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. Diagnosis and Management of Suspected IIH in New Patients

The following Modified Dandy Scale criteria are recommended when making a new diagnosis of IIH:
- Signs and symptoms of increased intracranial pressure.
- No localizing neurologic signs otherwise, with the single exception or unilateral or bilateral paresis in nerve VI.
- Cerebrospinal fluid can exhibit increased pressure but no cytologic or chemical abnormalities other than a low CSF protein which is very common in pseudotumor patients.
- Normal to small symmetric ventricles must be demonstrated.

#### Severe Findings
- Stage 4-5 papilledema
- Macular edema
- Retinal hemorrhages
- Acuity worse than 20/50 or change compared to baseline
- Marked visual field loss
- Rapid progression

#### Mild to Moderate Findings

**Mild:**
- Stage 1-2 papilledema
- Normal visual activity – transient obscurations of vision acceptable
- Normal visual field – except for enlarged blind spot

**Moderate:**
- Stage 3 papilledema
- Abnormal acuity
- Abnormal visual field – except for enlarged blind spot

#### MRI of head* if patient has not had a recent image within 6 months

- **Results consistent with IIH?**
  - **YES**
    - Lumbar puncture (LP)
    - Secondary etiology found, evaluate and treat accordingly.
    - Normal cerebrospinal fluid?
      - **YES**
        - Admit patient (ophthalmology)
        - Consult neurosurgery
      - **NO**
        - Secondary etiology found, evaluate and treat accordingly.
  - **NO**
    - Secondary etiology found, evaluate and treat accordingly.

- **MRI of head* if patient has not had a recent image within 6 months**

- **Results consistent with IIH?**
  - **YES**
    - Place patient in CDU for observation and treatment.
    - Manage as high pressure headache with medications recommended in **Appendix A** and **Appendix B**.
    - Do not use opioids
    - Outpatient follow up with ophthalmology and consideration for an outpatient LP.
  - **NO**
    - Secondary etiology found, evaluate and treat accordingly.

### Results consistent with IIH?

- **YES**
  - MRI of head* if patient has not had a recent image within 6 months
  - Secondary etiology found, evaluate and treat accordingly.
  - Normal cerebrospinal fluid?
    - **YES**
      - Admit patient (ophthalmology)
      - Consult neurosurgery
    - **NO**
      - Secondary etiology found, evaluate and treat accordingly.

- **NO**
  - Secondary etiology found, evaluate and treat accordingly.

**NOTE:** For patients who previously required LP under fluoroscopy, a bedside attempt at LP is NOT required prior to proceeding to fluoroscopy.

* CT scan if worse headache of patient’s life or atypical symptoms. If new diagnosis, full MRI / MRV often ordered.
Algorithm 2. Evaluation and Management of Established IIH Patients WITHOUT a Shunt

NOTE: Repeat neurological imaging is NOT recommended if the patient has had imaging (CT/MRI) done in the last 6 months, unless there are new neurological findings. For patients who previously required LP under fluoroscopy, a bedside attempt at LP is NOT required prior to proceeding to fluoroscopy.

Patient presents with headache symptoms

Patient presents with visual difficulties or changes in vision?

YES

Mild to Moderate Findings

Mild:
- Stage 1-2 papilledema
- Normal visual activity – transient obscurations of vision acceptable
- Normal visual field – except for enlarged blind spot

Moderate:
- Stage 3 papilledema
- Abnormal acuity
- Abnormal visual field – except for enlarged blind spot

Severe Findings

- Stage 4-5 papilledema
- Macular edema
- Retinal hemorrhages
- Acuity worse than 20/50 or change from baseline
- Marked visual field loss
- Rapid progression

MRI of head* if patient has not had a recent image within 6 months.
Perform LP (without fluoroscopy), if possible in the ED.

Admit patient (ophthalmology)
Consult neurosurgery

Normal cerebrospinal fluid?

NO
YES

Secondary etiology found – evaluate and treat accordingly.

Treat patient for IIH.

Do symptoms improve with use of medications?

NO
YES

PCRM schedule LP for within 7 days of discharge.
- If LP cannot be scheduled for within 7 days, consider completing procedure in the ED or increase dosage of outpatient medications in Appendix C.**
- Consider follow-up with ophthalmology and/or nutrition.

Discharge patient
Arrange for follow-up with ophthalmology and/or neurosurgery within three days.

* CT scan if worse headache of patient’s life or atypical symptoms. If new diagnosis, full MRI / MRV often ordered.
** Follow medication dosing instructions in Appendix C.
Algorithm 3. Evaluation and Management of Established IIH Patients WITH a Shunt

**NOTE:** Repeat neurological imaging is **NOT** recommended if the patient has had imaging (CT/MRI) done in the last 6 months, unless there are new neurological findings. Repeat shunt series is not recommended for patients with mild to moderate visual exam findings. For patients who previously required LP under fluoroscopy, a bedside attempt at LP is **NOT** required prior to proceeding to fluoroscopy.

1. **Patient presents with headache symptoms**
   - **High Pressure**
     - Patient presents with visual difficulties or changes in vision?
       - **NO**
         - Manage as headache
       - **YES**
         - Obtain Ophthalmology Consult
           - Make treatment decision based on visual exam findings.

2. **Severe Findings**
   - Stage 4-5 papilledema
   - Macular edema
   - Retinal hemorrhages
   - Acuity worse than 20/50 or change from baseline
   - Marked visual field loss
   - Rapid progression
   - MRI of head* if patient has not had a recent image within 6 months.
   - Perform shunt series if shunt has not been evaluated within the past month.
   - If shunt is programmable, consult neurosurgery for shunt adjustment.
   - Perform LP:
     - May perform without fluoroscopy in the ED if possible, unless patient previously required LP under fluoroscopy or patient has lumbar shunt.

3. **Mild to Moderate Findings**
   **Mild:**
   - Stage 1-2 papilledema
   - Normal visual activity – transient obscurations of vision acceptable
   - Normal visual field – except for enlarged blind spot
   **Moderate:**
   - Stage 3 papilledema
   - Abnormal acuity
   - Abnormal visual field – except for enlarged blind spot
   - Prescribe medications as indicated in Appendix A and B.
   - Do **not** use opioids
   - Perform shunt series if shunt has not been evaluated within the past month.
   - If shunt is programmable, consult neurosurgery for shunt adjustment.
   - If there is no sign of shunt problems such as dripping or abdominal pseudocysts due to fluid collection, keep patient as outpatient.
   - If patient has any signs of Carbapenem Resistant Enterobacteriaceae (CRE), an exposed shunt, or infection, admit the patient.

4. **Do symptoms improve with treatment?**
   - **NO**
     - Prescribe medications as indicated in Appendix A and B.
     - Perform shunt series if shunt has not been evaluated within the past month.
   - **YES**
     - Discharge patient.
      - Place patient in CDU for observation and treatment.
      - If headache is atypical, consult neurology for further treatment recommendations.

* CT scan if worse headache of patient’s life or atypical symptoms. If new diagnosis, full MRI / MRV often ordered.
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
- Percent of patients who receive narcotics
- Volume of patients seen in the ED
  - Number admitted
  - Number placed in CDU
  - Number discharged from ED
- Average time from ED visit to follow-up appointment
  - Ophthalmology
  - Neurosurgery

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

Guideline Approved
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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.

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## 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:  
  o Antiemetics  
  o 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:  
  o 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:  
  o Furosemide  
  o Topiramate  
  o Corticosteroids  
  • Shunt series, if shunt present or if applicable.  
  • If shunt is programmable, consult neurosurgery for shunt change. |
| All Patients | | • Assign patient to primary care provider if they do not already have one for guidance on:  
  o Pain management  
  o Education  
  • Provide counseling on steps for improved overall health:  
  o Weight loss  
  ▪ Bariatric surgery should be offered only to select patients  
  ▪ Please see OSUWMC surgical management of obesity guideline  
  o Improved nutrition  
  • Psychological evaluation  
  o Depression  
  ▪ Please see OSUWMC management of depression in adults guideline  
  o Anxiety |

* See OSUWMC acute non-life threatening headache guideline

** See OSUWMC acute non-life threatening headache guideline
## 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.</td>
<td>• Angina</td>
<td>• Pheochromocytoma</td>
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<tr>
<td></td>
<td>(Cafcit®)</td>
<td>• May repeat after 6 hours x 1</td>
<td>• Palpitations, tachycardia</td>
<td>• Significant cardiac history</td>
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<td></td>
<td></td>
<td></td>
<td>• Ventricular arrhythmia</td>
<td>• History of arrhythmias</td>
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<td></td>
<td></td>
<td></td>
<td>• Nausea, vomiting</td>
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<td></td>
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<td></td>
<td>• Insomnia</td>
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<td>• Dizziness, delirium</td>
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<td></td>
<td>Metoclopramide</td>
<td>• 10 mg IM or IVP over 1 to 2 minutes.</td>
<td>• Extrapyramidal reactions (give with diphenhydramine or benztropine)</td>
<td>• Pheochromocytoma</td>
</tr>
<tr>
<td></td>
<td>(Reglan®)</td>
<td>• May repeat in 8 hours as needed</td>
<td>• Drowsiness, fatigue</td>
<td>• Epilepsy</td>
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<td></td>
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<td></td>
<td>• Galactorrhea, amenorrhea</td>
<td>• Parkinson’s disease</td>
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<td></td>
<td></td>
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<td>• Hypotension, hypertension</td>
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<td></td>
<td></td>
<td></td>
<td>• Supraventricular tachycardia, bradycardia</td>
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<td></td>
<td></td>
<td></td>
<td>• Nausea, diarrhea</td>
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<td></td>
<td>• Urinary frequency, incontinence</td>
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<tr>
<td></td>
<td>Prochlorperazine</td>
<td>• 10 mg IVP over 2 minutes</td>
<td>• Constipation, dry mouth</td>
<td>• Parkinson’s disease</td>
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<tr>
<td></td>
<td>(Compazine®)</td>
<td></td>
<td>• Urinary retention</td>
<td>• Pheochromocytoma</td>
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<td></td>
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<td></td>
<td>• Drowsiness</td>
<td>• Myasthenia gravis</td>
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<td></td>
<td></td>
<td></td>
<td>• Extrapyramidal reactions (give with diphenhydramine or benztropine)</td>
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<tr>
<td></td>
<td>Valproate Sodium</td>
<td>• 500 mg IVPB over 30 minutes</td>
<td>• Rash</td>
<td>• Severe hepatic dysfunction</td>
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<td></td>
<td>(Depacon®)</td>
<td></td>
<td>• Dizziness</td>
<td>• Known hypersensitivity to valproate sodium</td>
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<td>• Nystagmus</td>
<td>• Pregnancy</td>
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<td>• Somnolence</td>
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<td></td>
<td>Magnesium Sulfate</td>
<td>• 1-2 grams IVPB over 30 to 60 minutes</td>
<td>• Flushing, sweating</td>
<td>• Caution in renal insufficiency</td>
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<td></td>
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<td></td>
<td>• Hypotension</td>
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<td></td>
<td>• Depressed reflexes</td>
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<td></td>
<td>• Flaccid paralysis</td>
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<td></td>
<td>Dexamethasone</td>
<td>• 10 mg IM or IVP over 3 to 5 minutes</td>
<td>• Fluid and electrolyte disturbances</td>
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<tr>
<td></td>
<td>(Decadron®)</td>
<td></td>
<td>• (hypokalemia)</td>
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<td></td>
<td></td>
<td></td>
<td>• Muscle weakness</td>
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<td>• Peptic ulcer</td>
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<td>• Burning or tingling in the perineal</td>
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<td></td>
<td></td>
<td></td>
<td>• area after IV administration</td>
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<td></td>
<td></td>
<td></td>
<td>• Impaired wound healing</td>
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<td></td>
<td>• Convulsions</td>
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<td></td>
<td>• Psychiatric disturbances</td>
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<td>• Hyperglycemia</td>
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<td></td>
<td>• Increased intraocular pressure</td>
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<td></td>
<td></td>
<td>• Hypersensitivity reactions</td>
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</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>High-Pressure</td>
<td>Acetazolamide</td>
<td>• 500 mg SR PO BID (or)</td>
<td>• Metabolic acidosis&lt;br&gt;• Electrolyte disturbances&lt;br&gt;• Taste alteration&lt;br&gt;• Tinnitus&lt;br&gt;• Polyuria&lt;br&gt;• Tingling paresthesia of the fingers and toes, and circumorally, are a near universal side effect.&lt;br&gt;• Consider Rx KCL 20mEQ/d for doses above 1 gm/d, or if muscle cramps develop.</td>
<td>• Hyponatremia&lt;br&gt;• Hypokalemia&lt;br&gt;• Significant kidney or liver disease&lt;br&gt;• History of kidney stones</td>
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<tr>
<td></td>
<td>(Diamox®)</td>
<td>• 250 mg PO q6h</td>
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<td>• Max daily dose: 2000 mg in divided doses.</td>
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<td>• Increased monitoring is required for dosages above 1000 mg due to risk of renal failure</td>
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<td></td>
<td>Furosemide</td>
<td>• 20 mg PO daily or 20 mg PO BID (Max daily dose: 40 mg TID)</td>
<td>• Orthostatic hypotension&lt;br&gt;• Hyperuricemia&lt;br&gt;• Hypokalemia&lt;br&gt;• Hypocalcemia&lt;br&gt;• Hypokalemia&lt;br&gt;• Hypomagnesemia</td>
<td>• Electrolyte abnormalities</td>
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<td></td>
<td>(Lasix®)</td>
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<td></td>
<td>Topiramate</td>
<td>• 25 mg daily. May increase by 25 mg weekly.&lt;br&gt;• (Max daily dose: 200 mg PO administered in two divided doses)</td>
<td>• Paresthesia&lt;br&gt;• Anorexia&lt;br&gt;• Somnolence&lt;br&gt;• Psychomotor slowing&lt;br&gt;• Abnormal vision&lt;br&gt;• Difficulty with memory&lt;br&gt;• Nausea, diarrhea&lt;br&gt;• Kidney stones (if used with other carbonic anhydrase inhibitors)</td>
<td>• Pregnancy&lt;br&gt;• Hepatic impairment</td>
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<tr>
<td></td>
<td>(Topamax®)</td>
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<tr>
<td></td>
<td>Prednisone</td>
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