Hydrocephalus: Emergency Department v.2.0

Assessment

• NPO
• Conduct complete physical exam including neuro exam
• Document Glasgow Coma Score

Unstable?

• NOTIFY ED ATTENDING IMMEDIATELY
  • Evaluate and stabilize: ABCD
  • Elevate head of bed to 30 degrees/head midline
  • Place on full monitors with BP to cycle every 15 minutes
  • If seizing initiate Seizure Acute Management Pathway
  • Notify Neurosurgery on call immediately
  • Proceed with assessment

Stable

Notify ED attending
For screening

Imaging

• DX shunt series
• CT Head w/o contrast STEALTH
• Abdominal signs or symptoms (acute abdomen)
  • Early (< 3 months of shunt placement): abdominal CT IV/PO contrast
  • Delayed (> 3 months of shunt placement): ultrasound abdomen – limited

Labs

• Concern for new hydrocephalus: CBC w/diff, PT/PTT
• Concern for shunt infection: CBC w/diff, ESR, CRP, serum glucose, BUN and serum creatinine, blood cultures.
  • Consider workup for other source of infection (could include respiratory panel, urinalysis, throat culture, urine culture, stool culture, chest XR if indicated)
• Concern for shunt malfunction: CBC w/diff

Notify Neurosurgery after labs are resulted

• Neurosurgery to determine if and when to begin antibiotics
• Determine if surgery needed
• Shunt/Reservoir tap per shunt tap protocol below
  • Shunt/Reservoir Tapping protocol
    • Tap to be done by Neurosurgery only
    • Tap if within 2 weeks of surgery with fever or
    • Tap if within 3 months of surgery with fever and no other source of infection
    • If > 3 months consider tap at neurosurgery discretion
    • If shunt/reservoir tapped send CSF for gram stain, culture, cell count, protein glucose, and bacterial PCR (use CSF Studies Orderset)

Surgery Needed

Surgery Preparation

• Start admission process
• Initiate admission orders
• Admit patient directly to OR or to inpatient unit—see ED Job Aid: ED-OR Transfer for Emergent or Urgent surgery (for SCH only)

Definitions

Clinical signs of shunt malfunction

• Children < 1 year old – bulging fontanelle, rapidly increasing OFC, downward deviation of the eyes, less interest in feeding
• Children > 1 year old – headaches, cranial nerve signs, changes in vision
• All ages – vomiting, lethargy, swelling along shunt tract, irritability without another recognized cause, lethargy
• Please note that an isolated seizure is not a prognostic indication of a shunt malfunction

Clinical signs of shunt infection

• Fever (38.5°C) and shunt placement within last 3 months and no other source of infection
• Fever (38.5°C) and shunt placement within the last 2 weeks
• Wound overlying the implanted shunt material
• Erythema, swelling or drainage from incisions
• Redness along the shunt tract

New Hydrocephalus

• Radiographic imaging with findings of new hydrocephalus

Unstable Patient

• Abnormal vital signs (bradycardia/ hypertension/ altered respiratory rate, obtunded/unresponsive)

Emergent Surgery: patient to OR in <1 hour

Urgent Surgery: patient to OR in 1-4 hours (can be transferred to inpatient unit)

Phase Change

• Postop Shunt Placement
• Postop ETV-CPC
• Postop Infection

For questions concerning this pathway, contact: Hydrocephalus@seattlechildrens.org

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Hydrocephalus: Clinic/Inpatient Admit v.2.0

**Inclusion Criteria**
- New hydrocephalus including scheduled referral, suspected shunt malfunction, or suspected shunt infection

**Exclusion Criteria**
- <1 Month of age, <44 weeks PMA
- New brain tumor
- Non-accidental trauma/truma
- Patients who undergo ETV who have had prior shunt

**Assessment**

- Send unstable patient to ER

**Imaging**
- New presentation hydrocephalus
  - CT brain without contrast STEALTH
  - MRI brain Hydrocephalus Protocol to evaluate if candidate for ETV, if indicated
- Shunt malfunction or shunt infection
  - CT brain without contrast STEALTH
  - If previous CT at Seattle Children’s: order low-dose CT
  - If clinically stable and has Strata valve, non-programmable valve, or no valve: order MRI brain HASTE
- XR Shunt Series
- Abdominal signs or symptoms (acute abdomen)
  - Early (<3 months of shunt replacement): abdominal CT IV/PO contrast
  - Delayed (>3 months of shunt placement): Ultrasound Abdomen – Limited

**Labs**
- Concern for new hydrocephalus: CBC w/ diff, PT/PTT
- Concern for shunt infection:
  - CBC w/ diff, ESR, CRP, serum glucose, BUN and serum creatinine, blood cultures
  - Consider workup for other source of infection (could include respiratory panel, urinalysis, throat culture, urine culture, stool culture, chest XR if indicated)
- Concern for shunt malfunction: CBC w/ diff
- Shunt/Reservoir tap per shunt tap protocol below
  - Shunt/Reservoir Tapping protocol
    - Tap to be done by Neurosurgery only
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    - Tap if within 3 months of surgery with fever and no other source of infection
    - If > 3 months consider tap at neurosurgery discretion
    - If shunt/reservoir tapped send CSF for gram stain, culture, cell count, protein glucose, and bacterial PCR (use CSF Studies Orderset)
- Determine if surgery needed

**Surgery Preparation**
- Start admission process
- Initiate admission orders
- Admit patient directly to OR or to inpatient unit (Emergent vs. Urgent)

**Definitions**

**Clinical signs of shunt malfunction**
- Children < 1 year old – bulging fontanelle, rapidly increasing OFC, downward deviation of the eyes, less interest in feeding
- Children > 1 year old – headaches, cranial nerve signs, changes in vision
- All ages – vomiting, lethargy, swelling along shunt tract, irritability without another recognized cause, lethargy
- Please note that an isolated seizure is not a prognostic indication of a shunt malfunction

**Clinical signs of shunt infection**
- Fever (38.5°C) and shunt placement within last 3 months and no other source of infection
- Fever (38.5°C) and shunt placement within the last 2 weeks
- Wound overlying the implanted shunt material
- Erythema, swelling or drainage from incisions
- Redness along the shunt tract

**New Hydrocephalus**
- Radiographic imaging with findings of new hydrocephalus

**Unstable Patient**
- Abnormal vital signs (bradycardia/ hypertension/ altered respiratory rate, obtunded/unresponsive)
Hydrocephalus: Postop Shunt Placement/Revision v.2.0

Management

- Patient transferred from OR to inpatient unit
- Vitals:
  - Neurological checks: q2 hours x 6 hours; q 4 hours till discharge
  - Monitors: Continuous cardiorespiratory monitors x 12 hours then only when asleep
- Activity: Ad Lib
- Nursing: see Hydrocephalus GOC (for SCH only)
  - Elevate head of bed to 30 degrees
  - Bathing: patient may bathe at 48 hours postoperative; no soaking
- Diet: Standard diet

- Fluids:
  - ≥ 1 month of age: D5NS + KCl 20 mEq/L
- Post-op Imaging:
  - CT scan
  - DX shunt series
- Medications
  - Miralax/Docusate scheduled, suppository or senna PRN
  - Pain:
    - Acetaminophen scheduled x 1 day then PRN starting in OR/PACU.
    - Alternate ibuprofen with acetaminophen, but delay starting ibuprofen until 4 hours postop to minimize bleeding risk
- Labs: none
- Place discharge orders (anticipated 24-48 hours)

Discharge Criteria

- Afebrile x 24 hours
- Tolerating PO/PO pain med
- No nausea or vomiting
- Tolerating up out of bed

Outpatient Follow-up

<table>
<thead>
<tr>
<th>Patient Age</th>
<th>Shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 5 years</td>
<td>Haste MRI yearly</td>
</tr>
<tr>
<td>≥ 5 years</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• Follow up every other year with a HASTE MRI and shunt series</td>
</tr>
<tr>
<td></td>
<td>• If patient has codman: CT every other year per provider discretion</td>
</tr>
</tbody>
</table>

Discharge Instructions

- Follow-up with neurosurgery NP in 2 weeks for wound check, and with neurosurgeon in 6 weeks for HASTE MR/CT

Inclusion Criteria

- Postop shunt placement

Exclusion Criteria

- < 1 Month of age, <44 weeks PMA
- New brain tumor
- Non-accidental trauma/trauma
- Patients who undergo ETV who have had prior shunt
Management

- Patient transferred from OR to inpatient unit
- Vitals:
  - Neurological checks: q2 hours x 6 hours; q 4 hours till discharge
  - Monitors: Continuous cardiorespiratory monitors x 12 hours then only when asleep
- Activity: Ad Lib
- Nursing: see Hydrocephalus GOC (for SCH only)
  - Elevate head of bed to 30 degrees
  - Bathing: patient may bathe at 48 hours postoperative; no soaking
- Diet: Standard diet

- Fluids:
  - ≥ 1 month of age: D5NS + KCl 20 mEq/L
- Post-op Imaging:
  - CT scan
  - DX shunt series
- Medications
  - Miralax/Docusate scheduled, suppository or senna PRN
  - Acetaminophen scheduled x 1 day then PRN starting in OR/PACU.
  - Alternate ibuprofen with acetaminophen, but delay starting ibuprofen until 24 hours postop to minimize bleeding risk
- Labs: Check sodium night of surgery and next AM
- Place discharge orders (anticipated 24-48 hours)

Discharge Criteria

- Afebrile x 24 hours
- Tolerating PO/PO pain med
- No nausea or vomiting
- Tolerating up out of bed

Discharge Instructions

- Follow-up with neurosurgery NP in 2 weeks for wound check, and with neurosurgeon in 6 weeks for HASTE MRI/CT

Inclusion Criteria

- Postop ETV/CPC

Exclusion Criteria

- <1 Month of age, <44 weeks PMA
- New brain tumor
- Non-accidental trauma/trauma
- Patients who undergo ETV who have had prior shunt

Outpatient Follow-up

<table>
<thead>
<tr>
<th>Patient Age</th>
<th>ETV</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 5 years</td>
<td>Haste MRI yearly</td>
</tr>
<tr>
<td>≥ 5 years</td>
<td>Order full CINE MRI first year, followed by HASTE MRI every other year when stable</td>
</tr>
</tbody>
</table>
Hydrocephalus: Suspected Shunt Infection v.2.0

Inclusion Criteria
- Postop suspected shunt or CSF infection

Exclusion Criteria
- <1 Month of age, <44 weeks PMA
- New brain tumor
- Non-accidental trauma/trauma
- Patients who undergo ETV who have had prior shunt

Management
- Patient transferred from OR to inpatient unit
- Vitals:
  - Neurological checks: q 2 hours x 6 hours then q 4 hours
  - Monitors: Continuous CR monitors
- Activity: Bedrest. Out of bed with drain clamps 30 mins x 3 times/day
- Nursing: see Hydrocephalus GOC and P&P: Ventriculostomy and Lumbar Drain Care (for SCH only)
  - PICC line
  - Elevate head of bed to 30 degrees
  - Bactiracin to insertion site BID
  - No bathing with EVD in place
  - Record strict I&O
- Refer to orders for EVD output parameters
- Diet: Regular Diet
- Fluids:
  - ≥1 month of age: D5NS +20KCl
  - Fluid replacement NS 1:1 for drain output
- Medications
  - Miralax/Docusate scheduled, suppository or senna PRN
  - Pain:
    - Acetaminophen scheduled x 1 day then PRN starting in OR/PACU
    - Oxycodone PRN for breakthrough pain
- Consult Infectious Disease
- Initiate antibiotics in consultation with ID

Monitor Response to Therapy
- Check CSF gram stain, culture, cell count, glucose, and protein
  - Obtain CSF from ventriculostomy catheter (not the bag).
  - Order daily until there are 3 negative cultures, then every Monday/Thursday
- Check CBC/differential daily for 7 days then Monday/Thursday
- Check ESR/CRP, electrolytes every other day for 7 days then Monday/Thursday
- For ventriculitis caused by gram-negative organisms, order MRI with and without contrast prior to discontinuing therapy
- For Complicated CSF Shunt Infection
  - Consider CT scan of head with contrast, bone scan, antibiotic levels in CSF to investigate reasons for persistent sites of infection
  - Additional or prolonged therapy may be necessary, consult outpatient ID
  - Insert new shunt after definitive completion of antibiotic therapy

Patient returns to OR for new shunt
How to Assess a Patient with Hydrocephalus

CT Scan

- MR cannot be used outside of clinic hours because it requires reprogramming of the shunt by clinic staff.
- CT scan is also necessary for OR planning. Given the high rate of shunt revisions from patients presenting to the ED, CT is justified.

[LOE: OOOO (Morton, 2013; Iskandar, 2004)]

Shunt Series

- Although the yield of the shunt series is low, the result of misdiagnosis is unacceptable, so shunt series should be continued.

[LOE: OOOO (Zorc, 2002; Lehnert, 2011; Pitetti, 2007; Desai, 2007)]
Care Following ETV

- Routine PICU admission is not recommended
  *Risk of postoperative complications is extremely low. [LOE: ★★★★★ (Bouras, 2013; Bouras 2011; Baykan 2005; Hader, 2008; Bhatia, 2009; Cinalli, 2007)]*

- Consider PICU admit if patient was previously shunted (exclude patient from pathway)
  *In a cohort of 131 children with ETV previously shunted, intracranial pressure was high following ETV, finding that some patients can develop severe intracranial hypertension. [LOE: ★★★★★ (Hader, 2008)]*

- Check sodium postoperatively and monitor for hyponatremia
  *Hyponatremia is one of the most common complications following ETV, occurring in as may as 16% of patients. [LOE: ★★★★★ (Baykan, 2005; Hader, 2008; Lang, 2012)]*
Empiric Antibiotic Therapy for Suspected Infection

Empiric Antibiotic Therapy

Initial therapy:
- Determine patient medication allergies prior to therapy
- Use vancomycin + ceftriaxone (unless history of past complicated infections with resistant gram-negative organisms or yeast)
  - If there is high suspicion for infection with a resistant gram-negative organism, consider substituting meropenem or cefepime for ceftriaxone as empiric therapy after consultation with the ID service

Alterations of initial therapy based on CSF Gram Stain Results:
- If the CSF gram stain shows gram-negative rods, continue vancomycin, discontinue ceftriaxone and begin meropenem
- If CSF gram stain reveals yeast, begin amphotericin B and consult ID

(ISDA Guidelines, 2004)

Antibiotic Treatment Duration

<table>
<thead>
<tr>
<th>Organism/CSF Findings</th>
<th>Continue antibiotic for x days of negative cultures</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No culture (+) after shunt removed</td>
</tr>
<tr>
<td>CoNS normal CSF</td>
<td>3 days</td>
</tr>
<tr>
<td>CoNS abnormal CSF</td>
<td>7 days</td>
</tr>
<tr>
<td>S. aureus</td>
<td>10 days</td>
</tr>
<tr>
<td>Other gram positive</td>
<td>7 days</td>
</tr>
<tr>
<td>Gram negative rod</td>
<td>10 days</td>
</tr>
<tr>
<td>Yeast</td>
<td></td>
</tr>
<tr>
<td>No growth normal CSF</td>
<td></td>
</tr>
<tr>
<td>No growth abnormal CSF</td>
<td></td>
</tr>
</tbody>
</table>

A culture is considered negative after 48 hours of incubation

(ISDA Guidelines, 2004)
Definitive Antibiotic Therapy

1. Consult Infectious Disease

2. Antibiotic therapy changes should be based on CSF culture and sensitivity. Results are typically available within 48 hours. Definitive antibiotic regimen should include the antimicrobial(s) with the narrowest spectrum and treat the isolated organism.

3. Intraventricular therapy is contraindicated for uncomplicated shunt infections.

4. A positive culture from the ventriculostomy after shunt removal is evidence of ventriculitis, and antibiotic therapy should be re-evaluated.

5. For cases of ventriculitis caused by gram-negative organisms known to carry ampC inducible beta lactamases (Serratia, Indole (+) proteus, Citrobacter, Enterobacter, Morganella), meropenem AND gentamicin or ciprofloxacin should be used. Dual therapy is not necessary for patients without ventriculitis. Alternative therapeutic regimens should be guided by antimicrobial sensitivities.

6. Consider adding rifampin to the antibiotic regimen to treat ventriculitis due to CoNS and S. aureus.

(ISDA Guidelines, 2004)
Complicated CSF Shunt Infections

Most CSF shunt infections clear with appropriate surgical and antibiotic interventions.

However, a small number of cases may be complicated and less likely to respond to usual therapy, and require additional evaluation or prolonged therapy. Indications include:

- Persistently positive cultures after institution of appropriate antibiotic therapy
- Persistently elevated CSF protein levels
- Increasing CSF cell count
- Infection with an organism with a propensity to cause abscess formation
- Evidence of osteomyelitis
- Retained foreign body
- Slow clinical response

(ISDA Guidelines, 2004)
Hydrocephalus Approval & Citation

Approved by the CSW Hydrocephalus for 3/1/2016

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Retrieval Website: http://www.seattlechildrens.org/pdf/hydrocephalus-pathway.pdf

Please cite as:
We used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial, or observational studies. The rating is then adjusted in the following manner:

Quality ratings are *downgraded* if studies:
- Have serious limitations
- Have inconsistent results
- If evidence does not directly address clinical questions
- If estimates are imprecise OR
- If it is felt that there is substantial publication bias

Quality ratings can be *upgraded* if it is felt that:
- The effect size is large
- If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
- If a dose-response gradient is evident

**Quality of Evidence:**
- 4★★★ High quality
- 3★★★ Moderate quality
- 2★★★ Low quality
- 1★★★★ Very low quality
- Expert Opinion (E)

Summary of Version Changes

- **Version 1.0 (3/1/2016):** Go live.
- **Version 2.0 (8/8/2022):** Update algorithm Outpatient Follow-up box for ETV.
Medical Disclaimer

Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children’s Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

Readers should confirm the information contained herein with other sources and are encouraged to consult with their health care provider before making any health care decision.
Studies were identified by searching electronic databases using search strategies developed and executed by a medical librarian, Susan Groshong. Searches were performed in August and September, 2013. The following databases were searched – on the Ovid platform: Medline, Cochrane Database of Systematic Reviews (2005 to date), Cochrane Central Register of Controlled Trials; elsewhere: Embase, CINAHL, Clinical Evidence, National Guideline Clearinghouse, TRIP, Cincinnati Children’s Evidence-Based Care Guidelines, Registered Nurses' Association of Ontario and Nursing+. Retrieval was limited to ages 0 – 18 (0 – 24 in Medline), English language and 2002 to current. In Medline and Embase, appropriate Medical Subject Headings (MeSH) and Emtree headings were used respectively, along with text words, and the search strategy was adapted for other databases using their controlled vocabularies, where available, along with text words. Concepts searched were hydrocephalus, ventricular or cerebrospinal shunts, ventriculostomy and any of the following: diagnostic imaging, laboratory tests, infections, postoperative care. All retrieval was further limited to certain evidence categories, such as relevant publication types, Clinical Queries, index terms for study types and other similar limits. Additional articles were identified by team members and added to results.

Susan Groshong, MLIS
December 2, 2013

Identification

782 records identified through database searching

2 additional records identified through other sources

Screening

764 records after duplicates removed

764 records screened

613 records excluded

Eligibility

151 records assessed for eligibility

110 full-text articles excluded,
101 did not answer clinical question
8 did not meet quality threshold
1 outdated relative to other included study

Included

41 studies included in pathway

Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535
Bibliography


