Idiopathic Intracranial Hypertension (Pseudotumor Cerebri) Management

- Idiopathic Intracranial Hypertension (IIH), also known as pseudotumor cerebri, is a neurological disorder attributable to elevated intracranial pressure. If not properly diagnosed and/or managed, IIH may lead to progressive – and possibly permanent – loss of vision. In addition, patients may be exposed to excessive radiation and/or may make frequent emergency department visits.
- The goals of IIH treatment are vision preservation, treatment of comorbid conditions (i.e., weight management), and symptom alleviation.
- Narcotics are NOT recommended for IIH headache.

### Algorithm 1. IIH New Suspected Diagnosis

#### Headache Triage

**Clinical suspicion or features concerning for increased ICP:**
- Holocranial or frontal headache, often worse when laying flat.
- Pulsatile tinnitus (“whooshing sound in ears”)
- Transient visual obscurations, particularly with position change

**Features suggestive of migraine:**
- Classic visual aura
- Episodic
- Photophobia
- Phonophobia
- Nausea/vomiting

**Excluding criteria:**
- Any cranial neuropathy other than CN II or CN VI
- Any other focal neurologic finding or symptom
- Trauma or altered LOC
- Sudden/worst headache of life
- Meningismus/fever

#### Ophthalmology Evaluation

Assessment and grading of optic disc edema

- Any grade bilateral edema (considered neuro ophthalmologic emergency)
- No edema: Consider alternative diagnosis (though increased ICP not excluded)

**Urine pregnancy test**

MRI brain w/o gadolinium

MRI venogram brain (MRV)

**Normal neuroimaging** with or without the following signs of increased ICP:
- Dilated optic nerve sheath
- Flattening of posterior globe
- Empty sella

**Lumbar puncture**

- Bedside LP attempted most experienced practitioner available, consider ultrasound guidance to minimize attempts and provide most accurate measurement. If patient BMI > 40 may proceed to fluoro without bedside attempt.
  - ICP measurement in lateral decubitus position (prone if fluoro)
  - CSF counts, protein, glucose, cytology, cultures*
  - *Do not discharge before counts are resulted

**Abnormal CSF studies** proceed with appropriate workup

**Abnormal neuroimaging** proceed with appropriate workup

**If lower risk/mild presentation, discuss with Neuro Ophthalmology to determine if Lumbar Puncture (LP) can be done on an outpatient basis (ED provider may enter outpatient order for LP under fluoro)**

#### Higher Risk Presentation

- Mild to moderate Papilledema + Elevated Opening Pressure, Normal CSF studies
  - Ophthalmology will provide dosing recommendations for medical management and outpatient follow-up
- Severe (vision threatening) Papilledema + Elevated Opening Pressure, Normal CSF studies
  - Admission to Ophthalmology service for initiation medical +/- surgical management, will involve Neurosurgery if indicated inpatient

---

*Copyright © 2015. The Ohio State University. All rights reserved. No part of this document may be reproduced, displayed, modified, or distributed in any form without a written agreement with the Ohio State University Technology Commercialization Office*
Clinical suspicion or features concerning for increased ICP:
- Holocranial or frontal headache, often worse when laying flat.
- Pulsatile tinnitus (“whooshing sound in ears”)
- Transient visual obscurations, particularly with position change

Features suggestive of migraine:
- Classic visual aura
- Episodic
- Photophobia
- Phonophobia
- Nausea/vomiting

“Red Flag” criteria:
- Any cranial neuropathy other than CN II or CN VI
- Any other focal neurologic finding or symptom
- Trauma or altered LOC
- Sudden/worst headache of life
- Meningismus/fever

Low Pressure Headache, patient with shunt or LP within the last two weeks:
- Positional headache often posterior head/neck worse when upright
- +/- nausea
- No fever
- No meningismus

Ophthalmology Evaluation
Assessment and grading of optic disc edema

Attempt non-narcotic medical management for migraine first

Inappropriate for IIH workup, proceed with standard emergent neuroimaging

Attempt conservative treatment:
- IV Fluids/Caffeine
- Non-narcotic pain meds

Neuroimaging required only if “Red Flag” criteria met

If symptoms resolve, discharge and recommend call to schedule outpatient follow-up

If patient has shunt and has fever, meningismus or pain at shunt site, obtain shunt series and consult Neurosurgery

Improved Symptoms?

If shunt is present:
- Neurosurgery consult
- Shunt change if possible

If recent LP:
- Consider blood patch

Patient has a shunt?

Medication titration per ophthalmology recommendation
- Outpatient follow up

Neuroimaging required only if “Red Flag” criteria met

Neurosurgery consult for empiric shunt adjustment if programmable.
- Addition of Diamox or Lasix
- Outpatient follow-up with ophthalmology and neurosurgery per their consult recommendations

Diagnostic/Therapeutic LP:
- Bedside LP attempted most experienced practitioner available, consider ultrasound guidance to minimize attempts and provide most accurate measurement.
- If patient BMI > 40 may proceed to fluoro without bedside attempt.
- Bedside LP attempt not required if LP shunt or if previously failed bedside attempt(s).

Patient has a shunt?

YES

Neurosurgery consult
- Consider shunt series
- Consider LP to evaluate opening pressure and consider shunt change. If elevated, it may indicate shunt failure.

NO

Diagnosed?

YES

NO

DC with outpatient follow-up

Copyright © 2015. The Ohio State University. All rights reserved. No part of this document may be reproduced, displayed, modified, or distributed in any form without a written agreement with the Ohio State University Technology Commercialization Office.
OSUWMC Resources

Guidelines

- Treatment of Acute Non-Life-Threatening Headache in the Emergency Department or Inpatient Settings
- Management of Spontaneous Intracerebral Hemorrhage (ICH) / Intraparenchymal Hemorrhage (IPH)
  - Increased Intracranial Pressure (ICP) Management Algorithm
- Management of Aneurysmal Subarachnoid Hemorrhage (SAH)

Ordersets

- OSU IP ED: Headache
- OSU IP ED: CDU/OBS Headache

Patient Education Materials

- Imaging Tests for Headaches-Choosing Wisely
- Pain Management (Krames):
  - Common Myths About Pain Medications
  - Medication for Pain
  - Managing Chronic Pain: Medications
- Lumbar Punctures (Krames):
  - What is a Lumbar Puncture
  - Having a Lumbar Puncture

Quality Measures

- Number of IIH patients treated
  - (% admitted, discharged, placed in observation)
- Percent of IIH patient who receive narcotics in the ED
- Percent of IIH patients with a Neurosurgery and/or Ophthalmology Consult in the ED
- Percent of IIH patients who return to the ED within 30 days
- Percent of IIH patients who receive CT
- Percent of IIH patients who receive LP
- Percent of IIH patients who receive Shuntogram
- Time spent in ED (LOS)

References

- Wall M. Idiopathic Intracranial Hypertension (pseudotumor cerebri). Current Neurology and Neuroscience Reports, 2008; 8: 87-93.

Guideline Authors

- Sue Bell, CNP
- Robert Cooper, MD
- John McGregor, MD
- Eric Bourekas, MD
- Abbe Craven, MD
- Eric Adkins, MD
- Peg Baylin, PharmD
- Erin Reichert, PharmD

Guideline Approved

November 29, 2017  Second Edition

Disclaimer: Clinical practice guidelines and algorithms at The Ohio State University Wexner Medical Center (OSUWMC) are standards that are intended to provide general guidance to clinicians. Patient choice and clinician judgment must remain central to the selection of diagnostic tests and therapy. OSUWMC’s guidelines and algorithms are reviewed periodically for consistency with new evidence; however, new developments may not be represented.
### Appendix A. Low-Pressure and High-Pressure Headache: Evaluation and Treatment

<table>
<thead>
<tr>
<th>Headache Type</th>
<th>Common Signs and Symptoms</th>
<th>Treatment Options</th>
</tr>
</thead>
</table>
| **Low-Pressure** | - Location of pain is posterior cervical  
- Feeling of achiness or throbbing  
- Pain is better when patient lies down  
- Nausea/vomiting  
- Blurry vision | - Fluids and caffeine  
- Medicinal options include:  
  - Antiemetics  
  - Headache cocktails  
- If shunt is programmable, consult neurosurgery for shunt change. |
| **High-Pressure** | - All-over headache  
- Headache feels explosive  
- Headache is worsened by coughing or sneezing  
- Tinnitus  
- Headache is not improved by lying down  
- Nausea/vomiting  
- Transient visual changes  
- Obscurations  
- Blurry vision  
- Double vision | - Consult ophthalmology:  
  - Patients with progressing visual loss may require surgical intervention with optic nerve sheath fenestration.  
- While the primary medication of choice is acetazolamide, other options include:  
  - Furosemide  
  - Topiramate  
- Shunt series, if shunt present or if applicable.  
- If shunt is programmable, consult neurosurgery for shunt change. |
| **All Patients Regardless of Headache Pressure Type** | | - Assign patient to primary care provider if they do not already have one for guidance on:  
  - Pain management  
  - Education  
- Provide counseling on steps for improved overall health:  
  - Weight loss  
    - Bariatric surgery should be offered only to select patients  
    - Please see OSUWMC surgical management of obesity guideline  
  - Improved nutrition  
- Psychological evaluation  
  - Depression  
    - Please see OSUWMC management of depression in adults guideline  
  - Anxiety |

* See OSUWMC [acute non-life threatening headache](https://www.osuwmc.edu) guideline

---

Copyright © 2015. The Ohio State University. All rights reserved. No part of this document may be reproduced, displayed, modified, or distributed in any form without a written agreement with the Ohio State University Technology Commercialization Office.
## Appendix B. Dosing Regimens for Treatment of Low-Pressure Headache

<table>
<thead>
<tr>
<th>Headache Type</th>
<th>Medication</th>
<th>Dose</th>
<th>Side Effects</th>
<th>Contraindications and Precautions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low-Pressure</td>
<td>Caffeine Citrate</td>
<td>480 mg IVPB over 60 minutes. May repeat</td>
<td>Angina, Palpitations, tachycardia, Ventricular arrhythmia, Nausea, vomiting, Insomnia, Dizziness, delirium</td>
<td>Pheochromocytoma, Significant cardiac history, History of arrhythmias</td>
</tr>
<tr>
<td></td>
<td>(Cafcit®)</td>
<td>after 6 hours x 1</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Metoclopramide</td>
<td>10 mg IM or IVP over 1 to 2 minutes. May</td>
<td>Extrapyramidal reactions (give with diphenhydramine or benztropine), Drowsiness, fatigue, Insomnia, Galactorrhea, amenorrhea, Hypotension, hypertension, Supraventricular tachycardia, bradycardia, Nausea, diarrhea, Urinary frequency, incontinence</td>
<td>Pheochromocytoma, Epilepsy, Parkinson’s disease</td>
</tr>
<tr>
<td></td>
<td>(Reglan®)</td>
<td>repeat in 8 hours as needed</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Prochlorperazine</td>
<td>10 mg IVP over 2 minutes</td>
<td>Constipation, dry mouth, Urinary retention, Drowsiness, Extrapyramidal reactions (give with diphenhydramine or benztropine), Cardiac arrhythmias</td>
<td>Parkinson’s disease, Pheochromocytoma, Myasthenia gravis</td>
</tr>
<tr>
<td></td>
<td>(Compazine®)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Valproate Sodium</td>
<td>500 mg IVPB over 30 minutes</td>
<td>Rash, Dizziness, Nystagmus, Somnolence, Tremor, Diplopia</td>
<td>Severe hepatic dysfunction, Known hypersensitivity to valproate sodium, Pregnancy</td>
</tr>
<tr>
<td></td>
<td>(Depacon®)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Magnesium Sulfate</td>
<td>1-2 grams IVPB over 30 to 60 minutes</td>
<td>Flushing, sweating, Hypotension, Depressed reflexes, Flaccid paralysis, Circulatory collapse</td>
<td>Caution in renal insufficiency</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Dexamethasone</td>
<td>10 mg IM or IVP over 3 to 5 minutes</td>
<td>Fluid and electrolyte disturbances, (hypokalemia), Muscle weakness, Peptic ulcer, Burning or tingling in the perineal, area after IV administration, Impaired wound healing, Convulsions, Psychiatric disturbances, Hyperglycemia, Increased intraocular pressure, Hypersensitivity reactions</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(Decadron®)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
## Appendix C. Outpatient Medications for Management of IIH Headaches

<table>
<thead>
<tr>
<th>Headache Type</th>
<th>Medication</th>
<th>Dose</th>
<th>Side Effects</th>
<th>Contraindications and Precautions</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Acetazolamide (Diamox®)</td>
<td>• 500 mg SR PO BID or 250 mg PO q6h</td>
<td>• Metabolic acidosis</td>
<td>• Hyponatremia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Max daily dose: 2000 mg in divided doses.</td>
<td>• Electrolyte disturbances</td>
<td>• Hypokalemia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Increased monitoring is required for dosages above 1000 mg due to risk of renal failure</td>
<td>• Taste alteration</td>
<td>• Significant kidney or liver disease</td>
</tr>
<tr>
<td></td>
<td>Furosemide (Lasix®)</td>
<td>• 20 mg PO daily or 20 mg PO BID (Max daily dose: 40 mg TID)</td>
<td>• Orthostatic hypotension</td>
<td>• Electrolyte abnormalities</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Hyperuricemia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Topiramate (Topamax®)</td>
<td>• 25 mg daily. May increase by 25 mg weekly.</td>
<td>• Paresthesia</td>
<td>• Pregnancy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(Max daily dose: 200 mg PO administered in two divided doses )</td>
<td>• Anorexia</td>
<td>• Hepatic impairment</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Somnolence</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Psychomotor slowing</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Abnormal vision</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Difficulty with memory</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Nausea, diarrhea</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Kidney stones (if used with other carbonic anhydrase inhibitors)</td>
<td></td>
</tr>
</tbody>
</table>

*Copyright © 2015. The Ohio State University. All rights reserved. No part of this document may be reproduced, displayed, modified, or distributed in any form without a written agreement with the Ohio State University Technology Commercialization Office*