

Suspected Shunt Malfunction/ETV Failure Clinical Pathway

June 2022

Outcomes/Goals	<ol style="list-style-type: none"> 1. Rapid identification and treatment of children with shunt or endoscopic third ventriculostomy (ETV) failure 2. Team-oriented approach to efficient, timely evaluation and workup 3. Decrease unnecessary radiation exposure
Inclusion Criteria	Patients aged ≤ 19 years with either a ventriculoperitoneal shunt or endoscopic third ventriculostomy (ETV) with symptoms of possible elevated intracranial pressure or shunt infection
Exclusion Criteria	Patients non-functional cerebrospinal fluid diverting shunts, patients with shunts or ETV presenting with symptoms not referable to their neurosurgical hardware
NURSE Documentation	Chief complaint. Onset of symptoms. Vital Signs, including pain assessment. Last relevant surgical revision. Neuro exam, note any deviations from baseline. Activity level, LOC. Seizure history. History of fever, external shunt drainage, shunt or shunt tubing swelling, history of shunt infections, recent trauma. Assess for signs of increased intracranial pressure and meningismus. Document head circumference, and NPO status. If shunt, ask/document programmable vs nonprogrammable shunt (if known).
INTERVENTIONS Initiate on arrival	<p>ESI Triage level II</p> <p>Full set of vitals per standard of care Complete and fax MRI screening form</p> <p>Place on continuous cardiac monitoring and document rhythm Notify LIP if hypertension, bradycardia, or decreased LOC noted Evaluate/consider Zofran for nausea/active vomiting</p> <p>Evaluate need for pain control</p> <p>Patient may go to MRI off monitor if stable (no bradycardia, apnea, LOC changes or sedation)</p> <p>Evaluate need for seizure pads NPO</p> <p>Place topical Lidocaine (LMX) or J-tip order in anticipation of peripheral IV start</p> <p>Elevate HOB 30 degrees</p>
DIAGNOSTICS	<p>Bedside CBG if altered mental status</p> <p>Consider BMP if vomiting, altered mental status, lethargy, behavioral change, etc.</p> <p>If concerns for infection, obtain CBC, CMP, CRP, ESR</p>
PHYSICIAN (LIP)	
Radiology	<p>Plain films: shunt series (NOT REQUIRED FOR ETV) – must be done prior to MRI if shunt is programmable or unknown.</p> <p>Quick brain MRI – within 60 minutes of presentation, first choice if able to obtain, order Head CT without contrast if unstable or MRI delayed</p>
Programmable Shunts	Consult Neurosurgery after MRI on patients with programmable shunts to restore to correct settings.
Medication(s)	<p>Zofran ODT: 0.1-0.2mg/kg/dose</p> <p>Tylenol PO/PR dose: 12.5 mg/kg</p> <p>Fentanyl (2 mcg/kg IN or 1-2 mcg/kg IV / IM)</p> <p>Avoid NSAIDS</p> <p><i>Opioids should be avoided if altered mental status or suspicion for elevated ICP</i></p>
Consults	<p>If patient in extremis, is lethargic, or has signs of Cushing's triad (i.e. bradycardia, hypertension), consult neurosurgery immediately.</p> <p>If patient is stable, consult neurosurgery if shunt failure or ETV failure remains in the differential after imaging</p> <p>Consult pediatric neurology if patient presents with new or worsened seizures</p>
ADMISSION	<p>Notify primary care physician</p> <p>Prepare family/infant for admission to PICU, ward, observation, or OR</p>
SPECIAL CONSIDERATIONS	<p>Admit to PICU if signs of increased ICP and ventriculomegaly</p> <p>Admit to OR for unstable shunt malfunction per neurosurgery</p> <p>Admission to 10N vs observation for possible failure with normal imaging</p>
DISPOSITION	Attending MD may need to see patient in person or via teleconsult prior to discharge.

Suspected Shunt Malfunction/ETV Failure Rationale and Data

Goals of Clinical Pathway

1. Rapid identification and treatment of children with shunt failure.
2. Team-oriented approach to efficient, timely evaluation and workup.
3. Identification of appropriate disposition.

Definitions

Hydrocephalus	Lack of absorption, over-production, or obstruction of flow of CSF. May be present at birth (congenital) or develop later in life (acquired). Occurs in approximately 1 out of every 1000 births. There is a 20-70% chance of developing hydrocephalus following intraventricular hemorrhage.
Communicating hydrocephalus	Ventricles are open but reabsorption of CSF back into the venous system is obstructed. Commonly occurs secondary to hemorrhage or infection. Shunts are often employed to palliate.
Noncommunicating hydrocephalus	Obstruction within ventricular system, typically secondary to congenital malformation or neoplasm. May be palliated with a shunt or in some cases an endoscopic third ventriculostomy.
Shunt Failure	Shunt failure rate is approximately 45-60% the first year following placement. Most common reasons for failure of shunting are infection, obstruction and disconnection. Failure can occur as a result of proximal malfunction (the intracranial catheter can become displaced, the valve can fail), shunt tubing disruption, or distal malfunction (migration of abdominal catheter or formation of CSF cyst at distal end with abdominal swelling, a so-called abdominal pseudocyst).
ETV failure	The overall success rate of ETV six months after the procedure was 66% in one large multicenter study; another large study yielded an 2-year operation-free survival of 58%.

Signs and Symptoms of Elevated ICP

Infant	Bulging fontanel, increasing head circumference, irritability, poor feeding, vomiting, scalp vein distension, setting sun sign (upgaze paresis), episodic bradycardia, apnea, and excessive sleepiness
Child	Headache, vomiting, irritability, change in personality, change in cognition, lethargy, hypersomnolence, discoordination, gait disturbance, seizure, nystagmus, upgaze paresis

Shunt Infection

Fever, irritability, lethargy, erythema at insertion site or tracking along shunt tubing are common findings. Abscesses at the drainage site (abdominal) usually occur in first 1-3 months after placement. Shunt infection is almost always associated with shunt malfunction.

Abdominal Pseudocyst

Abdominal pseudocysts are collections of fluid that develop at the tip of a ventriculoperitoneal shunt catheter within the peritoneum. They may be sterile or infected and typically cause predominantly abdominal symptoms such as distension, pain, nausea, vomiting, or constipation. A minority will also involve neurologic symptoms, though in one study half had increased ventricular size on neuroimaging.

Minimizing Ionizing Radiation

Children with ventricular shunts are likely to receive dozens of CT scans over their lifetimes, significantly adding to the lifetime risk of malignancy. Given this, alternative modalities for evaluating for ventricular shunt failure have been explored. At current the most promising modality is 'quick' or 'rapid' brain MRI. Multiple studies have demonstrated that quick brain MRI has comparable sensitivity and specificity to CT for the evaluation of hydrocephalus in this patient population. At Doernbecher ED, quick brain MRI is the neuroimaging of choice for the stable patient being evaluated for shunt and ETV failure.

References:

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- Erwood, A., Rindler, R. S., Motiwala, M., Ajmera, S., Vaughn, B., Klimo, P., Jr., & Chern, J. J. (2020). Management of sterile abdominal pseudocysts related to ventriculoperitoneal shunts, *Journal of Neurosurgery: Pediatrics PED*, 25(1), 57-61.
- Yue EL, Meckler GD, Fleischman RJ, Selden NR, Bardo DM, Chu O'Connor AK, Vu ET, Fu R, Spiro DM. Test characteristics of quick brain MRI for shunt evaluation in children: an alternative modality to avoid radiation. *J Neurosurg Pediatr*. 2015 Apr;15(4):420-6.
- Kulkarni AV, Drake JM, Mallucci CL, Sgouros S, Roth J, Constantini S; Canadian Pediatric Neurosurgery Study Group. Endoscopic third ventriculostomy in the treatment of childhood hydrocephalus. *J Pediatr*. 2009 Aug;155(2):254-9.e1.

Revision History Table

Document Number Rev. mmddyy	Final Approval by	Brief description of change/revision