Pearls, Pitfalls and Advances in Neuro-Ophthalmology

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Where? What? Now What?
Acute retinal ischemia
Different visual outcomes
Same systemic implications

<table>
<thead>
<tr>
<th>Study</th>
<th>MRI Results</th>
<th>Correlation</th>
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<tbody>
<tr>
<td>Boston 2012</td>
<td>DWI+ in 31/129 (24%)</td>
<td>Neuro sx+ Permanet VL &gt; TMVL</td>
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<td></td>
<td>Same vascular territory as visual loss in 28/31</td>
<td>Identified cause</td>
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<td></td>
<td>Small, multiple infarctions</td>
<td>Embolic cause</td>
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<td>Korea 2014</td>
<td>DWI+ in 8/33 (24.2%)</td>
<td>Neuro sx+</td>
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<td></td>
<td>Same vascular territory as visual loss in 8/8</td>
<td>CRAO &gt; BRAO</td>
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<td>Identified cause</td>
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<td>Embolic cause</td>
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<tr>
<td>Germany 2015</td>
<td>DWI+ in 49/213 (23%)</td>
<td>Neuro sx+</td>
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<td>Same vascular territory as visual loss in 55%</td>
<td>Identified cause</td>
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<td>Small, multiple infarctions</td>
<td>Embolic cause</td>
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Non-mydriatic fundus cameras

- Easy for non-ophthalmic trained individuals to use
- No pupillary dilation
- Able to take quality photographs of the posterior pole
- Reveals unrecognized findings in ED (Bruce et al. NEJM 2011; 364:387-9)
Optic Neuropathy

Classic Features

- Decreased visual acuity
- Abnormal visual field
- Relative afferent pupillary defect
- Can see through to the nerve
- Swollen or pale optic nerve

Optic Neuropathy

Disc Alternatives

Optic Neuropathy

Causes

- Inflammatory
- Vascular
- Compressive/Infiltrative
- Toxic/Nutritional
- Hereditary
- Traumatic
- Elevated intracranial pressure
- Elevated intraocular pressure
### Optic Neuropathy

**Causes**
- Inflammatory
- Vascular
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### Papilledema

**Causes**
- Intracranial mass lesions
- Hydrocephalus
- Meningeal processes
- Cerebral venous thrombosis
- Idiopathic (pseudotumor cerebri)

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### Idiopathic intracranial hypertension

**Causes**
- Papilledema
- Headaches
- No localizing neurologic symptoms/signs except for VIth
- No intracranial process, no venous sinus thrombosis
- Normal CSF contents
- **CSF opening pressure ≥25cm H₂O**
• Elevated ICP measured in the lateral decubitus position: neonates: >76 mm H2O, age 1–18 years: >280 mm H2O
• Normal CSF composition except in neonates who may have up to 19 WBC/mm³ if 0–28 days and up to 9 WBC/mm³ if between 29 and 56 days old; the protein may be as high as 150 mg/dl

IIH in 2017 - News

- Not just a diagnosis of exclusion
- New diagnostic criteria
  - Papilledema
  - Measure of intracranial pressure
  - Neuroimaging findings

IIH: Poor visual prognosis

- Patient’s characteristics
  - Black race. Neurology 2008; 70: 861-7
  - Severe obesity. J Neuro-Ophthalmol 2013; 33: 4-8
  - Anemia / sleep apnea syndrome / HTN
IIH in 2017 - News

IIH is everywhere there are obese people

Age-standardized prevalence of obesity in men aged 18 years and over (BMI ≥ 30 kg/m²), 2014

Low energy diet and intracranial pressure in women with idiopathic intracranial hypertension: prospective cohort study

Conclusions: Women with idiopathic intracranial hypertension who followed a low energy diet for three months had significantly reduced intracranial pressure compared with pressure measured in the three months before the diet, as well as improved symptoms and reduced papilledema. These reductions persisted for three months after they stopped the diet.

Bariatric surgery for the treatment of idiopathic intracranial hypertension

Effect of acetazolamide on visual function in patients with idiopathic intracranial hypertension and mild visual loss: the Idiopathic Intracranial Hypertension Treatment Trial

Quality of life in idiopathic intracranial hypertension at diagnosis

IHH Treatment Trial results

ABSTRACT

Objective: The study purpose was to examine vision-specific and overall health-related quality of life (QOL) at baseline in Idiopathic Intracranial Hypertension Treatment Trial patients who were newly diagnosed and had mild visual loss. We also sought to determine the associations between vision-specific QOL, visual scores and visual symptoms, visual function, pain, headache-related disability, and obesity.

Methods: We assessed QOL using the 36-Item Short Form Health Survey, National Eye Institute Visual Function Questionnaire-25 (NEI-VFQ-25), and 10-item NEI-VFQ-5 Neuro-Ophthalmic Quality of Life Index (BHI) QOL studies. We assessed relationships between QOL and other clinical characteristics. Results: Among 165 participants with IHH (161 women and 4 men with a mean age of 55.2 ± 7.5 years), visual QOL scores were reduced compared with published values for disease-free controls. Scores of participants were comparable to published values for patients with multiple sclerosis and an history of optic atrophy. A multiple linear regression model for the NEI-VFQ-25 composite score found that while age, sex, and visual scores were associated with QOL, no other predictors were associated with QOL.

Conclusions: IHH affects QOL at the time of diagnosis even in patients with mild visual impairment. Vision-specific QOL in patients with newly diagnosed IHH may be as decreased as that for patients with other neuro-ophthalmic disorders. IHH treatment should target visual loss and other symptoms of increased intracranial pressure associated with reduced QOL. Reduced QOL does not simply reflect obesity, an underlying IHH risk factor.
Clinical course of idiopathic intracranial hypertension with transverse sinus stenosis

Neurology 2013;80:289-95
Optic Neuropathy
Typical Optic Neuritis

- Inflammation of the optic nerve
- F:M 3:1
- Age: 15-45
- Pain on eye movement
- Normal or swollen disc
- Spontaneous improvement
- Associated with multiple sclerosis

ONTT

- No difference in visual acuity between steroid and placebo groups at 6 months.
- I.V. steroids may accelerate recovery by 2 to 3 weeks.
- P.O. steroids doubled the risk of recurrence in either eye.

(NEJM 326:581, 1992)
Clinical Features of Optic Neuritis with Low Risk of CDMS in Patients with No Brain MRI Lesions

No cases of CDMS have developed when any one of the following clinical features was present:

- Severe Disc Swelling (21 patients)
- Hemorrhage, disk or peripapillary (16 patients)
- Macular Exudates (8 patients)
- Painless (19 patients)
- No Light Perception (7 patients)

OCT: Retinal Nerve Fiber Layer (RNFL) Thickness

- Correlates with axonal loss
- Correlates with visual dysfunction
- Correlates with:
  - Brain atrophy in MS
  - Disability
  - Quality of life

Cellular composition of the retinal layers:
- ILM: inner limiting membrane
- RNFL: retinal nerve fiber layer
- GCL: ganglion cell layer
- IPL: inner plexiform layer
- INL: inner nuclear layer
- OPL: outer plexiform layer
- ONL: outer nuclear layer
- ELM: external limiting membrane
- IPS: inner photoreceptor segments
- OPS: outer photoreceptor segments
- PR: photoreceptors
- RPE: retinal pigment epithelium

Healthy subject. 3-dimensional macular volume cube generated by Cirrus HD-OCT from the macular region.

The individual layers of the retina are readily discernible, except for GCL and IPL, which are difficult to distinguish.

During the segmentation process (performed in 3-dimension), the segmentation software identifies the outer boundaries of the macular RNFL, IPL, and OPL, as well as the inner boundary of the RPE, which is identified by the conventional Cirrus HD-OCT algorithm. The identification of these boundaries facilitates OCT segmentation, enabling determination of the thicknesses of the macular RNFL, GCL + IPL, the INL + OPL, and the ONL including the inner and outer photoreceptor segments.
Fingolimod and Macular Edema

1. Incidence of macular edema is low (~1%); (uveitis, DM increase risk).
   Ophthalmology 2013; 120: 1432-1439

2. Screening evaluation for uveitis, macular or retinal vascular disease prior to starting, or within the first few weeks of starting fingolimod

3. Re-evaluation (complete eye exam +/- macular OCT) at 3-4 months of therapy (most reported cases of macular edema occurred within 3-4 months)

Optic Neuropathy

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Optic Neuritis and NMO Abs

- Bilateral
- Severe
- Poor recovery
- Recurrent

Should Most Patients With Optic Neuritis be Tested for Neuromyelitis Optica Antibodies and Should This Affect Their Treatment?

J Neurol Ophthalmol 2010; 30: 375-379