“I’S” IN NEURO-OPTOMETRY: ABNORMAL INTRACRANIAL PRESSURES

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DISCLOSURES

No financial relationship with any company or products mentioned in this presentation

The “I’s” in Neuro-Optometry

Iatrogenic #1: Medication Related Conditions
Iatrogenic #2: Perioperative Vision Loss
Inflammatory Thyroid Orbitopathy
Autoimmune Conditions
Abnormal Intracranial Pressures

OVERVIEW

Discuss the pathophysiology of intracranial hypertension and hypotension
Review the signs, symptoms, and ocular sequelae of abnormal intracranial pressures
Discuss proper diagnosis, treatment, and management

INTRACRANIAL HYPERTENSION

Abnormal Intracranial Pressures

Intracranial Hypertension

Intracranial Hypotension

Idiopathic Intracranial Hypertension (IHH) or Primary Pseudotumor Cerebri Syndrome
Secondary Intracranial Hypertension (SIHH) or Secondary Pseudotumor Cerebri Syndrome
IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH) PRIMARY PSEUDOTUMOR CEREBRI SYNDROME

Epidemiology
- Worldwide incidence of 12-20 per 100,000 per year for obese and child-bearing age women
- 0.5 to 2 per 100,000 per year in general population
- Boys and girls equally affected before puberty
- Male incidence 0.3 per 100,000 per year; 1.5 per 100,000 per year if obese
- Women are 9x more affected after puberty

Pathogenesis
- Unclear definitive pathogenesis, but several working theories:
  - Excess cerebrospinal fluid (CSF) production
  - Higher cerebral blood volume or brain water content
  - CSF obstruction and increased venous sinus
  - Endocrinological and metabolic factors

INTRACRANIAL HYPERTENSION (IH) SECONDARY PSEUDOTUMOR CEREBRI SYNDROME

Medication-Related
- Vitamin A
- Isotretinoin
- Antibiotics (tetracyclines, minocycline)
- Nalidixic acid
- Sulfur drugs
- Oral contraceptives
- Lithium
- Corticosteroid withdrawal

Systemic Conditions
- Endocrine disorders (Addison’s disease, hyperthyroidism, hypothyroidism)
- Hypercapnia (obstructive sleep apnea)
- Anemia
- Uremia
- Systemic Lupus Erythematosus

Venous Obstruction
- Cerebral venous sinus thrombosis (CVST)
- Hypercoagulability
- Arteriovenous fistulas
- Tumor compression
- Iatrogenic disruption of venous drainage

CLINICAL PRESENTATION

Symptoms
- Headaches (most common symptom)
- Transient visual symptoms
- Horizontal binocular diplopia (CN VI palsy)
- Pulsatile tinnitus
- Other: numbness, incoordination, decreased sense of smell, dizziness, and weakness

Signs
- Visual field defects
- Enlarged blind spot
- Localized nerve fiber bundle defect
- Arcuate defect
- Visual acuity is preserved until late stages
- Optic disc edema (hallmark sign)

OPTIC DISC EDEMA

- Optic disc edema: swelling of the optic disc from ANY cause (includes papilledema)
- Papilledema: swelling of the optic disc from increased intracranial pressure (confirmed by lumbar puncture)

PATHOPHYSIOLOGY OF PAPILLEDEMA

Increased intracranial pressure
- Axoplasmic flow stasis at lamina cribosa
- Swelling of optic nerve fibers and optic disc
- Compression of low-pressure venules
- Venous stasis and fluid leakage resulting in accumulation of extracellular fluid

FRISÉN GRADING SCALE

Grade 1: Mild disc elevation with c-shaped edema
Grade 2: Circumpapillary halo with folds (Paton’s line)
Grade 3: Obliteration of major blood vessels as they leave the disc
Grade 4: Loss of major blood vessels on the disc
Grade 5: Obliteration of blood vessels & obliteration of optic cup

Photo credit: http://eyerounds.org/article/IIH/pc_4.htm

Photo credit: https://www.eyerounds.org/cases/papilledema-grading.htm#g1
### DIAGNOSTIC CRITERIA

**Optic Disc Edema?**

- **YES**
- **NO**

**CNVI Palsy?**

- **YES**
- **NO**

**Definitive Diagnosis**

- Normal neurologic exam
- Normal neuroimaging on CT/MRI
- Normal CSF composition
- Elevated lumbar pressure:
  - ≥ 250mm H₂O in adults;
  - ≥ 280mm H₂O in children

**Suggests Diagnosis**

- Normal neurologic exam
- Neuroimaging showing at least one of the following:
  1. Empty Sella
  2. Flattening of the posterior aspect of the globe
  3. Distention of the peripapillary arachnoidal space with or without a tortuous optic nerve
  4. Transverse venous sinus stenosis
- Normal CSF composition
- Elevated lumbar pressure:
  - ≥ 250mm H₂O in adults;
  - ≥ 280mm H₂O in children

### DIAGNOSTIC TESTING

**Neuroradiography**

- Preferred: MRI with and without contrast
- Rule-out any underlying disease/intracranial mass
- Cerebral venous sinus thrombosis (CVST) may present similarly
- Must do MRI prior to LP to rule out intracranial mass

**Lumbar Puncture (LP)**

- Required to measure opening pressure and examine CSF contents
- Elevated pressures:
  - ≥ 250mm H₂O in adults
  - ≥ 280mm H₂O in children

### OPTICAL COHERENCE TOMOGRAPHY (OCT)

- Monitor for optic disc improvement and differentiate from pseudo-papilledema

<table>
<thead>
<tr>
<th>Condition</th>
<th>Contour</th>
<th>Hypo-Reflective Space</th>
<th>RNFL Thickness</th>
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<tbody>
<tr>
<td>Pseudo-papilledema (Optic nerve head drusen)</td>
<td><img src="image" alt="Contour" /></td>
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### VISUAL FIELD TESTING

- Enlarged Blind Spot
- Inferior Arcuate & Enlarged Blind Spot
- Superior Arcuate & Enlarged Blind Spot

### THE IDIOPATHIC INTRACRANIAL HYPERTENSION TREATMENT TRIAL (IIHTT)

- **Purpose:** assess efficacy of acetazolamide and weight loss in improving vision loss
- 165 patients (161 women, 4 men)
- 2 treatment groups:
  - Acetazolamide and weight management
  - Placebo and weight management
- Primary outcome measure = Perimetric Mean Deviation (PMD)
  - Average baseline PMD of -3.53dB in study eye, -2.28dB in fellow eye
  - 6 months:
    - Treatment: -2.10dB in study eye, -1.41dB in fellow eye
    - Placebo: -2.82dB in study eye, -1.86dB in fellow eye
- **Conclusion:** acetazolamide and weight management improved visual outcomes

### TREATMENT & MANAGEMENT

**Medical Treatment**

- Acetazolamide with weight/dietary management (gold standard)
- Topiramate (Topamax)

**Surgical Treatment**

- Considered if there is progressive vision loss and/or poor headache management
- Optic nerve sheath fenestration (ONSF)
- CSF shunting
- Venous sinus stenting
- Bariatric surgery (weight management)
CASE #1: 30YO WF

Chief complaint: Temporal blurry spot in each eye, constant headaches, and can hear blood whooshing behind her ears; began one week ago

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Equal, Round, 3+ reactive Pupils Equal, Round, 3+ reactive

FTFC CVF FTFC

Unremarkable Anterior Segments Unremarkable

13month (GAT) Tonometry 13month (GAT)

See photos Posterior Segments See photos

CASE #1: 30YO WF

Assessment and Plan:
- Bilateral Optic Disc Edema
- Bilateral scotomas in temporal visual field, pulsatile tinnitus, constant dull headaches and retrobulbar pressure for one week
- Weight gain of 15 pounds in past 3 months
- High suspicion of Idiopathic Intracranial Hypertension
  - sent to ED for urgent MRI/neurology consultation and possible lumbar puncture
- Follow-up in Optometry within 1-week for baseline HVF 24-2

MRI and RNF: Od Neuraxis Optic Disc Case 20x10 OD OS

Grade 3 Optic Disc Edema OD Grade 3 Optic Disc Edema OS

MRI with and without contrast:
- Negative for any abnormalities

Lumbar Puncture:
- Opening pressure: 370mm H2O, clear CSF contents
- Drained 10cc Closing pressure: 150mm H2O

Assessment/Plan:
- Idiopathic Intracranial Hypertension
- Begin acetazolamide 250mg BID PO until next neurology follow-up in 2-4 weeks
CASE #1: 30YO WF – 1 WEEK VF

- **Chief Complaint:** Less frequent headaches. No whooshing behind the ear. Still noticing bilateral temporal scotomas. Notes “speckles of glitter” which began 1-day after her LP. Currently taking acetazolamide 250mg BID PO.

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<tr>
<td>See HVF 24-2 VF See HVF 24-2</td>
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- **Assessment/Plan:** Bilateral optic disc edema with diagnosed Idiopathic Intracranial Hypertension
  - Recommended weight loss and continued use of acetazolamide 250mg BID PO
  - Continue care with Neurology
  - RTC 1-month repeat HVF 24-2 and DFE

CASE #1: 30YO WF – 3 MONTHS

- **Chief Complaint:** Was off acetazolamide for 4 weeks, recently restarted 2-weeks ago. Improvement in vision and headache symptoms.

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<tr>
<td>15mmHg (GAT) 15mmHg (GAT)</td>
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<td>See photos Posterior Segment See photos</td>
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Initial presentation: Grade 3 optic disc edema
3-month follow-up: Grade 1 optic disc edema

Initial presentation: Grade 3 optic disc edema
3-month follow-up: Grade 1 optic disc edema
CASE #1: 30YO WF – 3 MONTHS

- Assessment/Plan: Bilateral optic disc edema with diagnosed idiopathic intracranial hypertension
  - Lost about 15lb since initial diagnosis
  - Improvement in disc edema and visual fields
  - Subjective improvement in vision
  - Continue taking acetazolamide 250mg BID as directed by Neurology
  - RTC 2 months in Optometry for repeat HVF 24-2 and DFE

INTRACRANIAL HYPOTENSION

EPIDEMIOLOGY AND ETIOLOGY

<table>
<thead>
<tr>
<th>Epidemiology</th>
<th>Etiology</th>
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<tbody>
<tr>
<td>Incidence is estimated 5 per 100,000 per year</td>
<td>True hypervolemic state</td>
</tr>
<tr>
<td>Peak incidence around 40 years of age (may affect patients of any age)</td>
<td>Over-draining CSF shunts &amp; shunt complications</td>
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<tr>
<td>Increased prevalence in women</td>
<td>Traumatic CSF leaks</td>
</tr>
<tr>
<td>Female to male ratio of 1.5:1</td>
<td>Major injuries, aterogenic, post-surgical</td>
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<tr>
<td></td>
<td>Spontaneous CSF leaks</td>
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<td>Preexisting dural sac weakness</td>
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<tr>
<td></td>
<td>Connective tissue disorders (ie. Marfan’s Syndrome)</td>
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<td></td>
<td>Herniated discs/ spondylotic spurs</td>
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CLINICAL PRESENTATION

<table>
<thead>
<tr>
<th>Systemic Manifestations</th>
<th>Ocular Manifestations</th>
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<tbody>
<tr>
<td>Most common orthostatic headaches</td>
<td>Cranial nerve palsies (30-33%)</td>
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<tr>
<td>Spine pain</td>
<td>CN VI (80%) – 1 in 4 cases are bilateral</td>
</tr>
<tr>
<td>Cochleovestibular manifestations</td>
<td>CN III and CN IV (less common)</td>
</tr>
<tr>
<td>Nausea/vomiting</td>
<td>Decrease in visual acuity and visual field defects</td>
</tr>
<tr>
<td>Cognitive/behavioral changes</td>
<td>Nystagmus</td>
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<tr>
<td>Gait unsteadiness</td>
<td>Downbeat or gaze-evoked</td>
</tr>
<tr>
<td>Movement disorders</td>
<td>Upbeat nystagmus</td>
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</tbody>
</table>
CSF CIRCULATION

Lateral Ventricles
- Interventricular Foramen
- Third Ventricle
- Lateral Aperture
- Fourth Ventricle
- Medial Aperture
- Pontine Cistern
- Cerebomedullary Cistern
- Superior Cistern
- Interpeduncular Cistern
- Lumbar Cistern
- Subarachnoid Space
- Superior Sagittal Sinus
- Confluence of Sinus

CSF FLOW & CRANIAL NERVE PALSIES

HIGH ICP →
increased pressure/traction of nerve at the skull base

LOW ICP →
compression from brain sagging/sitting on nerves

INTERPEDUNCULAR CISTERN:
CN III

SUPERIOR CISTERN:
CN IV

PONTINE CISTERN:
CN VI

DIAGNOSTIC CRITERIA

International Classification of Headache Disorders, Third Edition (ICHD-3)

A. Headache fulfilling criterion 7.2 for headache attributed to low CSF pressure and criterion C below
B. Absence of a procedure or trauma known to be able to cause CSF leakage
C. Headache has developed in temporal relation to occurrence of low CSF pressure or CSF leakage, or has led to its discovery
D. Not better accounted for by another ICHD-3 diagnosis

Criteria 7.2

A. Any headache fulfilling criterion C
B. Either or both of the following:
   a. Low CSF pressure (<60mm H2O)
   b. Evidence of CSF leakage on imaging
C. Headache developed in relation to the low CSF pressure/leakage, or led to its discovery
D. Not better accounted for by another ICHD-3 diagnosis

Another set of diagnostic criteria has also been proposed which includes imaging if patients fail to satisfy ICHD-3 criteria:

1. Orthostatic headaches
2. The presence of at least ONE of the following:
   a. Low opening pressure (50mm H2O)
   b. Sustained improvement of symptoms after epidural blood patching
   c. Demonstration of an active spinal CSF leak
   d. Cranial MR changes of intracranial hypotension
3. No recent history of dural puncture
4. Not attributable to another disorder

NEUROIMAGING

- MRI with and without contrast of brain (and spine)
- Subdural fluid collections
- Enhancement of the pachymeninges
- Engorgement of the venous structures
- Pituitary engorgements
- Sagging of the brain
- Other techniques
  - Radioisotope cisternography
  - CT myelography
  - Digital subtraction myelography

TREATMENT, MANAGEMENT & COMPLICATIONS

Treatment & Management

- Conservative measures
  - Bed rest, coffee, hydration, time
  - Medications
  - Analgesics, corticosteroids, possibly Vitamin A, theophylline
  - Epidural blood patch and injections
  - Surgical repair of CSF leak

Complications

- Subdural hematoma
- Rebound intracranial hypertension
- Cerebral venous sinus thrombosis (CVT)
- Biventricular amyotrophy
- Superficial siderosis
- Syringomyelia
CASE #2: 71YO WM

- Chief Complaint: Recently seen by neurosurgery and ENT for:
  - Lumber decompression and ventriculoperitoneal shunt with repair for tegmen dehiscence with CSF leak
  - Reports positional headaches, new onset diplopia worse in left lateral gaze, and tastes fluid leaking from his ear

- Assessment/Plan:
  - RTC Optometry 2-weeks for diplopia follow-up
  - Patient to continue care with neurosurgery to manage CSF shunt leakage

- CN VI palsy resulting from intracranial hypotension due to post-surgical CSF leak
- MRI of brain with and without contrast: Normal
- CT of head/brain without contrast: shunt catheter is discontinuous, mild enlargement of the lateral ventricles (confirmed by neurosurgery)
- Follow-up with patients to manage diplopia symptoms
- Monitor for resolution of papilledema and visual field defects

Cranial Nerve Testing

<table>
<thead>
<tr>
<th>Case #2: 71YO WM</th>
<th>Case #2: 71YO WM – FOLLOW-UPS</th>
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<tbody>
<tr>
<td>Cover Text 10/12/2020</td>
<td>Cover Test 12/04/2020</td>
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<tr>
<td>18/1AE</td>
<td>15/4AET</td>
</tr>
<tr>
<td>20/1AE</td>
<td>14/2AET</td>
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<tr>
<td>30/1AE</td>
<td>13/1AET</td>
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<td>18/1AE</td>
<td>12/1AET</td>
</tr>
<tr>
<td>22/1AE</td>
<td>12/2P</td>
</tr>
<tr>
<td>Right head tilt: 35/1AET</td>
<td>Right head tilt: 12/1AET</td>
</tr>
<tr>
<td>Left head tilt: 25/1AET</td>
<td>Left head tilt: 14/1AET</td>
</tr>
<tr>
<td>Fresnel Prism: 15/1BO OS</td>
<td>Fresnel Prism: 15/1BO OS</td>
</tr>
<tr>
<td>EOM: -1 underaction left lateral gaze</td>
<td>EOM: -2 underaction left lateral gaze</td>
</tr>
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</table>

SUMMARY

- Pseudotumor cerebri syndrome is primarily seen in women of childbearing age and is becoming more prevalent due to the global obesity epidemic
- MRI with and without contrast of brain (and spine) is recommended to confirm diagnosis and rule-out any other signs of concomitant disease
- Cranial nerve palsies (most commonly CN VI) are seen with both high and low ICP
- Monitor for resolution of papilledema and visual field defects
- Follow-up with patients to manage diplopia symptoms

REFERENCES

THANK YOU!

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- Grace Tien, OD
- Russell Jee, OD
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QUESTIONS?

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