenosis.

Conclusion:
The incidence of NVI following acute CRAO in our population was 16.7% - a rate comparable with several other studies. The rate of NVD, however, was considerably higher than in other reports.

In the majority of our cases, neo-vascularisation occurred in the absence of a haemodynamically significant (>70%) carotid artery stenosis. We thus have insufficient evidence to conclude that this was a result of ocular ischaemia.

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Key Words: Central retinal artery occlusion, Neo-vascularization, Glaucoma

Financial Disclosure: NONE

Poster 38

“Tonic” but not “Adie’s” Pupil

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Introduction:
Today a tonic pupil—a non-syphilitic pupil that reacts poorly to light and slowly redilates-- is often labelled an “Adie’s pupil,” and the symptom complex of a tonic pupil with absent deep tendon reflexes as “Adie’s syndrome.” These eponyms give credit to William John Adie, an Australian born neurologist who practiced in Edinburgh in the early 1900s. To determine whether these attributions are historically correct, we investigated Adie’s original manuscripts.

Methods:
Adie’s original papers and a comprehensive review on the tonic pupil by Lowenstein and Lowenfeld were analyzed. Relevant papers referenced by those authors were also reviewed.

Results:
Saenger (1902), Strasburger (1902) and other physicians publishing before Adie (1931) described a non-luetic pupil with abnormally long-lasting pupillary constriction to the light reflex, or absent light reflex, paired with slow, extensive contraction to accommodation and sluggish redilation upon returning to far vision. In his first paper, Adie presented six patients each with a pupil that accommodated but did not react to light and absent tendon reflexes. In his second and third papers, Adie described a benign syndrome that in its complete form consisted of a tonic pupil and absent tendon reflexes. He also described four incomplete forms of this syndrome, one of which was a tonic pupil alone. In his three papers, Adie did not describe any new features of the tonic pupil.

Conclusion:
While our current use of the term “Adie’s Pupil” is technically not wrong—as it is a form of Adie’s Syndrome—a close reading of Adie’s publications shows that it does not do justice to the historical record. We believe that the abnormal pupil should be referred to as a “tonic pupil” rather than an “Adie’s pupil” and the eponym should be left for “Adie’s syndrome”.

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Key Words: Adie, Tonic, Pupil

Financial Disclosure: NONE

Poster 40

Patient Interviews and Focus Groups to Determine Outcome Measures for Idiopathic Intracranial Hypertension (IIH)

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Introduction:
To assess outcome measures for IIH, in order to develop a composite scale for assessing disease disability.

Methods:
Subjects with IIH (n=51) were identified by their neuro-ophthalmologist and recruited by letter or at their office visit from two sites in New York and one in Texas. 10 patients elected to participate. Semi-structured telephone interviews (n=4) and focus groups (n=6) were conducted to assess the content domains of the Beck Depression Inventory (BDI), VFQ-25, Patient Health Questionnaire, MIDAS, and HIT-6 scales for subjects with IIH. Open-ended questions were used to collect information on individuals' experiences with IIH and its effects on their lives.

Results:
Disease duration was 2-18 years (mean 10.5 years). Less than 70% of subjects endorsed two of the questions on the BDI, 6 questions on the VFQ-25, and one question on the MIDAS as being as relevant to IIH. 80% supported using HIT-6 over MIDAS for assessing headache disability. Participants noted that neither diplopia nor photosensitivity was adequately addressed in the VFQ-25. IIH negatively impacted subjects' relationships (40%), mood (70%), finances (60%), and work (50%). 70% of the subjects felt that their physicians blamed them for having IIH and only 20% received educational information on IIH from their physicians.

Conclusion:
This analysis suggests that while current headache, depression, health, and vision scales are relevant for assessing some areas of disability in IIH, other issues such as financial burden, effect on relationships and some visual symptoms are not adequately addressed. Larger numbers of subjects representing greater geographic diversity would enhance the reliability of our results. These data will be utilized with ongoing diary studies to develop and validate a composite outcome measure for IIH to use in clinical research.

References: NONE

Key Words: idiopathic intracranial hypertension, quality of life, Beck Depression Inventory, Headache disability, VFQ-25

Financial Disclosure: NONE

Poster 41

The Effect of Translaminar Pressure on Visual Field Outcomes in Pseudotumor Cerebri

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Introduction:
Pseudotumor cerebri (PTC) is a disorder of unknown etiology which results in elevated intracranial pressure (ICP). The optic disc is basically exposed to 2 pressurized compartments, the intraocular space and the subarachnoid space, which are separated by the lamina cribosa. The pressure difference between these compartments is known as the translaminar pressure difference. Recent studies have demonstrated that decreased ICP is associated with increased cup to disc ratio in patients with glaucoma (elevated translaminar pressure). Thus, it is our hypothesis that a worse visual outcome in PTC could also be associated with increased translaminar pressure in the other direction as a result of increased CSF pressure and/or decreased IOP. To our knowledge, there has been no study in PTC patients to determine whether translaminar pressure is related to visual field outcome. The objective of this study was to examine the relationship, if any, between translaminar pressure and visual field outcomes in PTC.

Methods:
Retrospective chart review was performed to examine the relationship between translaminar pressure in patients newly diagnosed with PTC at one institution and visual outcome in the form of Humphrey visual fields (HVF). Translaminar pressure was calculated according to the following formula: translaminar pressure (mm Hg) = ICP – IOP. All visual field data was collected including mean deviation (MD) and pattern standard deviation (PSD). Initial translaminar pressure was correlated to final visual field outcome (MD and PSD).

Results:
Data was collected on 60 eyes of 30 female patients with PTC. Follow-up ranged from 3 to 100 months with a mean of 25.4 months. Significant predictors of a worse HVF outcome (MD and PSD) were initial HVF MD and PSD ($p<.0001$), opening pressure ($p<.0001$), and translaminar pressure ($p=.0002$).

Conclusion:
There was a statistically significant correlation between translaminar pressure and visual field outcome in patients with pseudotumor cerebri.

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Key Words: Pseudotumor cerebri, Idiopathic intracranial hypertension, Translaminar pressure, Opening pressure, Visual field

Financial Disclosure: NONE

Poster 42

Idiopathic Intracranial Hypertension: A Canadian Tertiary Care Centre's Experience

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Introduction:
Idiopathic intracranial hypertension (IIH) is a syndrome of raised intracranial pressure for which no structural causative factor can be identified. As an initial study towards the larger goal of more rigorously evaluating therapies for IIH, the diagnosis, treatment and outcome of patients with IIH at a single Canadian tertiary care centre were reviewed.

Methods:
The terms "pseudotumour cerebri", "benign" or "idiopathic" "intracranial hypertension", "papilledema" or "optic neuropathy" were used to search the hospital patient registry between January 2002 and December 2008. Charts were reviewed and all patients diagnosed with IIH were included in the study.

Results:
Fifty-one patients with IIH were identified. As expected, the majority of patients were female (90%), of child-bearing age (31 years [14-64]) and overweight (mean 33 kg/m^2 [18-58]). Mean symptom duration prior to diagnosis was 14 weeks. Headache (82%) and visual blurring or loss (77%) were the most frequent symptoms at presentation. Hypothyroidism (21%) and obstructive sleep apnea (17%) were common co-morbid conditions. Patients over 40 years old were more likely to have sleep apnea and hypothyroidism and less likely to have headache at presentation. Recent minocycline or tetracycline use was identified in six patients, all under 40 years of age. Magnetic resonance brain imaging and venograms were available for review in 29 patients. Bilateral severe transverse sinus stenoses (75%), flattening of the posterior aspect of the globes (62%) and distension of the peri-optic nerve sheath (59%) were the most common neuro-imaging findings. Twenty one patients (41%) required surgical intervention due to progressive or severe visual loss and/or headache.

Conclusion:
Clinical presentation and co-morbid conditions distinguish older from younger patients with IIH. A high proportion of patients in this series required surgical intervention, likely reflecting referral bias of patients with more severe or refractory symptoms to this tertiary care centre.

References: NONE

Key Words: idiopathic intracranial hypertension, cohort study

Financial Disclosure: NONE

Poster 43

IDIOPATHIC INTRACRANIAL HYPERTENSION (IHH) IN APLASTIC ANEMIA

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Introduction:
IHH has been described in patients with different types of anemia: ferropenic, pernicious, hemolytic, paroxysmal nocturnal hemoglobinuria and aplastic anemia.
IHH is a very infrequent complication of acquired aplastic anemia, and few cases are described.

Methods:
Case presentation and review of relevant literature

Results:
A non obese 21-year-old woman developed headache, blurred vision and diplopia in the context of aplastic anemia. Visual acuity was 20/20 in both eyes. A fundoscopic examination revealed bilateral papilledema. A right sixth nerve palsy was present. Automated perimetry showed bilateral blind spot enlargement. Cerebral magnetic resonance imaging and venography -MRI revealed no abnormalities. A lumbar puncture showed an opening pressure of 350 mm Hg. Microbiological and cytological study of CSF was normal.
At that moment, she was receiving red blood transfusion and immunosuppressive therapy with cyclosporine. The cyclosporine was discontinued because persistence of papilledema and she was placed on antithymocytic-globulin. Because the persistence of clinical signs, she initiated treatment with acetazolamide 500 mg TID. After negative response to the conservative aplastic anemia treatment, she received bone marrow transplantation with good clinical results.
Acetazolamide was discontinued several weeks after transplantation because resolution of symptoms and signs of IHH.

Conclusion:
IHH has been described in patients with different types of anemia.
It has been reported that this association in aplastic anemia not always respond to corrective therapy but to IHH management.
In these patients with immunosupressive treatment, it has to be considered the role of cyclosporine, agent associated with intracranial hypertension, and it may be discontinued if papilledema persistence. Other causes have to be ruled-out as infectious diseases.
Papilledema must be ruled out in patients with aplastic anemia. On the other side, it is advisable to obtain a complete blood count in patients with idiopathic intracranial hypertension because of the possible association with anemia.

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Key Words: papilledema, peripapillary hemorrhages, intracranial-hypertension, anemia, aplastic

Financial Disclosure: NONE

Poster 44

Non-invasive Assessment of Intracranial Pressure Using Otoacoustic Emissions in Patients with Idiopathic Intracranial Hypertension

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Introduction:
Early treatment of elevated intracranial pressure (ICP) and the ability to follow response to treatment are crucial factors in the preservation of the optic nerve and visual function. Current methods of ICP monitoring require invasive entry into the skull or spinal cord. Distortion-product otoacoustic emissions (DPOAEs), measured non-invasively through the ear canal, appear to be correlated with ICP.[1] We sought to determine whether DPOAEs may be utilized as a reflection of elevated ICP in idiopathic intracranial hypertension (IIH).

Methods:
DPOAE magnitudes for a set of frequencies (range 516-3984 Hz), DPOAE angles, power reflectance, and transmittance were measured in 5 patients undergoing a medically necessary diagnostic lumbar puncture (LP). Normal hearing was confirmed by otoscopy and a tympanogram prior to data collection. Measurements were obtained before and after the LP using software from Mimosa Acoustics (HearID v3.1). Data including age, opening and closing ICPs, time of day were also recorded.

Results:
Three patients with IIH and two normal subjects were evaluated. Mean opening and closing pressures in IIH patients were 36 cm H_20 and 22 cm H_20, respectively; in controls, pressures were 18 cm H_20 and 12 cm H_20, respectively. DPOAE magnitudes were appreciably different between IIH and control patients for the frequencies recorded; low-frequency range measurements (< 1172 Hz) were more difficult to record and were confounded by background noise. The absolute difference between opening and closing pressure did not appear to be as important as the degree of initial ICP elevation.

Conclusion:
DPOAEs may offer a promising non-invasive alternative for monitoring ICP in normal hearing patients. Although measurements cannot specifically predict the actual ICP value, DPOAEs appear to reflect elevation in ICP. These preliminary findings provide a basis for more extensive exploration to determine the utility of DPOAE measurements in longitudinal monitoring of patients with IIH.

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Key Words: measurements cannot idiopathic intracranical hypertension, papilledman, ICP monitoring, otoacoustic emissions

Financial Disclosure: NONE

Poster 45

Weight Gain and Recurrence in Idiopathic Intracranial Hypertension: An Expanded Case Control Study

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Introduction:
Obesity and weight-gain are established risk factors for idiopathic intracranial hypertension (IIH). The association of weight gain with IIH recurrence has not been established. This study's purpose was to determine frequency and degree of weight gain associated with IIH recurrence and to test the hypothesis that patients with IIH who develop recurrence has a greater percent change in weight between initial resolution and recurrence than do IIH patients without recurrence (controls).

Methods:
We performed a case-control study of adult patients who fulfilled established clinical criteria for IIH within the neuro-ophthalmic practices at two academic institutions. Twenty-six patients fulfilled inclusion criteria for recurrence of IIH and 26 controls met IIH criteria without recurrence. Body mass index (BMI) was calculated at IIH initial presentation, resolution, and at time of recurrence; recurrence BMI's for controls were based on weight measurements performed at a time interval from resolution that was the same as the average period between resolution and recurrence in the patient group. Changes in BMI and percent changes in weight were compared between groups using the Wilcoxon rank-sum test.

Results:
In IIH recurrence patients (n=26, 100% female, age 33±11 years), BMI's at recurrence were significantly greater than those at resolution (p=0.0009, signed-rank test) and presentation (p=0.007). Patients with IIH recurrence demonstrated a greater degree of weight gain between resolution and recurrence (median BMI change +1.9 kg/m^2 [-1.53-10.8]) compared with controls (+0.18 kg/m^2 [-20.3-3.6], p=0.03, rank-sum test). On the basis of percent weight change, controls had stable weights between resolution and recurrence (0.18%± 5 loss), while IIH recurrence patients demonstrated a 5.6% weight gain (p=0.01). This corresponded to an average rate of BMI gain of 1.3 kg/m^2/year for recurrence patients, compared with -0.21 kg/m^2/year for controls.

Conclusion:
IIH recurrence was associated with significant gains of weight and BMI compared to IIH controls without recurrence in this cohort. Patients with resolved IIH should be advised that weight gain may be associated with IIH recurrence.

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Key Words: Idiopathic Intracranial Hypertension, Recurrence, Weight Gain

Financial Disclosure: NONE

Poster 46

A Big Problem: IgA Nephropathy Presenting as Idiopathic Intracranial Hypertension in a 15 Year Old

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Introduction:
When women in the child bearing years present with headache, visual loss and nerve fiber layer edema, a diagnosis of idiopathic intracranial hypertension is often entertained. We present a case of an obese 15 year old girl presenting with 3 week history of severe headache and progressive visual loss. This case emphasizes the differential diagnosis of nerve fiber layer edema associated with normal imaging and increased intracranial pressure.

Methods:
A single retrospective case report of clinical findings, neuro-imaging, laboratory studies and histopathological confirmation of diagnosis.

Results:
The patient developed headache with associated nausea and vomiting three weeks prior to presentation. Her eye doctor gave a prescription for a kit containing low-dose doxycycline (Alodox) and Tranquileyes goggles. She denied transient visual obscurations, pulsatile tinnitus or double vision.

Visual acuity was 20/400 and 20/150. Visual fields were constricted. Pupils were pharmacologically dilated. Motility was full OU. Profound bilateral nerve fiber layer edema with subretinal fluid was noted. She was alert and oriented x 4. Vitals included blood pressure 145/99, pulse 125, respiratory rate 24, weight 108 kg, temperature 98. Lumbar puncture opening pressure was 52 cm water. Blood count showed hemoglobin of 7.5 g/dl (nl 12.0 -16.0) and platelets of 88 thous/uL (nl 150-400). Metabolic profile showed creatinine of 5.94 mg/dl (nl <1.11) and blood urea nitrogen of 40 mg/dl (nl 5-25). Urinalysis revealed 4+ protein. A lumbar drain was placed. Optic nerve sheath fenestration was performed. A renal biopsy confirmed IgA nephropathy. Vision recovered to 20/30 OD and 20/60 OS.

Conclusion:
IgA nephropathy is the most common cause of glomerulonephritis. Prompt diagnosis can be organ saving. Over a third of IgA nephropathy patients present with hypertension and 15% of these patients present with malignant hypertension. Assessment of renal function is a critical component to the evaluation of a patient with nerve fiber layer edema.

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Key Words: Vision Loss, Optic Nerve, Increased Intracranial Pressure, Optic Nerve Sheath Fenestration, Systemic Hypertension

Financial Disclosure: NONE

Poster 47

Transverse Sinus Stenting As Primary Intervention In Fulminant Idiopathic Intracranial Hypertension

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Introduction:
A 24 yo severely obese woman (BMI 61.5) with unremarkable past medical and ocular histories presented with headaches and pulsatile tinnitus of one month duration. More recently she had developed horizontal diplopia and blurring of vision OS. Acuities measured 20/20- OD and 20/40 OS. Goldmann visual fields were severely constricted, worse on the left. CSF composition, opening pressure and neuroimaging were compatible with the diagnosis of idiopathic intracranial hypertension (IIH).

Methods:
The MRV suggested pronounced bilateral segmental transverse sinus (TS) narrowing. Venography demonstrated a 39mmHg pressure differential across the right TS and 43mmHg across the left TS. Stenting of the left (90%-narrowed) TS dropped the pressure gradient to 5mmHg on the left side. Interestingly enough, the right TS narrowing had resolved spontaneously by the end of the procedure during which only the left TS was stented.

Results:
The patient's symptoms improved nearly instantaneously, followed by more gradual visual field expansion and resolution of papilledema. Symptomatic control was maintained at 8 months follow-up. The average peripapillary retinal nerve fiber layer thickness measured 62 microns OD and 56 microns OS, 6 months after intervention.

Conclusion:
TS stenting can be successfully applied as primary intervention in sight threatening IIH. Unilateral stenting of a narrowed TS and secondary intracranial pressure (ICP) normalization can also resolve compressive TS flow obstruction on the opposite side. This mirrors the effects of ICP lowering on TS diameter achieved by lumbar puncture or shunt placement.

References: NONE

Key Words: idiopathic intracranial hypertension, dural venous sinus stenosis, dural venous sinus stenting

Financial Disclosure: NONE

Poster 48

Bilateral Abduction Deficits in Idiopathic Intracranial Hypertension and Improvement with Acetazolamide Treatment: An Infrared Oculography Study

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Introduction:
There has been no clinical trial to evaluate the efficacy of acetazolamide treatment for idiopathic intracranial hypertension (IIH). In this prospective study, we used high speed eye movement recording to characterize reflexive saccades in patients with IIH and to assess the effectiveness of acetazolamide.

Methods:
We recorded 25 consecutive patients referred for evaluation of IIH with 500-Hz binocular infrared oculography. Eighten patients fulfilled the Modified Dandy criteria for IIH[1,2,3,4], and data from 16 patients with good recordings (10 paired recordings performed before and on acetazolamide) were analyzed. Stimuli included reflexive centrifugal saccades. Each eye was calibrated independently, and data were analyzed with MATLAB and IgorPro.

Results:
In 16 IIH patients (15 women/1 man, age 39.8±3.2, body mass index 35.4±1.9), symptoms included headache (100%), vision loss (56%), tinnitus (69%), and diplopia (19%). All patients had visual acuity of 20/20 bilaterally, visual field loss grade ≤2[5], and papilledema. Alignment was 63% orthophoric and 31% esophoric. On infrared oculography, 90% of patients with paired recordings (n=10) had bilateral abduction deficits. In horizontal saccades, plateau amplitude showed 0.8±0.2 deg abduction deficit (center-to-left: OD 11.9 deg, OS 11.1 deg, P=0.006; center-to-right: OD 11.2 deg, OS 12.0 deg, P=0.01). Data from all patients (non-paired) also showed significant bilateral abduction deficits (center-to-left: P=0.02, center-to-right: P=0.005). Despite this abducton deficit in saccade amplitude, there was no difference in peak velocity. There was also no binocular mismatch in vertical saccades (center-to-up: P=1.0; center-to-down: P=0.9). Acetazolamide treatment eliminated this bilateral abduction deficit (center-to-left: P=0.02; center-to-right: P=0.03), with no effects on vertical saccades (center-to-up, P=0.5; center-to-down, P=0.5) or peak velocities in all directions.

Conclusion:
Despite the lack of symptomatic binocular diplopia and cranial nerve VI palsy, 90% of patients with IIH exhibited bilateral abduction deficits during reflexive saccades. This difference disappeared after acetazolamide treatment, along with symptomatic improvement of headache, vision loss, and diplopia.

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Key Words: idiopathic intracranial hypertension, saccade, eye movement, acetazolamide, papilledema

Financial Disclosure: NONE

Poster 49

Susac's Syndrome in a 27-Year Old Female: The First Case Report in Iran

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Introduction:
Susac's syndrome is an extremely rare neurological disorder with involving three systems: encephalopathy, branch retinal artery occlusion, and hearing loss. The mechanism of it is microangiopathy serving the brain, the retina and cochlea. This is the first report of this syndrome in Iran.

Methods:
A 27-year-old woman was referred by the neurologist, for ophthalmic examination. She had a history of headache, visual loss in her right eye, four-limb paresthesia and behavioral changes from 10 months ago. She also complained of tinnitus and hearing loss for 2 weeks.

Results:
The patient was initially diagnosed with multiple sclerosis, but auditory and retinal involvement raised the possibility of Susac's syndrome. Magnetic resonance imaging, audiometric tests, and retinal fluorescein angiography were performed (figure1, 2).

Conclusion:
Susac's syndrome is a rare disorder of unknown origin characterized by the triad of encephalopathy, hearing loss, and visual loss. This finding showed that patients with encephalopathy and hearing loss should have a dilated examination of their retina for occlusions of retinal artery branches.

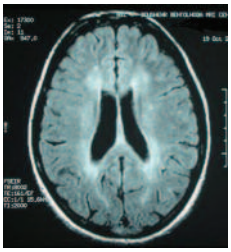

Fig. 1 Axial FLAIR image shows diffuse, bilateral, confluent increased signal intensity in the deep white matter

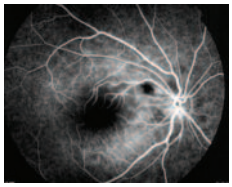

Fig. 2 fluorescein angiography shows small macular branch artery occlusion.

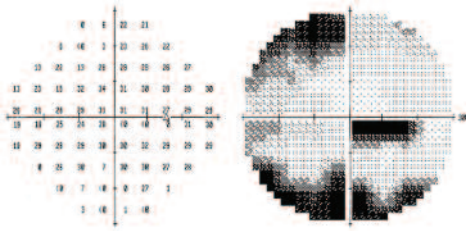

Fig. 3 Perimetry shows central scotoma consisting with macular problem

References: NONE

Key Words: Susac's syndrome, encephalopathy, branch retinal artery occlusion, hearing loss, multiple sclerosis

Financial Disclosure: NONE

Poster 50

Computed Tomography (CT) Demonstration Of Dorsal Midbrain Hemorrhage In Traumatic Fourth Cranial Nerve Palsy

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Introduction:
The location of injury to the fourth cranial nerve in closed head trauma is uncertain. In most cases, brain imaging fails to show a lesion. There are, however, seven reports demonstrating hemorrhage either in the dorsal midbrain or perimesencephalic cisterns. We describe a case of traumatic fourth cranial nerve palsy in which, for the first time, a dorsal midbrain haemorrhage was demonstrated on CT.

Methods:
A single case report and literature review with display of previous imaging reports of traumatic fourth cranial nerve palsy.

Results:
A 21-year-old man who suffered a traumatic brain injury from a motor vehicle accident recovered brain function except for an isolated left fourth cranial nerve palsy. Brain CT showed a large haemorrhage in the right dorsal midbrain directly in the path of what would become the left fourth cranial nerve. There has been previous imaging documentation of dorsal midbrain and superior cerebellar, quadrigeminal, or ambient cistern hemorrhage in patients with isolated post-traumatic fourth cranial nerve palsy. However, this is the first example of a large dorsal midbrain hemorrhage demonstrated on CT. The striking feature of our case is that a large midbrain hemorrhage, so easily seen on CT, would cause lingering damage limited to the fourth cranial nerve.

Conclusion:
Our case and the reported literature indicate that intrinsic midbrain or peri-mesencephalic hemorrhages may occasionally be seen on imaging in traumatic fourth cranial nerve palsy. Impact of the midbrain against the rigid free edge of the tentorium cerebelli appears to be the most plausible mechanism of injury. The intrinsic portion, or perhaps even the extrinsic portion, of the fourth cranial nerve would then be damaged when the dorsal midbrain or fourth cranial nerve exit zone in the anterior medullary velum collides with the tentorium.

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Key Words: Traumatic fourth cranial nerve palsy, dorsal midbrain hemorrhage, computed tomography, midbrain concussion, perimesencephalic hemorrhage

Financial Disclosure: NONE

Poster 51

Posterior Reversible Encephalopathy Syndrome (PRES) And Serous Retinal Detachment: A Common Pathogenesis?

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Introduction:
Posterior reversible encephalopathy syndrome (PRES) consists of neurologic manifestations associated with MRI findings of vasogenic edema predominantly in posterior cerebral hemispheres. In the setting of acute hypertension, there are two competing pathogenetic hypotheses: autoregulatory breakthrough leading to vasodilation and excessive autoregulation leading to vasoconstriction. We describe two patients with PRES and serous retinal detachments. Given evidence that favors vasoconstriction as the cause of serous retinal detachment in acute systemic hypertension, we propose that these ophthalmic manifestations and PRES share vasoconstriction as the underlying mechanism.

Methods:
Two case reports and review of the literature.

Results:
A 22 year old woman with angioimmunoblastic lymphoma developed renal failure, severe systemic hypertension, generalized seizures, and depressed consciousness. After blood pressure was normalized and seizures were controlled, she returned to full consciousness but had a right homonymous hemianopia and peripapillary serous retinal detachments in both eyes. Brain MRI demonstrated posterior parietal and occipital high T2/FLAIR signal without restricted diffusion, findings compatible with PRES. Within weeks, the serous detachments disappeared, but the homonymous hemianopia persisted. Repeat brain MRI showed that the right hemisphere signal abnormalities had disappeared, but high T2/FLAIR signal in the left hemisphere persisted, findings interpreted as left occipital infarction following PRES.

A 15 year old boy with binocular blurred vision was treated with corticosteroids for presumed Vogt-Koyanagi-Harada disease based on the finding of peripapillary serous retinal detachments in both eyes. He developed a seizure and a blood pressure of 250/140 owing to renal failure. Renal biopsy showed membranous glomerulonephritis. MRI showed signs of PRES. As blood pressure and seizures were controlled, vision rapidly recovered. Six months later, examination showed Elschnig spots and corresponding scotomatous visual loss.

Conclusion:
The combination of PRES and serous retinal detachments, reported here in two hypertensive patients for the first time, suggests vasoconstriction as a common underlying mechanism.

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Key Words: Choroidopathy, PRES, Vasoconstriction, Hypertension, Serous Retinal detachment

Financial Disclosure: NONE

Poster 52

Clinical Heidenhain Variant Of Sporadic Creutzfeldt-Jakob Disease (CJD) With Co-occurrence Of Prion Protein Types 1 and 2

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Background:
Sporadic Creutzfeldt-Jacob disease (sCJD) is a rare neurodegenerative illness comprising five phenotypically distinct subtypes based on the methionine(M)/valine(V) polymorphism at codon 129 of the prion protein (PrP) gene and presence of either one of the two protease-resistant PrP (PrPres), referred to as PrPres type 1 and type 2.1, 2 The most common of these five subtypes includes the sCJDMM(MV)1 that matches the "classic" sCJD and comprises the so-called Heidenhain variant (HsCJD) characterized by early and prominent visual symptoms. Recently, an additional subtype of sCJD has been described and identified as sCJDMM1-2 in which both types 1 and 2 histopathological changes are found in the same brain.3 The clinical Heidenhain phenotype in association with histopathological sCJDMM1-2 subtype has not been described.

Methods:
Clinical, neuroimaging, EEG findings, histopathological and immunostaining as well as PrPres characterization of 20 cases of sCJDMM1-2 were reviewed.

Results:
Two cases (10%) of HsCJD with ages at onset of 51 and 66 years and disease durations of 4 and 12 months were identified. Immunohistopathology and PrPres type determination were consistent with the features of the sCJDMM1-2 subtype.3 The visual cortex was more severely affected than the frontal cortex and was found to carry both PrPres types.4 The study of the quantitative distribution of PrPres types, expressed as mean percentage of total PrPres types 1 and 2 (1:2), was 79%:21% for the sCJDMM1-2 with 4 month and 45%:55% for the case with 12 months disease duration respectively. The cerebellum showed both PrPres types and PrP immunohistochemical patterns of the sCJDMM1-2.

Conclusion:
To our knowledge, this is the first histopathological finding of sCJDMM1-2 in Heidenhain variant of sporadic CJD. Also, the amount of PrPres type 2 and to a lesser extent that of the MM2-like immunohistochemical features increase with the disease duration, as seen for the sCJDMM1-2 .

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Key Words: Heidenhain variant of sporadic CJD disease duration, co-existence of Prion proteins types 1 and 2, disease duration

Financial Disclosure: NONE

Poster 53

Cortical Blindness Following a Near-Drowning Incident

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Introduction:
Near-drowning is a term applied when a patient survives for at least some period of time after suffocation from submersion in a liquid[1]. The primary mechanism of injury is pulmonary failure caused by fluid aspiration, which results in severe arterial hypoxemia and secondary ischemic damage to other organs, including the brain[2]. Victims of near-drowning incidents often suffer neurologic injury with long-term sequelae secondary to hypoxic-ischemic injury[3] .The regions of the brain that are particularly susceptible to such injury are the vascular end zones, hippocampus, insular cortex, and basal ganglia[4]. Thus far, visual loss as a consequence of hypoxic-ischemic damage from near-drowning has not been reported. We describe a case of profound visual loss due to bilateral occipital lobe infarcts following a near-drowning incident.

Methods:
Case report and literature review.

Results:
A 23-year-old male was found unresponsive at the beach after a near-drowning incident while surfing. He was intubated and admitted to the Trauma Intensive Care Unit. Computed Tomography (CT) of the chest showed bilateral airspace disease consistent with aspiration. Magnetic Resonance Imaging (MRI) of the cervical spine showed a fracture of the C-5 vertebral body, which did not require surgical intervention. Shortly after the patient was extubated on the fourth day of his hospitalization, he complained of profound bilateral visual loss. His visual acuity was noted to be counting fingers in both eyes, and an ophthalmologic exam was otherwise normal. An MRI of the brain with diffusion- and perfusion-weighted protocols revealed multifocal infarcts involving both occipital lobes.

Conclusion:
Near-drowning incidents may result in profound visual loss on the basis of hypoxic brain injury with resultant occipital lobe infarcts.

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Key Words: Near-drowning, Visual Loss, Occipital infarcts

Financial Disclosure: NONE

Poster 54

Neuro-ophthalmologic Abnormalities in a de novo Terminal 6q Deletion

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Introduction:
Terminal deletions of the long arm of chromosome 6 are relatively rare [1-2], but a typical phenotype has been proposed to include mental retardation, esotropia, developmental hypotonia, microcephaly, brain anomalies, seizures, and facial dysmorphism [2-5]. We describe here a 14 year old boy with a deletion of the long arm of chromosome 6 who has novel ophthalmologic features that might have been present but not recognized in previous patients.

Methods:
Careful clinical evaluation including MRI; conventional cytogenetic analysis on GTG-banded chromosomes; and 244K array CGH analysis.

Results:
This 14 year old boy had modest mental retardation, microcephaly, cortical dysplasia, seizures, facial dysmorphism, substantial myopia, and thick corneas. He also had a large angle esotropia appearing in infancy, congenital nystagmus, cross fixation, and impersistent eccentric gaze associated in this case with an abnormal brainstem but similar to the infantile esotropia syndrome seen more commonly in neurologically normal children. He had a *de novo* 10.79 Mb deletion on chromosome 6 from 6q25.3 to 6qter, a region encompassing 87 genes**.** Eleven genes remained for analysis after excluding genes coding for unrecognized or uncharacterized proteins and genes located in the deleted area that have been reported in phenotypically normal individuals. Of these only the *TBP* (TATA box binding protein) gene (probably acting in combination with *PSMB1*) has been associated with the neurologic signs observed in this patient.

Conclusion:
This boy had clinical features universally reported in patients with the 6q terminal deletion syndrome, including mental retardation and an esotropia. His unusual ocular motility pattern and thick corneas are two features that may be more common in 6q deletions than previously recognized. The characteristics of his ocular motility raise the question of whether the infantile esotropia syndrome might be caused in part by under expression of genes in 6q.

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Key Words: chromosome deletion, mental retardation, congenital nystagmus, esotropia, thick cornea

Financial Disclosure: NONE

Poster 55

Prevalence of Clinical Neuro-Ophthalmic Abnormalities in an ALS Cohort

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Introduction:
ALS-Plus syndromes are clinically diagnosed on the basis of impairments not accounted for by motor system pathology. Quantitative ocular motor abnormalities such as decreased smooth pursuit gain and increased antisaccade errors have been proposed as biomarkers for such pathological spread. Case series have also reported absent Bell's phenomenon and eyelid apraxia in ALS patients. We sought to evaluate the prevalence of clinical ocular motor and visual acuity abnormalities in a population of ALS patients.

Methods:
Patients at an academic ALS center and their caregivers participated in this study. Past ocular and neurological history were collected. High-contrast visual acuity (VA) and low-contrast letter acuity testing (2.5% and 1.25% levels) were performed. We also assessed ocular alignment. Ductions, smooth pursuit, voluntary saccades, anti-saccades, lid function, and eye deviation upon forced eyelid closure (e.g. Bell's phenomenon), were recorded on videos, which were evaluated by two senior neuro-ophthalmologists.

Results:
Data was collected from 16 patients with ALS (mean age 56 years, 56% female) and 7 age-matched controls. Bell's phenomenon was present less frequently in the ALS cohort (6/12[50%] vs. 5/5[100%], p=0.04). Gaze impersistence was observed in 3/16[19%] patients and 0/7 controls. Square wave jerks were observed in 5/16[31%] patients and 0/7 controls. Saccadic smooth pursuit was observed in both groups in both vertical (14/16[88%] ALS, 7/7[100%] controls) and horizontal (11/16[69%] ALS, 4/7[57%]) directions. There were no significant differences for other clinical findings.

Conclusion:
We have identified absent Bell's phenomenon, gaze impersistence and square wave jerks to be more common in individuals with ALS. These results support pathology outside the motor system, or so called ALS-plus, in a subset of patients. We are unable to differentiate ALS patients from normal subjects based upon the antisaccadic and smooth pursuit abnormalities that were identified in recordings by other investigators. This may reflect the sensitivity and resolution of bedside examination.

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Key Words: Amyotrophic lateral sclerosis, Clinical examination, Bell's phenomenon

Financial Disclosure: NONE

Poster 56

METASTASIS OF AN ADENOCARCINOMA TO AN INTRACRANIAL MENINGIOMA

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Introduction:
Meningioma is the most common intracranial tumour to harbor metastasis.
Breast cancer is the most common tumor that metastasizes into meningiomas, followed by lung cancer. Other types of tumour are unusual.

Methods:
Case report

Results:
A 57-year-old man presented with a two-month progressive visual loss in his left eye and a mild visual loss in his right eye. On physical examination, a left relative efferent pupillary defect and bilateral temporal optic disk pallor was noticed. A magnetic resonance imaging study (MRI) demonstrated a large homogenous mass compatible with meningioma in the olfactory sulcus affecting optic chiasm and both optic and olfactory nerves.
Pathological diagnosis confirmed a transitional meningioma that hosted a mucinous adenocarcinoma with signet ring cells. The inmunohistochemistry showed to be positive for CEA and CK CAM 5.2.
A total body computed tomography showed a spiculated pulmonary nodule, an intraluminal sigmoid colon mass, retroperitoneal adenopathies and lithic bone lesions.
Tumor-to-tumor metastasis is described in the literature. In our case, the different histopathology features of both tumors and inmunohistochemical profile of a colonic adenocarcinoma (positive for CEA y CK CAM5.2) suggested the metastasis to an intracranial meningioma. Our case fulfilled the Campbell and Pamphlett criteria.
Imaging is not reliable to exclude the metastasis within meningiomas.
MR-spectroscopy could be an ancillary tool to identify malignancy in a meningioma.
Cases of intrameningioma metastasis as the first clinical manifestation of occult primary cancer, as our case, have been described but are less frequent.

Conclusion:
Though unusual, metastasis in a meningioma has been reported and, in rare cases, the meningioma diagnosis is made first. A systemic work up study should be done to localize the primary tumor. The diagnosis of a metastasis to a meningioma is usually done with the histopathological examination.

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Key Words: meningioma, metastasis, colon adenocarcinoma, tumor-to-tumor, MRI

Financial Disclosure: NONE

Poster 57

Metastatic Cavernous Sinus Syndrome from a Carotid Body Tumor Undergoing Delayed Malignant Transformation

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Introduction:
Carotid body tumors (CBTs) are the most common paraganglioma of the head and neck and present as a slow growing, non-tender neck mass. They arise from accumulation of chemoreceptor cells of neural crest region during embryogenesis. Malignant transformation may occur in 6% of patients, but metastatic spread to the brain is very rare.

Methods:
Case report and review of the literature.

Results:
A 70 year old man presented to our service with diplopia. Thirty years prior to presentation, he noticed a bump on the left side of his neck while shaving. Biopsy revealed a benign CBT but complete resection was not performed. Approximately two years prior to presentation the bump increased in size and firmness. Repeat biopsy revealed a malignant paraganglioma with lymph node involvement. He underwent neck radiation and chemotherapy including cisplatin and etopisode. In January 2008, he noticed horizontal binocular diplopia and drooping of the left lid. A brain MRI revealed multiple enhancing brain tumors, with a dominant left temporal lobe mass but no explanation for his complaints.

Our examination revealed a left abducens palsy and Horner's syndrome. This led to a repeat MRI with thin cuts through the cavernous sinus which showed asymmetric enhancement on its left side. Over weeks, a right oculomotor palsy developed and repeat imaging showed spread of tumor to the right cavernous sinus. Radiation to the region was followed by slow improvement in his symptoms.

Conclusion:
This case is a reminder that benign CBTs may rarely undergo malignant transformation even decades after discovery and should be resected upon discovery. Extra-cervical metastases from malignant CBTs are rare but may occur in the brain. To our knowledge, this is the first report of cavernous sinus invasion by metastatic spread from a transformed CBT although a paraganglioma arising from the cavernous sinus has been reported.

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Key Words: Carotid body tumor, Cavernous Sinus Syndrome, Abducens Palsy, Horner's Syndrome, Paraganglioma

Financial Disclosure: NONE

Poster 58

Depression and Anxiety in Migraine Patients with and without Chronic Photophobia

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Introduction:
Nearly all patients with migraine report photophobia during a headache, but a subset of these patients report chronic photophobia. These patients are always light sensitive, even during headache-free periods. In this study, we assessed the prevalence of anxiety and depression in migraine patients with chronic photophobia and in migraine patients without chronic photophobia.

Methods:
IRB approval and informed consent was obtained from all subjects prior to participation. Sixteen subjects with migraine and chronic photophobia were compared to sixteen age- and gender-matched migraine subjects without chronic photophobia. Each subject met the International Headache Society classification criteria for episodic migraine. A photophobia questionnaire, the Beck Depression Inventory (BDI), and the Beck Anxiety Inventory (BAI) were administered to each subject.

Results:
The average photophobia score (S.D.) for 16 photophobic migraineurs was 8 (4), while the average for the non-photophobic migraineurs was 1 (0.5). ($p<0.000002$, 2-tailed t-test) The average BDI score (S.D.) for 16 photophobic migraineurs was 13 (10), while the average for the non-photophobic migraineurs was 6 (5). ($p<0.02$, 2-tailed t-test) The average BAI score (S.D.) for 16 photophobic migraineurs was 16 (10), while the average for the non-photophobic migraineurs was 4 (3). ($p<0.0004$, 2-tailed t-test)

Conclusion:
Migraine patients with chronic photophobia are more likely to manifest mild to moderate depression and anxiety compared to migraine patients without chronic photophobia. Physicians caring for these patients should be aware of the increased prevalence of depression and anxiety. Future research could assess if treatment of anxiety and depression improves symptoms of photophobia and headache in these patients.

References: NONE

Key Words: Migraine, Photophobia, Light Sensitivity, Anxiety, Depression

Financial Disclosure: NONE

Poster 59

Recurrence of Symptoms After Successful Carotid Cavernous Fistula Endovascular Treatment

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Introduction:
Carotid cavernous fistulas (CCF) are abnormal communications between the carotid arterial and the cavernous venous systems which occur spontaneously or with trauma. Some patients have recurrent symptoms after successful treatment. Patients with cortical venous drainage (CVD) are at higher risk for intracranial complications and may have different symptoms after successful treatment. This could be recurrence of the disease (reopening of the fistula), a new fistula at a different location, or aberrant nerve regeneration. The purpose of the study is to evaluate recurrence of symptoms with and without CVD after successful fistula closure.

Methods:
Retrospective chart review was performed on 19 patients from three academic centers who had been treated for CCF in the last 9 years. We contacted patients by telephone to assess for recurrent symptoms since their last clinic visit.

Results:
Of 19 patients, we were able to follow up 17 patients with a mean follow-up time of 35 weeks (range 1-112). Four patients (23%) had recurrent symptoms requiring further treatment. Recurrent symptoms included the following: diplopia (75%), pain (75%), vision loss (50%), red eye (25%). All exacerbations were recurrences of prior symptoms. All patients had documented CVD, while none with anterior drainage (via the superior ophthalmic vein) had recurrent symptoms ($p<0.011$).

Conclusion:
CCFs with CVD are more likely to present with severe symptoms and to reoccur after successful treatment. Further investigations regarding optimal treatment in patients with CVD on initial presentation should be pursued to prevent recurrence and need for retreatment.

References: NONE

Key Words: Carotid Cavernous Fistulas, Diplopia, Endovascular treatments

Financial Disclosure: NONE

Poster 60

Reliability of Magnetic Resonance Imaging for Diagnosis of Hypopituitarism in Children with Optic Nerve Hypoplasia

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Introduction:
The purpose of this study is to determine the reliability of pituitary abnormalities on magnetic resonance imaging (MRI) as an indicator of hypopituitarism in children with optic nerve hypoplasia. Although we have described the association of pituitary abnormalities on MRI and hypopituitarism in children with optic nerve hypoplasia, the diagnostic evaluation was not standardized in our previous studies.[1,2] In the current study, all of the children received an MRI of the pituitary interpreted by a masked neuroradiologist and clinical evaluation by a pediatric endocrinologist with extensive serologic testing.

Methods:
Retrospective analysis of 41 children with optic nerve hypoplasia evaluated at Arkansas Children's Hospital between 2002 - 2009. All patients had MRI of the brain/sella interpreted by a neuroradiologist masked to the patient's endocrinologic status. All patients had clinical evaluation by a pediatric endocrinologist and a standardized panel of serologic testing that included a complete blood count, electrolytes, cortisol, ACTH, TSH, Free T4, IGF-1, IGF-BP3 levels.

Results:
Among the 41 children, 16 were diagnosed with hypopituitarism. All 16 patients had pituitary abnormalities on MRI that included absence of the pituitary infundibulum, ectopic location or absence of the posterior pituitary gland, and pituitary gland hypoplasia. Among the 25 patients with normal endocrinologic function, only 2 had pituitary abnormalities on MRI (thin infundibulum and absence of infundibulum).

Conclusion:
Pituitary abnormalities on MRI are sensitive (100%) and specific (92%) indicators of hypopituitarism in children with optic nerve hypoplasia.

References:
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2. Phillips PH, Spear C, Brodsky MC. Magnetic resonance diagnosis of congenital hypopituitarism in children with optic nerve hypoplasia. J AAPOS 5:275-280, 2001.
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Key Words: hypopituitarism, optic nerve hypoplasia

Financial Disclosure: NONE

Poster 61

Transient Optic Perineuritis as the Initial Presentation of CNS involvement of pre-B Cell Acute Lymphocytic Leukemia

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Introduction:
Modern chemotherapeutic modalities for leukemia have dramatically increased survival as well as increased recurrence rates and involvement of secondary sites, including the eyes and optic nerves. We present a patient who underwent systemic and prophylactic intrathecal chemotherapy for pre-B cell ALL who presented with a transient optic perineuritis and subsequently developed an infiltrative optic neuropathy with central retinal artery occlusion (CRAO).

Methods:
The patient underwent serial ophthalmic exams, visual field testing, MRI's, systemic infectious workup, lumbar puncture, vitreous biopsy, and optic nerve biopsy.

Results:
A 20-year-old man initially had right eye pain, 20/20 vision OU, no RAPD, full but mildly painful motility on the right, and right optic disc edema. MRI revealed enhancement of the right optic nerve sheath and possible thickening of the optic nerve. CSF was negative for neoplastic cells and infectious agents. With oral prednisone, the vision remained stable and the pain and optic disc edema resolved by day 10. Subsequent CT revealed suspicious pulmonary nodules, but systemic workup was negative for aspergillosis. The patient remained without ocular complaints until ten weeks later when he had acute visual loss in the right eye. Exam revealed no light perception with CRAO and venous stasis in the right eye. Repeat MRI showed slightly decreased nerve sheath enhancement. Two days later, flocculent material was found protruding from the right optic nerve head. Vitreous biopsy was inconclusive with negative culture. Subsequent optic nerve biopsy revealed a monomorphic infiltrate and B-cell monoclonal population on flow cytometry. The patient received radiation therapy, high-dose intrathecal chemotherapy, and systemic chemotherapy.

Conclusion:
The optic nerve and perineural structures can serve as a sanctuary for leukemic cells. Opportunistic infections and toxicity from antileukemic drugs can present a diagnostic dilemma in patients with ALL, highlighting the importance of a multispecialty approach including the oncologist, ophthalmologist and radiologist.

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Key Words: optic perineuritis, leukemia, infiltrative optic neuropathy, central retinal artery occlusion, acute lymphocytic leukemia

Financial Disclosure: NONE

Poster 62

Toxocariasis: A Diagnostic Dilemma

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Introduction: Toxocariasis is a parasitic infection that affects various organs due to the larvae's ability to travel to various organs. The main target organs include liver, lung, eyes and the CNS and can be found clinically by elevated serum eosinophilia and positive toxocara antibodies. Larvae only survive a few months and are difficult obtain via biopsy. Our case report examines an atypical case of toxocara and examines differential diagnoses.

Case Report: A 33-year-old male reported lower extremity parasthesias and 6 months following, a central scotoma in left eye. His past medical history is significant for Bell palsy in 2006 that resolved within one week without treatment. Initial exam demonstrated 20/20 in the right eye and count fingers at 10' in the left eye, Ishihara chromatic plates yielded full color on the right and 7/11 on the left, an afferent pupillary defect on the left, funduscopy showed a lesion superiorly juxtaposed to the optic nerve. Magnetic resonance imaging of the brain revealed multiple enhancing lesions on T1 with gadolinium, and 6-8 ventricular white matter abnormalities while laboratory work up yielded serum and cerebral spinal fluid eosinophilia (32K/uL), along with 2 oligoclonal bands in the cerebral spinal fluid and positive serum toxocara titers. Suspecting a diagnosis of toxocara, a pars plana vitrectomy found no larvae, however protienaceous eosinophilic tissue consistent with Splendore-Hoeppli phenomenon was described.

Discussion: Although this case may be explained by the diagnosis of toxocara, it is also important to rule out other diagnoses such as demyelinating disorders and eosinophilic granulomatosis. These two diseases can present with some similar characteristics, further testing and evaluation is warranted.

References: NONE

Key Words: Toxocara canis, Eosinophilia

Financial Disclosure: NONE

Poster 63

Treatment of Geotropic Horizontal Canal Benign Paroxysmal Positional Vertigo; Comparison of Barbeque Rotation and Gufoni

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Background:
There are a number of repositioning maneuvers in treating HC-BPPV, but they lack standardization.
Objective: The purpose of this study was to determine and compare the immediate efficacy of barbecue rotation and Gufoni maneuver in treating geotropic horizontal canal benign paroxysmal positional vertigo (HC-BPPV).
Design: A randomized prospective sham-controlled study of patients with geotropic HC-BPPV.

Methods:
Patients (n=124) with geotropic HC-BPPV were randomized to one of three groups: barbecue rotation, Gufoni maneuver and sham maneuver. Immediate response was determined 30 minutes after each maneuver by positioning maneuver based on resolution of vertigo and positional nystagmus. The repositioning maneuver was repeated once more when the symptom and signs persisted one hour later. The patients also had weekly follow-ups for one month after the initial maneuver.

Statistics:
X2 and t test were used to compare the clinical characteristics of the patients allocated to each treatment group and to determine the therapeutic efficacies of maneuvers. Cox proportional hazards regression analysis was used to determine the effect of independent variables on the outcome. We used Kaplan-Meier survival analysis for estimating the survival curve reflection of the long-term effect, and the log rank test for a statistical comparison of two groups. A two-tailed P value of less than 0.05 was considered significant.

Results:
Of the 124 patients, 82 were women with a women-to-men ratio of 1.9:1. The age ranged from 11 to 93 years (mean±SD = 62.2±15.5, median = 62.0) without difference in the mean age between women and men (59.1±16.4 vs 66.3±13.4, p>0.05). The right ear was determined to be involved in 64 patients (51.6%). After excluding 6 patients with refused study or resolution of vertigo and nystagmus during the confirmatory positioning maneuver, 124 patients were randomly allocated to barbecue rotation (n = 40), Gufoni's maneuvers (n = 43) and sham groups (n = 41). After maximum two maneuvers, 28 of 40 (71.8%) patients showed resolution of vertigo and nystagmus in barbecue rotation group while 18 of 43 (45.0%) and 15 of 41 (37.5%) patients had resolution in the Gufoni maneuver and sham groups (p = 0.005). The overall Kaplan-Meier survival curve for the long-term effects also showed a better outcome with barbecue rotation compared with Gufoni (p = 0.02) and sham maneuvers (p = 0.02). However, therapeutic efficacy of Gufoni maneuver did not differ from that of sham maneuver in terms of both immediate (p = 0.37) and long-term (p = 0.65) outcomes.

Conclusion:
Using a prospective randomized trial, we demonstrated that barbeque rotation was superior to Gufoni's or sham maneuver in treating geotropic HC-BPPV. Therapeutic efficacy of Gufoni's maneuver, however, did not differ from that of sham maneuver. Barbeque rotation should be applied first in patients with geotropic HC-BPPV if applicable.

References: NONE

Key Words: BPPV = benign paroxysmal positional vertigo, HC-BPPV = BPPV involving the horizontal semicircular canal, Barbecue rotation, Gufoni's maneuver

Financial Disclosure: NONE

Poster 64

Surgical Management of Gliomas with the Perineural Arachnoidal Gliomatosis Growth Pattern

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Introduction:
Benign gliomas may adopt one of two distinct growth patterns. Most familiar is the classic intraneural glial proliferation where neoplastic growth occurs within individual fascicles. Contrastingly, perineural arachnoidal gliomatosis (PAG) is characterized by florid invasion of the leptomeninges, with relative sparing of the nerve itself. We describe successful surgical management of such a tumor.

Methods:
Interventional case description

Results:
A healthy 4 YO female presented with rapidly progressive right-sided proptosis. Magnetic resonance imaging (MRI) revealed an enhancing fusiform intraconal lesion: characteristic of PAG the optic nerve was visualized coursing through the tumor. Despite conservative measures, the tumor progressed in size resulting in a visual acuity of 20/70 and mild optic pallor. She became progressively disfigured and by the age of 7 had withdrawn socially, with declining academic performance.

Via a lateral orbitotomy, a window was cut into the thickened dura of the optic nerve. A rouge spongy infiltrate was able to be suctioned from the subarachnoid space. Small more solidified areas were excised with unipolar cautery. Care was taken to avoid identifiable blood vessels and the optic nerve itself, which was easily visualized. An estimated 60 to 70% of the tumor was removed and the dural window was sewn shut. Postoperatively, proptosis improved from 10.5 to 2 mm and visual acuity improved to 20/60.

Conclusion:
Optic nerve gliomas are benign tumors that in most cases require no intervention. Occasionally, progressive enlargement is encountered. Non-surgical options, radiation therapy and chemotherapy, are in most cases minimally effective. Surgery is effective in controlling tumor spread, but in general sacrifices vision. In this case, with PAG, substantial debulking of the tumor was achieved with preservation of vision. This suggests that this may be a viable option for management of gliomas with this specific growth pattern. Admittedly, this is a single case and should be interpreting accordingly.

References: NONE

Key Words: Optic nerve glioma, Surgical excision, perineural arachnoid gliomatosis

Financial Disclosure: NONE

Poster 65

Superior Oblique Paralysis: Eyes Cyclotorsion Difference Is More Important in Left Eye Paralysis

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Introduction:
Unilateral superior oblique palsies causes an interocular torsion imbalance. Cyclotorsion asymmetry in favor of the left eye was established, by our team, during a preceding measurement in healthy subjects (50). Origin of this oculomotor disorder is peripheral, so we can expect to find the sign of this asymmetry in case of unilateral superior oblique palsie.

Methods:
Thirty one patients, aged from 14 to 50 years, reached unilateral superior oblique palsies (14 right, 17 left) were examined. Each one had an oculomotor examination which objectified a unilateral superior oblique paralysis. A scanning laser ophthalmoscopy (SLO) was taken in standard condition. Congenital and acquired palsies were not distinguished

Results:
Patients cyclotorsions average is higher and more variable than healthy subjects. Inter ocular difference, is more important in the left eye palsie group than healthy subjects, and reach of zero in the right eye group.

Conclusion:
Right analysis of ocular cyclotorsion must take account of this asymmetry especially in the event of unilateral superior oblique paralysis.

References: NONE

Key Words: Cyclotorsion, Superior Oblique Paralysis

Financial Disclosure: NONE

Poster 66

Acute Comitant Esotropia in a Child with a Cerebellar Tumor

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Introduction: Acute comitant esotropia is generally thought to be benign and further neurological investigationis not warranted.

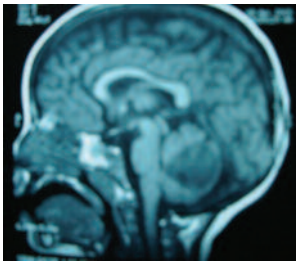
Method: A 4-year-old girl was referred for management of acute acquired comitant esotropia. Her family noticed sudden inward deviation of his left eye 3 months ago. On presentation, the angle of esodeviation was 40 prism diopters (PD) at distant and near fixation. The patient's visual acuity was 20/20 in both eyes with alternate fixation. There was no lateral incomitance and no deficit of abduction bilaterally (figure1a,b,c). No difference in the size of the deviation was found with fixation of the right versus the left eye.

Result: He was referred to a pediatric neurologist, but the neurological examination did not reveal any additional pathological findings. A brain MRI revealed a large mass with an enhanced peripheral rim at the midline of the cerebellum (figure2a,b). The tumor was completely excised andhistological examination confirmed the diagnosis of pilocytic astrocytoma. Esotropia resolved after tumor excision.

Conclsion: Acute comitant esotropia is unusual sign of brain tumor.

Fig1a,b,c:

Figure 2a,b:



References: NONE

Key Words: acute comitant esotropia, cerebellar tumor

Financial Disclosure: NONE

Poster 67

Diagnosis and Treatment of Convergent Strabismus Fixus

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Introduction:
Convergent strabismus fixus is an ocular motor abnormality where the eye is fixed in adduction. It is usually also hypotropic with a limitation of elevation. Typically associated with moderate to high myopia, it is a progressive disorder that can initially mimic a paralytic strabismus but then becomes restrictive. Orbital imaging studies demonstrate a slippage or displacement of the extraocular muscle path and pulley; the lateral rectus displaced inferiorly and the superior rectus displaced medially. Loop myopexy has become the treatment of choice for this disorder.

Methods:
We describe the presenting signs and symptoms of 5 patients with convergent strabismus fixus. All had large angle esotropia and hypotropia with a limitation to abduction and elevation.

Results:
Orbital imaging studies demonstrated displacement of the lateral rectus muscle path inferiorly and the superior muscle path nasally. Treatment was by loop myopexy with significant improvement in their ocular alignment.

Conclusion:
Convergent strabismus fixus is a shift in muscle pathways leading to the globe fixed in an esotropic and hypotropic position. It is important that neuro-ophthalmologists recognize this condition and obtain orbital imagining confirming the shift in extraocular muscle locations. Appropriate referral for loop myopexy should be considered.

References:
1. Wong, Leo, Khoo, Loop myopexy for the treatment of myopic strabismus fixus, J AAPOS, 9, 589, 2005.

Key Words: Strabismus fixus, Loop myopexy, Orbitopathy

Financial Disclosure: NONE

Poster 68

Synovial Sarcoma of the Left Arm Affecting Bilateral Extraocular Muscles: A Paraneoplastic Syndrome?

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Introduction:
Synovial sarcoma is an aggressive malignancy occurring in the soft tissues of young adults that compromises up to 10% of adult sarcomas. Although paraneoplastic syndrome induced by this type of cancer has not been reported to date, we recently encountered a patient with synovial sarcoma of the left arm who manifested ophthalmic symptoms in a manner resembling paraneoplastic syndrome.

Methods:
Case report

Results:
A 30-year-old man presented with bilateral swelling of the upper eyelids and painful eye movement that had progressively worsened for 2 months. He was aware of a mass in his left arm prior to these symptoms. His best corrected visual acuity was RE: 20/20 and LE: 20/20 at initial examination. Bilateral swelling of the upper eyelids and exophthalmos were noted, and movement in both eyes was limited to the upper field because of pain. Orbital magnetic resonance imaging (MRI) demonstrated enlargement of the bilateral extraocular muscles. Intraocular pressure was normal. A biopsy of the mass on the left arm was performed, leading to a diagnosis of synovial sarcoma based on characteristic histologic findings. The patient was treated by surgical resection and chemotherapy. Soon afterwards, ocular symptoms improved along with the orbital lesions seen in MRI findings.

Conclusion:
It appears that the patient's synovial sarcoma affected the bilateral extraocular muscles as a result of paraneoplastic syndrome. This may represent the first case of paraneoplastic syndrome caused by synovial sarcoma.

References: NONE

Key Words: synovial sarcoma, extraocular muscles, paraneoplastic syndrome

Financial Disclosure: NONE

Poster 69

Ocular Oscillations and Ataxia in Antiganglioside Antibody Syndrome

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Introduction:
Anti-ganglioside antibodies are found in various neurological disorders which constitute a continuum from peripheral neuropathy to encephalitis. However, ocular oscillations and ataxia without other neurological deficits have rarely been described in association with anti-ganglioside antibodies.

Methods:
From January 2008 to July 2009, we identified four patients with acute ataxia and ocular oscillations, who were found to have anti-GD1b or anti-GM1 antibodies, in three University Hospitals of Korea.

Results:
All four patients had acute ataxia and ocular oscillation. Three of them showed upbeat nystagmus, periodic alternating nystagmus, gaze-evoked nystagmus, or direction-changing positional nystagmus and one patient exhibited opsoclonus. All patients had serum IgG or IgM antibodies to GD1b and three of them also showed antibodies to GM1. Cerebrospinal fluid examinations, nerve conduction studies, and brain MRI were normal. In all patients, the symptoms and signs resolved over the following several months after symptom onset without specific treatment.

Conclusion:
Acute ataxia with various forms of ocular oscillations may be a sole or predominant manifestation of anti-ganglioside antibody syndrome. The development of ocular oscillations in association with ataxia indicates a central pathology involving the cerebellum or brainstem in this antibody-mediated disorder. Anti-ganglioside antibodies should be measured in patients with acute ataxia and ocular oscillation of undetermined etiology.

References: NONE

Key Words: Ataxia, Nystagmus, Opsoclonus, anti-ganglioside antibody

Financial Disclosure: NONE

Poster 70

Sixth Nerve Palsy in Pregnancy Due To Occult Intracavernous Meningioma

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Introduction:
Meningiomas may exhibit accelerated growth during pregnancy. Small intracavernous lesions may pose a diagnostic challenge on neuroimaging, especially when contrast agents are avoided. We present two cases of sixth nerve palsies presenting or worsening during pregnancy due to small intracavernous meningiomas that were missed on initial neuroradiologic review.

Methods:
Observational case reports

Results:
Case 1: A 40-year-old woman in her 21st week of pregnancy complained of binocular horizontal diplopia and headaches for one week. Her ophthalmologic exam revealed a partial right sixth nerve palsy and was otherwise normal. An unenhanced MRI the brain and orbits was reportedly normal, as was a lumbar puncture. Three weeks later, she developed a complete right sixth nerve palsy. The MRI was re-reviewed, and a small lesion was noted in the right cavernous sinus consistent with a meningioma. She was followed for the remainder of her pregnancy with serial exams and MRI's, both of which were stable, and underwent Gamma knife radiosurgery postpartum.
Case 2: A 34-year-old woman complained of binocular horizontal diplopia and headaches for 3 weeks. Her ophthalmologic exam revealed a partial left sixth nerve palsy. An MRI of the brain and orbits with gadolinium was reportedly normal, as were a lumber puncture and workup for myasthenia gravis. On follow-up 5 months later, she was 14 weeks pregnant, and her abduction deficit had nearly resolved but then worsened again by her 8th month of pregnancy. A brain MRI postpartum revealed a small left intracavernous meningioma, and she subsequently underwent Gamma knife radiosurgery.

Conclusion:
An intracavernous meningioma should be considered in the differential diagnosis of a sixth nerve palsy presenting or worsening during pregnancy. Workup should include an unenhanced MRI of the brain with attention to the cavernous sinus, and a high index of suspicion is warranted in such cases.

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Key Words: sixth nerve palsy, cavernous sinus, meningioma, pregnancy

Financial Disclosure: NONE

Poster 71

Management of Patients with Bilateral Nystagmus and Head Posture Due to Monocular Blindness

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Introduction:
Monocular infantile blindness may be associated with bilateral horizontal nystagmus and pronounced head posture that requires treatment. This nystagmus has been characterized as a manifest nystagmus of the latent type in patients who have inherited a genetic predisposition for infantile strabismus[1]. It increases during abduction and dampens in adduction of the seeing eye. This clinical entity has also been reported in the literature as unilateral Ciancia syndrome[2].

Methods:
We reviewed the records of eight consecutive patients with unilateral Ciancia syndrome from a single center who underwent extraocular muscle surgery to eliminate an abnormal head posture and/or to improve ocular alignment. Outcome measures included amount of head posture and deviation at last follow-up.

Results:
Combined horizontal surgery (recession of the medial rectus muscle and resection of the lateral rectus muscle) of the fixing eye regularly resulted in a pronounced reduction or elimination of the abnormal head posture (face turn). In cases of coexisting esotropia this single procedure also markedly reduced the angle of deviation. In four patients surgical management included bilateral operations either concurrently or in two separate sessions.

Conclusion:
Surgical management of unilateral Ciancia syndrome has so far not been specifically addressed in the literature. Based on our series, combined horizontal surgery of the sound eye according to the principles of Kestenbaum[3] is mandatory and effective if improvement of a significant face turn is the goal of surgery. This single procedure also leads to an improvement of accompanying esotropia. In some patients bilateral surgery is advisable: in the fixing eye for correction of face turn and in the blind eye to achieve satisfactory alignment.

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2. Jampolsky, A: Strategies in strabismus surgery. Pediatric Ophthalmology and Strabismus: Transactions of the New Orleans Academy of Ophthalmology, New York, NY: Raven Press, 363, 1986.
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Key Words: Nystagmus, Head posture, Ciancia syndrome, Monocular blindness, Extraocular muscle surgery

Financial Disclosure: NONE

Poster 72

Left Third Nerve Palsy and Bilateral Internuclear Ophthalmoplegia in Colon Cancer Patient : Rare Clinical Manifestation and Non-invasive Diagnostic Approach in Multifocal Inflammatory Leukoencephalopathy

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Introduction:
5-fluorouracil (5-FU) and levamisole have been used in adenocarcinoma of colon for adjuvant therapy. These can rarely induce a demyelinating disease of the brain such as the multifocal inflammatory leukoencephalopathy (MIL). We report a rare ophthalmologic finding of patient with adenocarcinoma of colon.

Methods:
A 45-years-old woman had taken 12 weeks of 5-FU and levamisole treatment for moderately differentiated Stage IIIb adenocarcinoma of the cecum after right hemicolectomy. On admission she revealed nearly complete third nerve palsy on left eye and also showed bilateral internuclear ophtalmoplegia. Several diagnostic procedures were taken.

Results:
Brain MRI revealed a midbrain lesion and multiple cerebral white matter lesions. Diffusion tensor image (DTI) and Positron Emission Tomography-Computed Tomography (PET-CT) were taken. PET-CT presented decreased fluorodeoxyglucose uptake of brain stem and multiple white matter lesions. DTI revealed decreased fractional anisotropy (FA) value and diffusivity of the left midbrain compared with those of contralateral side. And disruption of tract is observed in 3D-tractogram. After cessation of 5-FU and levamisole administration, Methylprednisolone was injected intravenously. 5 weeks later, eye movement of patient was nearly recovered.

Conclusion:
The MIL lesion involving left oculomotor nucleus and neighboring both medial longitudinal fasciculi is responsible for left third nerve palsy with bilateral internuclearophthalmoplegia. We report a rare ophthalmologic finding in a patient during the treatment of the adenocarcinoma of colon.

References: NONE

Key Words: multifocal inflammatory leukoencephalopathy, colon cancer, third nerve palsy, internuclear ophthalmoplegia, diffusion tensor image

Financial Disclosure: NONE

Poster 73

Omnidirectional Gaze-evoked Nystagmus in Cavernous Hemangioma in Brainstem

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Introduction:
Gaze-evoked nystagmus (GEN) is caused by a “leaky” neural integrator, which fails to maintain eccentric gaze positions after centrifugal eye movements. It is usually observed as the results of toxic, metabolic or paraneoplastic disorders, rather than single structural lesions. We report a patient with omnidirectional GEN due to cavernous hemangioma in the brainstem.

Methods:
A previously healthy 28-year-old man presented with progressive dizziness and diplopia for one year. Neurological examinations revealed head tilt to the left and symmetrical GEN in both horizontal and vertical directions, and partial abduction deficit of the right eye. Rightward saccades were slow.

Results:
Results of routine chemical analyses were normal. MRI of the posterior fossa showed a cavernous hemangioma expanding from the dorsal medulla to pons.

Conclusion:
The lesions in bilateral nucleus prepositus hypoglossi and medial vestibular nucleus or in the paramedian tract neurons, which have been recently recognized as a site of neural integration, may lead to omnidirectional GEN of our patient.

References: NONE

Key Words: Gaze-evoked nystagmus, Nucleus prepositus hypoglossi, Medial vestibular nucleus, Paramedian cell tract

Financial Disclosure: NONE

Poster 74

Mydriasis Caused by Oculomotor Nerve Compression by the Tortuous Posterior Cerebral Artery (P2)

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Introduction:
It is well known that anisocoria is produced by an internal carotid-posterior communicating artery (ICPCom) aneurysm. We report a case of unilateral mydriasis without external ophthalmoplegia caused by the compression of the oculomotor nerve by a tortuous posterior cerebral artery (P2).

Methods:
Case presentation and review of the relevant literature

Results:
A 79-year-old woman noted several episodes of transient upper eye lid drooping that lasted for a few seconds 10 days prior to presentation at her local ophthalmologist. Mydriatic pupil was noted in the left eye (OS) and there was no constriction in response to a flash light in either direct or consensual ways. There were no limitations in the eye movements of either eye (OU). She was referred to us under a suspected diagnosis of tonic pupil OS. Her pupils showed 2 mm OD and 4 mm OS in bright room illumination. There was no difference in the position of the upper eye lid margin between the two eyes. Her extraocular movements were normal OU. Because of the history of episodic ptosis accompanied by an enlarged pupil, 3D-CT angiography were obtained and demonstrated a saccular aneurysm (2.4x3.9 mm) in the left ICPCom region. A frontotemporal craniotomy was performed to treat the aneurysm. It was found that the oculmotor nerve was not compressed by the aneurysm but that the nerve was compressed by the tortuous P2. By dissecting the arachnoid membrane surrounding the oculomotor nerve (the liliquest membrane), the oculomotor nerve was successfully decompressed, which subsequently resulted in restoring the size and function of the affected pupil.

Conclusion:
There was a previous case report describing congenital anomalous vascular compression of the oculmotor nerve was suggested to be the cause of mydriasis without ophthamoplegia based on MRI findings (1). This is the first case that showed that anisocoria could be corrected by releasing the vascular compression.

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Key Words: Mydriasis without ophthalmoplegia, vascular compression of the oculomotor nerve , microvascular decompression

Financial Disclosure: NONE

Poster 75

Botulinum Toxin Type A as a Reversible Treatment for Residual Fourth Cranial Nerve Palsies, Secondary to Intracranial Hypertension in the Context of a Ventriculo-Peritoneal Shunt Failure

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Introduction:
This case report illustrates bilateral fourth cranial nerve palsies in the setting of an intracranial hypertension, secondary to a ventriculo-peritoneal shunt failure. More importantly, it shows how diminishing the impact of a fourth cranial nerve palsy in an active patient's life can be obtained by a single injection of botulinum toxin type A in the ipsilateral inferior oblique muscle. The resolution of symptoms made further intervention unnecessary in this case.

Methods:
A 29 year old man presented with gradually increasing, binocular, vertical double vision and lingering headaches for 4 months.
His relevant medical history revealed a congenital biliary atresia that required liver transplantation.
18 years earlier, the patient had suffered a subarachnoid hemorrhage due to a ruptured aneurism with a secondary hydrocephalus requiring ventriculo-peritoneal shunting, and lead to the discovery of 3 additional intracranial aneurisms.
The brain imaging was performed, revealing a recurrence of a tetraventricular hydrocephalus with pressure on both fourth cranial nerves. Surgical revision of the shunt failed to restore single vision.
The vertical misalignment and cyclotorsion rendered especially reading challenging, thus keeping the patient from studying, even several months after surgery, despite one side had recovered.
In attempt to alleviate the symptoms a single injection of botulinum toxin type A was performed in the ipsilateral inferior rectus muscle.

Results:
The previous symptomatology had resolved even one year after the treatment and the oculomoteurn control was normalized.

Conclusion:
This patient's quality of life was considerably improved by one injection to the ipsilateral inferior oblique muscle. Botulinum toxin type A can be a powerful treatment for reversible paresis of the antagonist muscle in the setting of fourth cranial nerve palsy due to intracranial hypertension.

References: NONE

Key Words: Fourth Cranial Nerve palsies, Intracranial hypertension, Ventriculo-peritoneal shunt failure, Treatment, Botulinum toxin Type A

Financial Disclosure: NONE

Poster 76

Training to Improve Reading Speed in Patients with Central Scotoma - A Randomized and Controlled Study

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Background: We examined two training methods for optimizing reading ability in patients with central scotoma due to juvenile macular dystrophy with established eccentric retinal fixation locus and optimal use of low vision aids.

Method: 36 patients were randomized into two groups: Group 1: training to read during single-word presentation (rapid serial visual presentation, RSVP) with elimination of eye movements (n=20), Group 2: Training to optimize reading eye movements (sensomotoric training SM, n=16). Training was performed for 4 weeks, ½ hour per day and five days a week. Reading speed (page reading aloud) was measured before and after training. Eye movements during silent reading were recorded before and after training in 11 patients.

Results: Median reading speed was 83 words per minute (wpm) (interquartile range 74-105 wpm) in the RSVP training group and 102 (interquartile range 63-126 wpm) in the SM group before training and increased significantly to 104 (interquartile range 81-124 wpm) and 122, respectively (interquartile range 102-137 wpm; p=0.01 and 0.001) after training, i.e. patients with RSVP training increased their reading speed by a median of 21 wpm, while it was 20 wpm in the SM group. There were individual patients, who benefited strongly from the training. In the RVSP group, increasing reading speed correlated with decreasing fixation duration (r= -0.75, p= 0.03), whereas in the SM group, increasing reading speed correlated with a decreasing number of forward saccades (r=-0.9, p=0.01).

Conclusion: Although the median effect of both training methods was limited, individual patients benefited very much. Each training method indicates a specific effect on reading strategy: the RVSP method reduces fixation duration, the SM method decreases the number of forward saccades. Patients can apply their newly learned reading strategy in the natural reading situation, e.g. in page reading without special presentation of the text.

For future studies, based on their eye movement pattern at baseline, patients can be selected and assigned to the appropriate training method. These results can be used as a basis for further improvement of training methods for optimizing reading performance in patients with a central scotoma.

ConclusionPreconditions of a training effect are the optimal adaptation of low vision aids and the motivation of the patients.

References: NONE

Key Words: reading speed, reading training, central scotoma, eye movements, juvenile macular dystrophy

Financial Disclosure: Pro-Retina Germany

Poster 77

Diagnosis of Chronic Progressive External Ophthalmoplegia Without Muscle Biopsy

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Introduction:
Chronic progressive external ophthalmoplegia (CPEO) features insiduous development of slowly progressive external ophthalmoplegia and ptosis. Its diagnosis is based on the results of a muscle biopsy including ragged-red fibers. CPEO can also be diagnosed by verifying specific alterations of mtDNA in the skeletal muscle. However, such deletions may not be found in leukocytes; therefore the diagnosis requires an invasive procedure to obtain the skeletal muscle. To investigate the efficacy of long range polymerase chain reaction (PCR), a much more simplified easy method, to diagnose CPEO mtDNA deletions in buccal cells and urinary epithelium.

Methods:
A molecular genetic analysis in buccal cells and urinary epithelium of patients with CPEO mtDNA deletions in medial rectus muscle was performed. Total mtDNA was extracted from peripheral blood, medial rectus muscle, buccal mucosa, urinary epithelium and hair follicles. Long range PCR encompassing entire mtDNA was carried out. If long range PCR revealed a deletion of mtDNA, the exact deletion range and breakpoints were identified by sequencing.

Results:
MtDNA deletion associated with CPEO could be successfully found in two patients' medial rectus muscle as well as in buccal cells, urinary epithelium and other tissues.

Conclusion:
The traditional method used for the diagnosis of CPEO, the muscle biopsy, might be substituted by long range PCR in other tissues, which are more easily obtained.

References: NONE

Key Words: Chronic progressive external ophthalmoplegia, mitochondrial DNA deletions, muscle, buccal cells, urinary epithelium

Financial Disclosure: NONE

Poster 78

Pituitary Stalk Hemangioblastoma: an Uncommon Tumor Associated with Visual Loss

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Introduction:
Hemangioblastomas (HBLs) represent about 1.5 to 2.5% of all primary intracranial tumors and occur most commonly in the cerebellum. Supratentorial HBLs are rare and 80% occur as sporadic tumors, unrelated to von Hippel Lindau (VHL) disease in adults. We describe a patient with a pituitary stalk HBL who presents with blurred vision and bitemporal field defects.

Methods:
This 33-year old man present with a 4-month history of gradual progressive visual loss and mild headaches. His best-corrected visual acuity was 20/60 OU and bitemporal defects. Pupils, color vision, and extraocular motility were normal. Anterior and posterior segment exams were unremarkable. Neurological exam was normal.

Results:
MRI of the brain revealed a 2.8-cm globular, cystic lesion isointense on T1, hyperintense on T2, and enhancing with contrast. MRA revealed a vascular blush supplied by superior hypophyseal arteries. Serum and cerebrospinal fluid markers, including alpha-fetoprotein, human chorionic gonadotropin, and placental alkaline phosphatase, were negative. PCR testing for VHL gene was negative.

Subtotal resection by right pterional craniotomy revealed a red cluster of blood vessels growing within the pituitary stalk. Histopathologic sections stained with hematoxylin and eosin revealed a rich capillary network lined with endothelial cells interspersed with eosinophilic and vacuolated stromal cells. Histologic stains for epithelial membrane antigen, cytokeratin, CD34, CD31, factor VIII-related antigen, and glial fibrillary acidic protein were all negative, but the stain for inhibin was positive, which was consistent with the diagnosis of HBL. His residual tumor has remained stable for the past 6 years.

Conclusion:
Our patient presented with visual loss related to a hemangioblastoma, an uncommon tumor of the pituitary stalk. Only 5 patients with such tumors have been described in the literature to date. Similar tumors, including angioblastic meningioma and metastatic renal cell carcinoma, must be carefully ruled out by appropriate histologic stains.

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Key Words: suprasellar hemangioblastoma, visual field defects, pituitary stalk

Financial Disclosure: NONE

Poster 79

Orbital Decompression for Optic Nerve Sheath Meningioma: A Case Report

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Introduction:
Optic nerve sheath meningiomas (ONSMs) are rare tumors of the anterior visual pathway which untreated often lead to vision loss. Stereotactic fractionated radiotherapy can improve or stabilize vision. Radiation retinopathy can occur as a complication.

Methods:
A 69 year old white woman presented with a visual acuity 20/30 OD and 20/20 OS. There was 3.5mm of exophthalmos by Hertel exophthalmometry.

Results:
An MRI scan revealed an ONSM without intracranial extension. During a fourteen year period her visual acuity decreased to 20/60, and discomfort worsened. The patient did not want radiation therapy. She underwent a right sided three wall bone orbital decompression. Post-operatively her visual acuity improved to 20/30 that is unchanged after more than three years.

Conclusion:
The management of ONSM is controversial. Surgical removal of tumor often results in blindness. Turbin and Kennerdell et. al. reported radiotherapy with optic nerve sheath fenestration. The author used orbital decompression to reduce optic nerve compression, improve appearance and well being.

References:
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Key Words:
Orbital decompression, Optic nerve sheath meningioma, Vision loss, Orbital discomfort, Exophthalmos

Financial Disclosure: NONE

Poster 80

Benign Unilateral Apraxia of Eyelid Opening

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Introduction:
Transient ptosis in clinical practice can cause a clinical dilemma and often results in an extensive evaluation. We report a case series of patients with unilateral transient ptosis after sleep that is being identified as a benign clinical entity and defined as benign unilateral apraxia of eyelid opening (BUAEO).

Methods:
A list of 208 patients with ptosis seen at a single neuro-ophthalmology practice was drawn through the computer coding system. Transcribed reports of the outpatient visits of these patients were reviewed retrospectively from this database to identify the final diagnoses. All patients with a diagnosis of ptosis due to underlying neurologic, myopathic or neuromuscular problems were excluded. Five patients with ptosis, which occurred on awakening and resolved after mechanical eyelid elevation, were included.

Results:
Three out of five patients were women. Ages ranged from 49-71 years. All patients experienced complete ptosis only upon awakening after sleep. The ptosis was unilateral and did not recur after manual elevation of the eyelid. None of the patients had any underlying neurological problem that could be contributory and were thoroughly investigated with neuroimaging and laboratory testing including acetylcholine receptor antibodies. Examination revealed no ocular cause for the ptosis in each patient. In all patients, follow-up telephone reports noted no subsequent condition that could be responsible for ptosis.

Conclusion:
We propose that in patients with complete unilateral ptosis occurring only on awakening and resolving with mechanical eyelid opening, and in the absence of pupillary or ocular motor abnormalities, BUAEO should be considered. The identification and diagnosis of this benign condition is of importance in that extensive evaluation may not be warranted unless there is development of other unexplained neurological findings.

References: None

Key Words: Ptosis, Apraxia, Eyelid

Financial Disclosure: NONE

Poster 81

Thyroid Eye Disease in the Elderly

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Introduction:
The time of onset of thyroid eye disease (TED), an autoimmune orbital disorder, is known to follow a bimodal peak: 40-44 years and 60-64 years in women, as well as 45-49 and 65-69 years in men. Women are affected 4-5 times more often than men. It is rare in children and very old people. The purpose of this study was to evaluate the nature of TED in patients older than 80 years at time of disease onset.

Methods:
A retrospective review of files of nine patients 80 years or older at the time of TED diagnosis, in three neuro-ophthalmology units.

Results:
Nine out of 491 (1.8%) patients with TED were 80 years old or older at the time of diagnosis. Five were men and four were women. Mean age was 85 years (80-94). The most common symptom was diplopia (6/9). Only one patient had severe eyelid swelling and chemosis with progressive restriction of ocular motility and thus needed steroids. Sight threatening complications such as compressive optic neuropathy and/or severe corneal exposure were not observed. In one patient the diagnosis of Graves' was concurrent with the ocular symptoms, in seven Graves' disease was known 6-240 (mean 76.7) months prior to TED appearance, and one patient never developed thyroid gland disease. Three patients were hyperthyroid at time of TED onset, two were hypothyroid and five were euthyroid.

Conclusion:
TED after the age of 80 is rare and its course seems to be more benign than in younger patients. Female predominance was not observed. Most patients are euthyroid at the time of diagnosis and ocular involvement follows the thyroid gland disease.

Key Words: TED, Elderly

Financial Disclosure: NONE

Poster 82

Thyroid Eye Disease in Young Patients Compared with Older

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Introduction:
To compare clinical features of thyroid eye disease (TED) patients younger than 40 years of age with older patients.

Methods:
A retrospective chart review of 131 consecutive TED patients from two medical centers. Demographics and medical history as well as ophthalmic manifestation, management and outcome were recorded. The groups were compared using t-test and Chi-Square tests**.**

Results:
Twenty three out of 131 patients (17.5%) were 40 years old or younger at the time of TED diagnosis. Mean age at onset was 29.7 ± 9.1(8-40) years in the younger group and 56.4 ± 10.9 (41-94) years in the older group. Female predominance was noted in both groups (2:1). Majority of patients in both groups had hyperthyroidism at ophthalmopaty onset (86% of the young patients and 82% in the older group). Treatment for Graves' disease was similar in both groups. Proptosis and lid retraction were more frequent in the young patients group compared with diplopia and periocular edema, which were more frequent in the advanced age group ($p \leq 0.03$). Bilaterality and imaging results were similar in both groups. Treatment with immunosuppressant drugs, radiation and decompression, although not statistically significant, was more common in older patients.

Conclusion:
TED is less common under the age of 40 among our patients. Its clinical presentation is different i.e. more lid retraction and proptosis and less restrictive myopathy, which has not been previously described. It seems their disease was less severe and this is similar to previous reports.

References: NONE

Key Words: Thyroid Eye Disease, Age

Financial Disclosure: NONE

Poster 83

Assessment of Facial Nerve Injury after Temporal Artery Biopsy

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Introduction:
Temporal artery biopsy is considered the gold standard for diagnosis of giant cell arteritis. This procedure may be considered to be benign by some referring physicians. Reported complications include hematoma, infection, would dehiscence, skin necrosis, cerebral infarction. In this report, we describe 4 patients who suffered from facial nerve injury after biopsy.

Methods:
A review of the records in the tertiary care referral neuro-ophthalmology practices at the University of California – San Francisco and Stanford University from October 2004 to January 2009 was performed. Patients who had facial nerve injury after temporal artery biopsy were included.

Results:
Four patients (2 men and 2 women) ranging in age fro 60 to 87 years (mean 72.8 years) were included. All patients were referred for evaluation of facial nerve palsy, following temporal artery biopsy performed in other departments. 3 patients had unilateral biopsy, 1 patient had bilateral biopsy. Based on clinical photographs and examination records, all patients had biopsy performed in the pre-auricular or pre-trichal temporal regions. Self-reported improvement in frontalis function ranged from zero (no improvement) to 75 percent (mean 35%). No patients underwent surgery to correct brow position. There were no other complications of biopsy.

Conclusion:
Understanding the fascial planes and anatomic relationships is essential when performing surgery. The “danger zone” is a region of vulnerability of the facial nerve branches to injury. The deeper facial nerve branches are separated from the superficial temporal artery by only the superficial temporal fascia. All four patients in this series had biopsies within the “danger zone.” This is a rare, and likely underreported complication of this procedure. Recovery of frontalis function was variable, consistent with previous reports.

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Key Words: Temporal arteritis, Surgery, Complications

Financial Disclosure: NONE

Poster 84

Visual Neuropraxia and Progressive Vision Loss fromThyroid-Associated Stretch Optic Neuropathy

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Introduction:
Animal models have provided information on the tensile strength of the optic nerve, but to our knowledge no *in vivo* study of the tensile strength of the human optic nerve has been reported. Accordingly, we present a case series of stretch optic neuropathy, an often unrecognized cause of vision loss from thyroid eye disease.

Methods:
Observational study of thyroid-associated stretch optic neuropathy.

Results:
Three cases of stretch optic neuropathy were identified. Visual acuity was better than 20/40. Two patients had arcuate scotoma. Moderate to severe proptosis of 25 to 33 mm was present, without evidence of apical orbital compression. Two patients had retinal hemorrhages suggesting venous stasis retinopathy; the venous stasis retinopathy resolved after orbital decompression. Orbital decompression resulted in improvement of visual function. The rate of decibel sensitivity loss on automated perimetry was estimated at -0.042 dB/da in one case, with complete blindness projected to occur within 785 days from the onset of visual symptoms.

Conclusion:
Stretch optic neuropathy presents initially as neuropraxia with temporary visual loss. Orbital decompression should be considered for treatment before permanent and irreversible visual loss ensues.

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Key Words: stretch optic neuropathy, thyroid eye disease, neuropraxia, vision loss

Financial Disclosure: NONE

Poster 85

Spheno-orbital Meningioma: A Multidisciplinary Approach

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Introduction:
Traditionally, patients with meningiomas extending from the sphenoid ridge and middle cranial fossa into the orbit undergo extensive surgery with complications ranging from cosmetic deformity, double vision, vision loss, or death. We have a series of patients operated on over the last 10 years by a modified pterional craniotomy with excellent cosmetic and functional outcome. This presentation will highlight the surgical approach in three of these patients who presented with ophthalmic manifestations.

Methods:
Institutional Review Board approval was obtained to conduct a retrospective chart review of our patients with spheno-orbital meningiomas. The subject population was defined all patients with an ICD-9 diagnosis of meningioma (225.2) who underwent craniotomy for resection, specifically, CPT code 61608 (resection lesions parasellar area, cavernous sinus, clivus, middle skull base, intradural), CPT code 61592 (Orbitocranial zygomatic approach to middle cranial fossa), or CPT code 61605 (resection lesion infratemporal fossa, parapharyngeal space, petrous apex, extradural) seen at Vanderbilt University Medical Center in the last six years. The data collected includes age, sex, tumor location, pre-admission diagnosis, ophthalmology consult, radiologic evidence of bone involvement, hospital course, surgical intervention and duration, pathologic diagnosis, discharge time and neurologic outcome.

Results:
Forty-seven patients with spheno-orbital meningiomas operated upon between 2003 and 2009 were identified. Three of these patients had a combined neurosurgery and orbital surgery approach. In spite of extension into the superior orbital fissure and dissection in this area, none of our patients had any cranial neuropathies post-operatively. All of them had improvement of their pre-operative complaint.

Conclusion:
Patient's with spheno-orbital meningioma may benefit from a combined neurosurgical and orbital surgical approach.

References: NONE

Key Words: Intracranial Tumors, Skull Base Tumors, Proptosis, Superior Orbital Fissure, Middle Cranial Fossa

Financial Disclosure: NONE

Poster 86

Transcranial Supralateral Orbital Wall Reconstruction Using Titanium Mesh Embedded in Porous Polyethylene (Medpor Titan™)

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Introduction:
Pterional-orbitozygomatic craniotomies provide access to both the orbital and intracranial spaces. When utilized in the resection of neoplastic disease, large defects in the superior and lateral orbital walls result. A recently developed implant embeds a titanium mesh within porous polyethylene. We describe our experience using this implant.

Methods:
In this retrospective university based study, we reviewed the medical records of eight consecutive patients (mean age 54, range 34 to 78 years of age; 1 man, 7 women) during the time period, July 2008 to July 2009. All patients underwent pterional-orbitozygomatic craniotomy with resection portions of the superior and lateral orbital walls with reconstruction using the Medpor Titan implant.

Results:
Indications for surgery were sphenoid wing meningioma (n=6), adenoid cystic carcinoma (n=1), and pachymeningitis (n=1).

In all patients surgery was performed similarly. Following completion of tumour resection, the size of the defect in the orbital wall was assessed (3 to 12 cm^2). Slightly larger sized implants were cut, such that 3 to 4 mm of overlap was achieved medially and laterally, and shaped, matching the shape of the excised bone. Edges are secured under (occasionally over) solid bone medially and laterally. Anterior, one to three titanium screws are used to fix the implant to the superior orbital rim. The craniotomy bone flap is then secured in a standard fashion.

In all patients proptosis was either improved or normalized. No implant related complications were seen. This includes implant migration/displacement, infection, abnormalities in extraocular motility or the development of enophthalmos or pulsatile proptosis.

Conclusion:
Based on our initial experience, due in part to its malleability and ability to retain shape, the Titan Medpor implant appears to be well suited to replace complex orbital wall defects of varying size and shape. Admittedly, this is a small retrospective study and its role in orbito-cranial reconstruction continues to evolve.

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Key Words: Orbital reconstruction, Porous polyetheylene with titanium mesh, Tumor resection

Financial Disclosure: NONE

Poster 87

Measurement of Distance Objective Visual Acuity with the Computerized Optokinetic Nystagmus Test in Patients with Ocular Diseases

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Introduction:
To investigate the efficacy of a computerized optokinetic nystagmus (OKN) test in evaluating the objective distant visual acuity, and to determine the correlation between subjective and objective visual acuities in patients with various ocular diseases.

Methods:
Eighty-five eyes in 71 patients were included and categorized as media opacity (20 patients), generalized retinal disease (19), glaucoma (17), maculopathy (15), or optic neuropathy (14). To measure visual acuity at a distance, OKN visual stimuli were presented using a laser beam projector on a 127-inch white screen located 10 feet from a subject. Eye movements were recorded and analyzed by computerized infrared oculography. Objective visual acuity was determined using both the induction and suppression method. The correlations and relationship between objective and subjective distance visual acuities were evaluated.

Results:
Linear regression analysis showed that objective visual acuity measured by OKN induction and suppression methods correlated with subjective visual acuity ($p<0.05$). There were also significant correlations between subjective visual acuity and objective visual acuity using induction and suppression methods in each group.

Conclusion:
The objective visual acuities measured by distance OKN methods correlated well with subjective visual acuities in patients with ocular diseases. These results indicate the feasibility of OKN induction and suppression in measuring objective visual distance function in ocular disease.

References: NONE

Key Words: computerized optokinetic nystagmus, objective distant visual acuity

Financial Disclosure: NONE

Poster 88*

Quantitative Diplopia Assessment and Correlation with Visual Quality of Life

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Introduction:
Patients with adult-onset diplopia have trouble with many routine activities like driving, reading, and using the computer. These difficulties may persist despite improved objective ocular alignment (*i.e.* after strabismus surgery). We are developing a simple method to quantitatively and reproducibly characterize oculomotor deviations and their effect on quality of life in adult patients with diplopia.

Methods:
Subjects with diplopia of any etiology were assessed in primary and eccentric distance gaze positions using the CROM device to determine precisely the head position in space(1). The area of single binocular vision was plotted on a modified Goldmann perimetery chart, and the area was quantitated using *ImageJ* software. Stereopsis was measured at distance and at near. A diplopia score also was calculated(1). Subjects completed visual quality of life assessments [NEI VFQ-25 and AS-20(2)]. Correlations between diplopic area at distance, diplopia score, and quality of life were calculated.

Results:
All 13 subjects were able to report diplopia reliably, and the methods proved reproducible even in the hands of non-expert physician trainees. Initial results demonstrate that the diplopia score has a moderate correlation (R=0.43) with psychosocial quality of life (QOL) measures on the AS-20 and a lesser (R=0.29) correlation with functional QOL. Both general and visual QOL measured by the VFQ-25 declined with worsening diplopia scores. The diplopic area had similar but less strong correlations with the QOL assessments.

Conclusion:
Adult-onset diplopia results in reduced visual QOL as measured by general (VFQ-25) and strabismus-specific (AS-20) QOL instruments. The diplopia severity has a linear correlation with worsening psychosocial and general and vision-specific QOL measures. The diplopia score determination is simple and reproducible, and its use with QOL instruments will be useful in determining outcomes of future clinical trials.

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Key Words: diplopia, quality of life, outcomes assessment

Financial Disclosure: NONE

Poster 89*

Obstructive Sleep Apnea (OSA) In Idiopathic Intracranial Hypertension (IIH): A Prospective Study

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Introduction:
OSA is strongly associated with obesity and is thought to be associated with IIH. However, the prevalence of OSA in IIH patients is unknown. Patients at high risk for OSA can be identified using the Berlin questionnaire, but it is also unknown if the Berlin questionnaire reliably identifies IIH patients at high risk for OSA. We aimed to determine the prevalence of OSA in newly-diagnosed IIH patients and to validate the Berlin questionnaire as a screening tool for OSA in IIH patients.

Methods:
Prospective study of consecutive patients with newly-diagnosed IIH. Pregnant patients and patients less than 16 years old were excluded. All patients completed the Berlin questionnaire and had a diagnostic sleep study. We collected demographic/anthropometric data (age, sex, race, height, weight, and BMI), Berlin questionnaire results, sleep study results, and clinical data (symptoms, signs, automated perimetry results, and CSF opening pressure). OSA was defined as a respiratory disturbance index of >15.

Results:
We included 15 patients for this interim analysis; 11 female (73.3%), 6 Caucasian (40%), 9 African-American (60%). Age ranged from 17-54 (median 32). BMI ranged from 27.3-49.9 (median 40.4). Seven patients (47%) had OSA diagnosed by sleep study. Berlin questionnaire scores were “high risk” for OSA in 9 patients (60%), of which 6 had OSA by sleep study. Only one of 6 patients with a “low risk” score had OSA by sleep study. The sensitivity of the Berlin questionnaire for OSA in IIH patients was 85.7%, while the specificity was 62.5%.

Conclusion:
Our preliminary findings suggest that the prevalence of OSA may be higher in IIH patients than in the BMI-matched general population, where it is about 30%. A “high risk” score on the Berlin questionnaire suggests OSA in IIH patients and should prompt a sleep study. Findings from a larger sample will be presented at the meeting.

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Key Words: Idiopathic Intracranial Hypertension, Obstructive Sleep Apnea, Berlin Questionnaire

Financial Disclosure: NONE

Poster 90*

Imaging Strategies in Isolated Third Nerve Palsy (IIIrd): Detecting Intracranial Aneurysms

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Introduction:
Expert interpretation of modern non-invasive neuroimaging should detect nearly all aneurysms responsible for isolated IIIrds. Our objective was to review modern diagnostic strategies used to evaluate patients with isolated IIIrds.

Methods:
Retrospective review of all PCOM aneurysmal IIIrds seen at our institution since 2001, by searching diagnosis codes of IIIrd, aneurysm, and subarachnoid hemorrhage (SAH).

Results:
16/416 cases presented with an acute isolated painful IIIrd from a PCOM aneurysm. 4/16 had SAH on initial CT, facilitating diagnosis of aneurysmal IIIrd. 6/12 cases with isolated IIIrd and normal non-contrast head CT were correctly diagnosed with PCOM aneurysms [5 with non-invasive imaging (2 CTA, 3 MRI/MRA), 1 with catheter angiography (prior aneurysm clipping, contra-indicating MRI and creating CT artifact)]. The 6 remaining cases had reportedly negative non-invasive studies (CT, MRI/MRA in 5; CT, CTA, MRI/MRA in 1) at outside institutions. In one case, outside studies were not available and repeat MRI/MRA showed a PCOM aneurysm; in 5 cases, review of the outside non-invasive studies by us/neuroradiology/neurosurgery identified a PCOM aneurysm, confirming misinterpretation by outside radiologists rather than incorrect technique. For those misinterpreted, 5/6 were interpreted by general radiologists; in 1/6, the radiologist received neuroradiology training but was practicing mostly as a general radiologist. The smallest aneurysm correctly detected by initial CTA was 3.5x2.5x2.5 mm. The size range for PCOM aneurysms initially missed was 5x3x3mm to 6x8x3 mm.

Conclusion:
Since 2001, we have performed only one catheter angiogram to correctly diagnose a PCOM aneurysmal IIIrd (patient could not have non-invasive imaging). All presumed negative non-invasive studies were misinterpreted by general radiologists who had incomplete clinical information. We agree with recent reports[1,2] suggesting that meticulous interpretation of CTA/MRI/MRA by trained neuroradiologists[5] aware of the clinical reason for vascular imaging is essential in obtaining the highest sensitivity[3,4] of non-invasive imaging of isolated IIIrds.

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Key Words: third nerve palsy, non-invasive imaging, intracranial aneurysm, posterior communicating artery, subarachnoid hemorrhage

Financial Disclosure: NONE

Poster 91*

Leber Hereditary Optic Neuropathy Gene Therapy Clinical Trial for G11778A Mutation

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Introduction:
The purpose of this presentation is to provide information and to improve enrollment in the preparatory phase of the LHON gene therapy clinical trial for the G11778A mutation. The aim is to identify and characterize affected patients and carriers for planned gene therapy studies that will utilize “allotopic expression” by delivering copies of normal nuclear-encoded ND4 gene into the nuclei of retinal ganglion cells via an adeno-associated virus vector. The normal ND4 protein expressed in the cytoplasm is then imported into the mitochondria.

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

Results:
From September 2008 to September 2009, 45 subjects with LHON G11778A (24 affected, mean age 30.1 years (range 9.9-62.5); 21 carriers, mean age 43.2 (9.3-84.7) years) were recruited. Median time from visual loss was 33 months. Mean OCT3 RFNL thickness was 62.7 μm for affected and 100.7 μm for carriers ($p<0.01$). Mean PERG amplitude was 0.48 μV for affected versus 0.98 μV for carriers ($p<0.01$). There was a subset of carriers with PERG amplitudes 40% less than the normal expected age-related mean amplitude of 1.00 μV.

Conclusion:
Based on the first year recruitment results, we may consider selecting affected patients with mildly reduced RNFL or carriers with low PERG amplitudes (if this test is a predictor of conversion to LHON) for the intraocular ND4 gene injections in the future trial. We are actively recruiting participants and welcome referrals of affected and carriers of LHON G11778A. Contact information and detailed information are available by googling “BPEI LHON”.

References: NONE

Key Words: Leber hereditary optic neuropathy, gene therapy, clinical trial

Financial Disclosure: NONE

Poster 92*

Etiology and Prognosis of Sixth Cranial Nerve Palsy

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

Methods:
Records of all sixth cranial nerve palsy patients seen between 1995 and 2009 were reviewed.

Results:
Of 514 records reviewed, there were 540 recorded episodes of sixth cranial nerve palsy. Age of onset ranged from 4 months-94 years. Microvascular ischemia was the etiology in 49% overall (265/540), but was the most common cause in 64% over age 65 (158/245), and in 66% age 50-65 (91/138). Neoplasm was the cause in 9% overall (48/540), and did not vary significantly by age group. Trauma was the cause in 8% overall (44/540), but was the most common etiology for 42% of 7-18 year olds (10/24) and for 31% of 4 month-7 year olds (4/13). Demyelinating disease was the cause in 6% overall (31/540), but was the most common etiology in 22% of 18-50 year olds (27/122). Increased intracranial pressure was the etiology in 21% of 4 month-18 year olds (8/38), and 5% of 18 to 50 year olds (6/122). Overall, 14% of cases were undetermined (77/540), and did not vary significantly by age group. Prognosis varied by etiology but not by age of onset. Complete recovery was seen in 45% and no recovery was seen in 12% with an average of 11 month follow-up.

Conclusion:
This post-MRI era study shows a higher incidence of microvascular than neoplastic etiology compared to pre-MRI era studies. No other cause of sixth cranial nerve palsy exceeded 10%, including neoplasm, trauma, demyelinating disease, and increased intracranial pressure. Etiology depends on age, but prognosis depends on etiology.

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Key Words: diplopia, sixth nerve palsy, cranial nerves, microvascular ischemia, demyelinating disease

Financial Disclosure: NONE

Poster 93*

Effects of Target Motion Perturbation during Initiation and Steady-State Pursuit in Humans with Infantile Esotropia

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Introduction:
Smooth pursuit consists of two phases: initiation and steady-state. During initiation, the eye starts from zero velocity and accelerates to match target velocity. In steady-state pursuit, the eye continues to track a moving target, and accelerates or decelerates when target velocity changes. Patients with infantile esotropia exhibit a nasotemporal asymmetry of pursuit initiation favouring nasalward movements. The goal of this study is to investigate whether an asymmetry also exists during steady-state pursuit.

Methods:
Six patients with infantile esotropia and 11 control subjects were recruited. Eye movements were recorded using a video-based system while subjects tracked a red laser target during monocular viewing. For the initiation phase, a "step-ramp" paradigm was used, with targets moving at 5, 10 or 15 deg/s nasally or temporally. During steady-state pursuit, a "ramp-in-ramp" paradigm was used—the target moved at an initial velocity of 5 or 15 deg/s and then accelerated or decelerated to 10 deg/s after 500 ms.

Results:
Large nasotemporal asymmetries were found in patients during pursuit initiation ($p=0.0017$), with temporalward gains 48% lower than nasalward gains. No asymmetry was found during steady-state pursuit. Pursuit latency was increased by 21% during initiation ($p=0.0001$) and 25% during steady-state pursuit ($p=0.0011$) in patients when compared to control subjects, but no asymmetry was found.

Conclusion:
This is the first study to demonstrate the absence of a nasotemporal asymmetry during steady-state pursuit in humans with infantile esotropia. Previous studies have shown that pursuit initiation is driven primarily by retinal velocity error, whereas both retinal velocity and position errors are used to modulate steady-state pursuit. The presence of a nasotemporal asymmetry during pursuit initiation suggests that processing of retinal *velocity* error signal is impaired, while its absence during steady-state pursuit indicates that processing of retinal *position* error signal is intact in patients with infantile esotropia.

References: NONE

Key Words: infantile esotropia, smooth pursuit, pursuit initiation, pursuit maintenance, pursuit asymmetry

Financial Disclosure: NONE

Poster 94*

Anti-Aquaporin-4 Antibody in Chinese Severe Optic Neuritis

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Introduction:
Optic nerve and spinal cord are more commonly affected in Asian MS, which is called optic-spinal MS (OSMS). Anti-AQP4 antibody plays an important role in differentiation of multiple sclerosis(MS) and neuromyelitis optica (NMO) in recent years. To investigate the anti-AQP4 antibody distribution in Chinese severe optic neuritis patients and its prognostic value to visual and neurological outcome.

Methods:
Single center, prospective, cohort study. Anti-AQP4 antibody was detected by indirect immunofluorescence method using human AQP4-transfected cell on Chinese severe ON. Clinical features at baseline and follow-up were compared between seropositive and seronegative groups. The visual and neurological outcome was evaluated according to the sero-positivity.

Results:
Anti-AQP4 antibody was detected in 11 of 34 severe ON patients (32.4%). Five sero-positive recurrent ON patients had significantly higher anti-AQP4 antibody titers (range 512 times to 65536 times, median 512 times) than those of 6 seropositive patients with only one episode (ranged 16-512 times, median 32 times) (P=0.021). Female to male ratio(10:1) and antinuclear autoantibody (ANA) positivity in anti-AQP4 antibody seropositive patients (3/9, 33.3%) were higher than those of negative group (12:11; 0/19; p=0.026). The seropositive patients had poorer visual outcomes than seronegative patients (p=0.025). During averaged 32 months follow-up, 2 of 11 seropositive patients (18.2%) developed clinically incomplete transverse myelitis, while no similar symptoms reported in the seronegative group.

Conclusion:
Anti-AQP4 antibody positivity is relatively high in Chinese severe optic neuritis. Anti-AQP4 antibody positivity suggested a poorer visual outcome, higher risk of developing spinal cord lesion, and a closer association between severe ON and systemic autoimmune disorders.

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Key Words: optic neuritis, neuromyelitis optica, anti-aquaporin 4 antibody, multiple sclerosis, outcome

Financial Disclosure: NONE

Poster 95*

Macular Carotenoids in Patients with Photophobia

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Introduction:
The carotenoids lutein and zeaxanthin are concentrated in the macula, protect the retina from oxidative damage, and filter out blue light. We previously used Raman resonance spectroscopy to demonstrate elevated levels of carotenoids in patients with migraine and benign essential blepharospasm (BEB). In this investigation we used a complementary, image-based method, autofluorescence imaging, to quantify carotenoids in subjects with migraine and BEB. Our objective was to determine if macular carotenoids play a role in the pathogenesis of photophobia.

Methods:
Two groups of photophobic subjects were recruited with age- and gender-matched controls: subjects with BEB and subjects with migraine. These subjects reported light sensitivity, even when they were spasm- or headache-free. We excluded eyes with macular disease, cataracts, or a best-corrected acuity <20/80. We used a novel CCD-camera-based instrument that detects fluorescence from retinal lipofuscin chromophores ("auto fluorescence") to indirectly quantify and spatially image the distribution of macular pigment. Customized image processing software was used to convert pixel intensity maps to proportional files, and to quantify macular pigment optical density (MPOD).

Results:
The average (S.D.) MPOD for 21 migraine patients was 0.34 (0.15) and 0.20 (0.13) for controls. ($p<0.0058$, 2-tailed t-test) The average MPOD for 16 blepharospasm patients was 0.14 (0.12) and 0.21 (0.13) for controls. ($p<0.19$, 2-tailed t-test)

Conclusion:
Patients with migraine and chronic photophobia have macular carotenoid levels higher than controls. Patients with BEB and photophobia have carotenoid levels that are not significantly different from controls. It is not clear what role, if any, these compounds play in the pathogenesis of photophobia. One hypothesis states that patients with migraine and chronic photophobia accumulate these compounds to protect themselves from frequencies of light that are noxious. Further investigations will be conducted to determine if dietary supplementation of these compounds improves symptoms of photophobia.

References: NONE

Key Words: Light Sensitivity, Migraine, Blepharospasm, Autofluorescence, Photophobia

Financial Disclosure: NONE

Poster 96*

Macular Homonymous Hemianopsia

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Introduction:
Patients with an optic tract lesion develop a homonymous hemianopsia associated with bow tie optic atrophy. Optical Coherence Tomography(OCT) has been proven to be a useful clinical tool in the diagnosis and follow-up of optic neuropathies and optic atrophy. This study's purpose is to evaluate if bow tie atrophy can be detected by OCT.

Methods:
Four patients aged 7 to 36 presented to the neuro-ophthalmology clinic at the XXX Eye Center with lesions involving the optic tracts on one side. All patients had a complete neuro-ophthalmic examination, including formal visual fields testing which showed a homonymous hemianopsia that respected the vertical midline. OCT-3 measures of the Fast Macular Thickness as well as the Fast Retinal Nerve Fiber Layer (RNFL) 3.4 Thickness protocols were collected. RNFL and macular scans were compared to aged matched normative database and observed for signs of optic atrophy.

Results:
RNFL thickness was not significantly decreased from normal in any quadrant. The fast macular thickness map, however, displayed temporal thinning of the macula in the eye ipsilateral to the lesion, and nasal thinning in the eye contralateral to the lesion. The pattern of the thinning respected the vertical midline in all eyes.

Conclusion:
In our small sample of patients with bow tie atrophy due to an optic tract lesion, OCT was able to detect homonymous macular atrophy that respects the vertical midline.

References: NONE

Key Words: Optic tract, retinal nerve fiber layer, optical coherence tomography, optic atrophy, homonymous hemianopsia

Financial Disclosure: NONE

Poster 97*

Management of Vertical Ocular Misalignment in Thyroid Eye Disease using Adjustable Suture Techniques

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Introduction:
Restrictive vertical strabismus due to thyroid eye disease (TED) is challenging to correct. We report the clinical outcomes of a management approach employing adjustable sutures.

Methods:
Fifty-five subjects with TED and hypertropia underwent adjustable-suture inferior rectus recession (IRRec) by one surgeon (NJV). Surgery variably included superior rectus recession (SRRec) on the hypertropic eye. Forced-ductions determined the number of vertical muscles recessed. Post-adjustment goal was orthotropia or undercorrection in primary gaze without hyperdeviation in downgaze ipsilateral to IRRec. Outcomes were vertical alignment within 5pd of orthotropia, diplopia, reoperation, and need for prisms.

Results:
Median preoperative primary-position deviation was 14pd (range 4-50). 38(69%) subjects had unilateral IRRec; 17(31%) bilateral. 24(44%) subjects had SRRec. 11 patients were adjusted same-day, 41 next-day. On long-term follow-up (mean 38 wks), no subjects had diplopia, although two required re-operation (3.4%, 95%CI 0.4%-12.5%) and 18(33%) required prism (13/18 were <6pd). 11 of these 20 subjects had overcorrections. 46(84%) subjects had successful vertical alignment after one surgery, of which 30 were orthotropic. In multivariate regression, day of adjustment, muscles recessed, and previous decompression did not affect outcomes

Conclusion:
Previous studies report a reoperation rate of 17-45%. Using adjustable-sutures, operating on multiple muscles if indicated, and planning for postoperative IR weakening can reduce this rate. While 1/3 of patients required prisms, long-term deviations were small. TED-associated hypertropia can be successfully managed with unilateral and bilateral adjustable-suture IRRec, with or without supplemental SRRec.

References: NONE

Key Words: double vision, hypertropia, thyroid eye disease, strabismus surgery, surgical outcomes

Financial Disclosure: NONE

Poster 98

Familial Multiple Sclerosis Presenting With Familial Intracranial Hypertension

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Introduction:
A 24 year-old obese female who presented with several months of headaches and blurry vision. She first noticed some floaters and then her vision OS worsened several weeks later. She was seen by an ophthalmologist and a neurologist who noted bilateral optic nerve swelling. Her vision was 20/70 and 20/800 with an APD OS. She was thought to have either pseudotumor cerebri or optic neuritis.

Because of the diagnostic uncertainty, she was sent for further evaluation and treatment to our clinic.

Methods:
Case report and review of the literature.

Results:
A 24 year-old obese female who presented with several months of headaches and blurry vision. She first noticed some floaters and then her vision OS worsened several weeks later. She was seen by an ophthalmologist and a neurologist who noted bilateral optic nerve swelling. Her vision was 20/70 and 20/800 with an APD OS. She was thought to have either IIH or optic neuritis.

Because of the diagnostic uncertainty, she was referred for further evaluation.

Case report and review of the literature.

In October, 2007, her visual acuities were 20/40 and 20/200. Visual fields showed a central scotoma OS and generalized field loss OU. She still had bilateral disc swelling. She had vibratory sensation loss in the right foot and an up-going toe on the right.

Her spinal tap OP was >550 mm Hg. CSF studies showed 4 WBC, 46 RBC, TP 71, glucose 87, +16 oligoclonal bands and IgG index +20.4. Cytology was negative.

She had another course of IV steroids and a VP shunt was placed.

There was a high suspicion she had multiple sclerosis in addition to IIH.

After the VP shunt was placed, her vision stabilized at 20/40 OD and 20/400 OS.

One year later, her older sister noticed transient vision loss OS.

She had no headaches. Her VA were 20/20 OD and 20/200 OS with an APD OS. Fundus exam showed bilateral disc swelling.

HVF showed bilateral blind spot enlargement.

Her spinal tap OP was 370 mmHg. CSF IgG synthesis rate of +63, IgG index +2.48 and > 5 OCB were present. Cytology was negative. She was started on acetazolamide 500mg bid. Her vision stabilized at 20/20 and 20/60, and her papilledema resolved.

The sisters had a paternal great-aunt with unexplained blindness.

Conclusion:
The final diagnosis was familial multiple sclerosis presenting as familial intracranial hypertension.

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Key Words: papilledema, intracranial hypertension, vision loss, multiple sclerosis,

Financial Disclosure: Yes

Poster 99

Low Grade Lymphoma with Isolated Bilateral Optic Nerve Involvement

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Introduction:
CNS involvement with lymphoplasmacytic lymphoma with IgM paraproteinemia (previously known as Waldenstrom's macroglobulinemia) is usually a feature of more aggressive/transformed disease. We present an unusual case of indolent lymphoma causing isolated disc edema and bilateral optic neuropathy, with no evidence of disease transformation or systemic progression.

Methods:
Case report

Results:
Our patient was diagnosed with lymphoplasmacytic lymphoma based on bone marrow biopsy results and paraproteinemia. He developed chronic bilateral disc edema, with a left RAPD, decreased central vision and severely constricted visual fields. He had no increased intracranial pressure or constitutional symptoms, and was otherwise well. MRI brain and orbits with and without contrast showed normal brain parenchyma and meninges, but the optic nerve-sheath complexes were thickened and edematous. Lumbar puncture showed a normal opening pressure of 15 cmH20. Molecular analysis of CSF and peripheral blood showed the same B-cell clone in both anatomical sites. Therefore, dexamethasone and weekly intrathecal methotrexate was started as a diagnostic trial. There was complete resolution of the papilledema, with segmental pallor OD, and secondary atrophy OS.

Conclusion:
Low grade lymphoma can present with CNS involvement limited to the optic nerve-sheath complex. Disc edema with visual loss should not be left untreated, even if the etiology is not well understood, since permanent visual loss can ensue. In the setting of Waldenstrom's macroglobulinemia, disc edema is not always due to hyperviscosity, central retinal vein occlusion, IgM deposition, or clogging of the arachnoid villi with increased intracranial pressure.

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Key Words: disc edema, optic neuropathy, complications of cancer, lymphoma, Waldenstrom's macroglobulinemia

Financial Disclosure: NONE

Poster 100

Intravascular Lymphoma Presenting as a Posterior Ischemic Optic Neuropathy

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Introduction:
Intravascular lymphoma (IVL) is a rare condition in which lymphoma is confined to the small vessels of brain and skin resulting in ischemia. We report a case in which IVL presented as an acute posterior optic neuropathy.

Methods:
Case report and review of the literature.

Results:
A 68 year old experienced painless visual loss OS for 5 hours. MRI showed a small left parietal DWI+ infarct and a right cerebellar T2 bright lesion. Painless visual loss recurred OS 9 days later and persisted. There was an inferior field defect OD, severe diffuse field loss OS. A papillitis and arteriole attenuation were noted OS. ESR and CRP were negative. She was diagnosed with a non-arteritic posterior ischemic optic neuropathy (PION) OS. Vision worsened over weeks and optic nerve pallor developed. Orbital MRI showed enhancement of the left intra-orbital optic nerve.

MRI brain showed T2-hyperintense lesions in the corpus callosum and some enhancement over the left parietal infarct. Orbital MRI showed persistent peri-neural enhancement OS at 6 months.
Eleven months after presentation, confusion, urinary incontinence and ataxia developed with progression of brain lesions. CSF cytology / flow cytometry were negative twice. Cerebellar biopsy revealed neoplastic lymphoid cells present only within vascular spaces consistent IVL. Surrounding tissue showed foci of infarction with complete loss of Purkinje cells. Neurological symptoms and lesions improved with chemotherapy.

Conclusion:
IVL may cause cortical infarcts that do not conform to large vessel territories. Since brain lesions were already present at the time of the optic neuropathy, it is probable that its cause was occlusion of small vessels to the optic nerve by the disease. This is one of the first reports in the literature of IVL associated with an optic neuropathy and helps expand the spectrum of both IVL and PION.

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Key Words: Ischemic Optic Neuropathy, Intravascular Lymphoma, Neoplastic Optic Neuropathy, Optic Nerve Head Drusen, Transient Monocular Blindness

Financial Disclosure: Yes

Poster 101

Radiation Induced Cavernous Hemangioma of the Oculomotor Nerve

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Introduction:
A 63 year old woman presented with progressive double vision over 1 year. Her past medical history was remarkable for a pituitary tumor diagnosed in the 1970's. She underwent transphenoidal resection in 1979 and proton beam irradiation in 1982.

Methods:
On examination her best corrected vision was 20/20. There was 3mm of ptosis involving the left upper lid and the left pupil was 1.5mm greater than the right and poorly reactive to light. There was no RAPD. In primary position the eye was 35 diopters exotropic and 16 diopters hypotropic. Ductions revealed a significant elevation deficit, a mild depression deficit and a mild adduction deficit.

The patient was diagnosed with a partial third nerve palsy on the left. Serial MRI studies showed progressive enlargement of a suprasellar lesion.

Results:
A left pterional craniotomy and exploration of the left third nerve was performed. Where the third nerve enters the roof of the cavernous sinus, a red-purple, multilobulated mass was seen within the nerve. Nerve fibers were splayed circumferentially around the lesion, which by appearance was very typical of a cavernous hemangioma. No dissecting plane could be found and the surgery was terminated without complication.

Conclusion:
Radiation induced cavernous hemangioma of the central nervous system is a rare entity. Occuring primarily in children, it is seen on average 9 years following radiotherapy most commonly for medulloblastoma, glioma or ALL. None of the cases reported involved a cranial nerve. Our patient presented with a progressive third nerve palsy. On direct visualization, a cavernous hemangioma of the oculomotor nerve was found.

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Key Words: Third cranial nerve palsy, Cavernous hemangioma, Diplopia, Radiation treatment

Financial Disclosure: None

Poster 102

Progressive Visual Loss in an Adult with an Optic Pathway Glioma

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Introduction:
Low grade gliomas of the visual pathway, termed optic pathway glioma (OPG), occur most frequently in children with Neurofibromatosis type 1 (NF1). OPG also occur sporadically, but regardless of etiology, they frequently do not become symptomatic in adulthood. We report a case of a 42 year old male with a newly symptomatic OPG.

Methods:
A 43 year old police officer and expert marksman, noticed vision in his right eye had changed over one year. He sought evaluation and was diagnosed as a glaucoma suspect based on increased optic cup size. The patient continued to have worsening vision and eventually noticed a significant field defect. Neuro-ophthalmologic consultation was sought when a glaucoma specialist was concerned about the hemianopic character of his visual field defect.

Results:
On neuro-ophthalmologic examination, his best corrected visual acuity was 20/40 OD and 20/25 OS. 11/15 Ishihara color plates were identified OU. Automated perimetry revealed a superior bitemporal quadrantanopsia. There was a trace afferent pupillary defect OD. Dilated fundus exam revealed mild pallor of the optic rim OU. MRI revealed a non-enhancing lesion involving the optic chiasm and hypothalamus. Biopsy of the mass surrounding the right optic nerve revealed a WHO Grade I juvenile pilocytic astrocytoma. Genetic testing for NF1 was normal.

Conclusion:
This case highlights new onset vision loss from an OPG in a 42 year old male without NF1. To our knowledge, this is the oldest individual to report new onset vision loss from an OPG. In a retrospective analysis of adult patients with OPG, Ellis and colleagues (2009) reported new visual symptoms in adults in their third decade, but no one in their fifth decade. Given these prior reports and the findings in our patient, any new visual complaint should prompt a thorough neuro-ophthalmologic exam in all patients with OPG, regardless of age.

References: None

Key Words: Optic nerve tumors, Vision loss, Optic atrophy, Optic chiasm

Financial Disclosure: None

Poster 103

Compressive Optic Neuropathy from Renal Osteodystrophy Arrested after Parathyroidectomy

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Introduction:
Chronic renal failure (CRF) cause excessive bone resorption and new bone formation called renal osteodystrophy (RO). Calavarial hypertrophy and narrowing of neural canals and foramina are features. Parathyroidectomy stabilizes RO. Uremic leontiasis ossea is a rare, severe form of RO with distinctive facial features: prominent maxilla and mandible, widely spaced teeth, widened nares, and flattened nasal bridge. We report bilateral compressive optic neuropathy secondary to RO.

Methods:
A 23 year-old woman with Branchio-Oto-Renal syndrome and CRF who developed insidious bilateral blurred vision and occasional headaches 3 months earlier was evaluated for optic disc swelling and visual loss.

Results:
Examination showed prominent cheekbones, widened nares and flattened nasal bridge. Visual acuity was finger counting at 6 feet OU. Pupils measured 4 mm with bilateral afferent defects. Color vision was 2/17 Ishihara plates OU. Funduscopic examination revealed elevated nasal disc margins, temporal pallor and no spontaneous venous pulsations OU. Humphrey fields revealed central scotomas and intact peripheral fields in both eyes. Hearing was moderately impaired bilaterally. Lumbar puncture opening pressure and routine CSF studies were normal. MRI revealed narrowing of optic canals with bilateral optic nerve compression. While awaiting neurosurgical evaluation, she had partial parathyroidectomy. Her vision spontaneously improved to 20/200 OD and 20/70 OS with some improvement in the visual fields at follow-up 5 months after symptom onset.

Conclusion:
Our patient had bilateral compressive optic neuropathy resulting from RO with features of leontiasis ossea. Simultaneous involvement of both optic nerves caused by RO can be confused with intracranial hypertension from parathyroid disorders. There are no clear indications for decompression surgery in compressive optic neuropathies caused by thickened optic canals, but are probably limited to acute loss or progressive deterioration of vision. Parathyroidectomy probably relieved the optic compression and resulted in the visual improvement in our case.

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Key Words: Compressive optic neuropathy, Renal osteodystrophy, Optic canal narrowing, Branchio-Oto-Renal syndrome, Uremic leontiasis ossea

Financial Disclosure: None

Poster 104

The Depths of Darkness

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Introduction:
Creutzfeldt-Jakob disease (CJD) is a rare, fatal neurological degenerative disease associated with a rapid, progressive spongiform encephalopathy. A proteinaceous infectious agent (prion) is believed to be the cause of this disease. Heidenhain variant of CJD frequently presents with vague visual symptoms, which can produce diagnostic uncertainty.

Methods:
A 67-year-old woman with history of migraine presented with 3 weeks of "dark" vision in both eyes on May 17, 2007. An initial MRI on was unrevealing. The Neuro-ophthalmic examination showed the corrected visual acuities were 20/15 OD and 20/15 OS with full colour vision. In the following 2-3 months, her visual acuities worsened to 20/40 OD and 20/50 OS, then 20/400 OU and dyschromatopsia developed OU. She also developed difficulty with memory and naming objects. She was hospitalized because of the progressive vision loss and decline of mental status. On admission, she was "blind with normally reactive pupils". Neurological examination confirmed subnormal memory and found a slightly ataxic gait. During the course of admission, her speech and memory progressively declined. She developed myoclonic jerks and marked disorientation, and then became non-verbal and unresponsive. On August 28, four months after the onset of visual symptoms, she expired.

Results:
The diagnosis of Heidenhain variant of CJD was suggested by followings: 1) an EEG with frequent sharp waves bilaterally in the occipital area with some theta and delta activity; and 2) elevated CSF protein with positive 14-3-3 protein. In addition, the diffusion-weighted image showed "possible" hyperintensity diffusely in the occipital cortex bilaterally, which was thought to be consistent with the diagnosis of CJD.

Conclusion:
The report of "dark" vision is interesting and cannot be specifically explained, other than perhaps reflecting some type of higher cortical abnormality. Heidenhain variant of CJD should be considered in any adult with unexplained bilateral visual loss who develops a change in mental status.

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Key Words: Visual loss, Occipital cortex, MRI, EEG, CSF

Financial Disclosure: None