Ventriculoatrial Shunt for Complicated Chronic Hydrocephalus

Left-sided Fluoroscopic Guidewire Placement of Ventriculoatrial Shunt

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Background

- Hydrocephalus is a pathologic accumulation of cerebrospinal fluid (CSF) in the brain, with an active distension of the ventricular system resulting from inadequate passage of CSF from its point of production to its point of absorption.[1]
- It has an estimated prevalence of 85/100,000 in the United states, with an estimated 60,000 related hospital discharges reported annually.[1]
- Treatment depends on etiology but is generally via CSF shunting, through ventriculoperitoneal shunting (VPS), or less commonly through ventriculoatrial shunting (VAS) in which the distal catheter is placed in the right atrium. Other techniques include ventriculopleural shunting or endoscopic third ventriculostomy.[2]
- Despite being one of the most common neurosurgical procedures with an incidence of 5.5/100,000 in the US, shunts remain as prone to failure now as they were decades ago, with 40% of pediatric and 29% of adult shunts failing in the first year of placement, and 45-81% of patients with a shunt requiring 1 or more surgical revisions in their lifetime. Shunt revisions are associated with higher failure rates compared to primary shunts, and shunt failures are treated with multiple shunt revisions in >50% of patients.[1,3,4,5]
- Normal venous anatomy favors the right-sided approach, as the path from the right EJV or direct cutdown on right IJV to the superior vena cava and then the right atrium is more direct when compared with approaching from the left EJV/IJV.

Case Presentation

History:
39 year old nonverbal male with past medical history of cerebral palsy (CP), chronic spastic quadriparesis, hydrocephalus shunted at 10 months of age, abdominal surgery for adhesion lysis, multiple VPS revisions, and conversion to ventriculopleural shunt s/p failure of VPS revision 4 months prior presented with 1 day of lethargy, multiple episodes of emesis, and poor PO intake consistent with past instances of shunt failure. Parent denied recent sick contacts, cough, fever, trauma, shortness of breath, or distress.

Last VPS revision in October 2019 demonstrated failure of CSF absorption, and patient was subsequently converted to right ventriculopleural shunt. The ventriculopleural shunt was patent but was complicated by pleural effusion in December 2019.

Physical:
Vitals: T: 35.9 C HR: 98 RR: 22 BP: 137/100 Wt: 32.3 kg
General: No acute distress, alert
HEENT: Pupils equal, round, and reactive to light, with intact extraocular movements and normal conjunctiva. Dry oral mucosa, left parietal shunt refills briskly. Neck is non-tender, and without erythema. Shunt track is palpable.
Respiratory: Respirations are non-labored, with symmetrical chest wall expansion and a right upper chest hernia.
GI: Soft and nontender
Musculoskeletal: No tenderness, extremities contracted x4.
Integumentary: Warm and dry
Neurologic: NAD, nonverbal, awake and alert, PERRL, tracks with eyes, FSM, contractures in all extremities.

Imaging

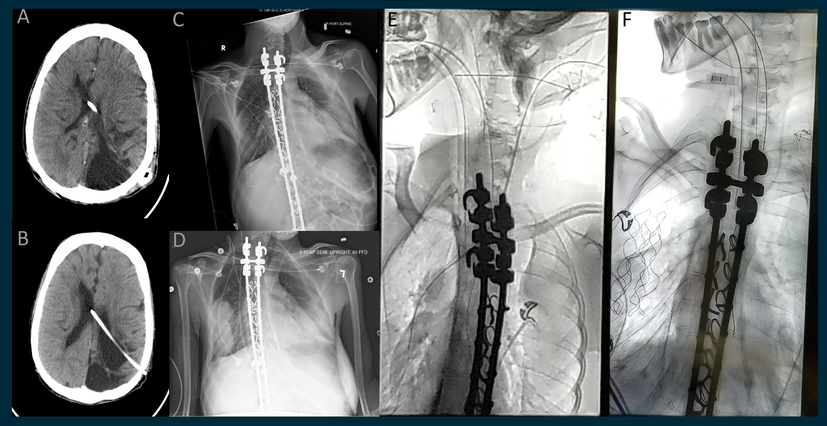

Figure 1. CT images at baseline (A) and after distal shunt failure with ventricular dilatation (B). CXR s/p thoracentesis (C) with reaccumulation of fluid on repeat CXR (D). Fluoroscopic images showing guidewire (E) and distal tip of shunt catheter (F) in right atrium.

Patient Outcome

The patient was admitted to SICU for observation with close neuromonitoring. Over the next few days, the patient remained stable with intermittent emesis. A thoracentesis was performed on the second day of admission to drain accumulated fluid in the right pleural space, but subsequent reaccumulation of fluid suggested poor absorption of CSF in the pleural space. CMP and CBC were remarkable only for low WBC, high glucose, and low creatinine. CSF cultures taken at time of pleurocentesis were negative. Serial CT brain imaging showed slowly progressing ventricular dilation.

With symptoms consistent with shunt failure, and poor absorption of CSF in the pleural space, a revision with ventriculoatrial shunt placement was performed on the third day of admission.

With fluoroscopic guidance, the patient's existing ventriculopleural shunt was externalized and verified to be patent. Vascular access to the left internal jugular vein was obtained and a wire introducer sheath was fed to the right atrium. The catheter length was measured and then a peel-away sheath catheter was introduced and the guidewire retrieved. The ventricular catheter was trimmed to the length of the guidewire at 20 cm, and then passed through the sheath system to the midbody of the atrium, with the peel-away sheath subsequently removed and the incisions closed. The patient tolerated the procedure well and was transferred to the surgical ICU with stable vital signs.

The patient subsequently showed improvement in lethargy with no further emesis. He was discharged in good condition shortly afterward with regular follow up.

Discussion

- Specific abdominal complications of VPS include peritoneal scarring/adhesions, abdominal pseudocysts, intraperitoneal infections, and ascites, in addition to a general unsuitability for further CSF diversion, thus requiring alternative distal catheter placement such as with ventriculopleural shunts or VAS.[5,7]
- Ventriculoplueral shunt associated complications include pleural effusion, pneumothorax, hydrothorax, intrapleural pseudocyts, cardiac tamponade with heart failure, or trapped lung. In rare cases, the distal catheter can migrate and erode into lung parenchyma, resulting in a fistula with subsequent recurrent pneumonia and persistent cough.[6]
- Our patient developed VPS failure after revision with poor peritoneal absorption of CSF, likely due to significant scarring and adhesions secondary to multiple abdominal surgeries and VPS revisions.
- Second line ventriculopleural shunt placement was performed, with generous provision of space in the right pleural space for shunt deposition via thoracoscopy. The shunt revision was initially successful, but after 3 months pleural absorption of CSF became poor with subsequent clinical presentation suggestive of shunt failure.
- Consequently the ventriculopleural shunt was revised to a ventriculoatrial shunt via the left internal jugular vein.
- Ventriculoatrial shunts have their own set of associated complications, such as pulmonary hypertension, arrhythmias, thromboembolic complications, shunt infections, and shunt nephritis, with one single-center study from Austria reporting 12.1% of VAS revisions for shunt infection and 1.6% (a single case) of revision for shunt nephritis.[7]
- Shunt nephritis is a severe VAS specific complication which presents as a secondary renal disease with proteinuria which may progress to end-stage renal disease and possibly death, with overall incidence ranging from 0.7% to 2.3%.[7]
- Most VAS-specific complications are potentially serious but can be reversible with early detection and treatment, and thus a VAS is an adequate alternative to VPS and ventriculopleural shunting with lifelong follow-up to avoid cardiopulmonary or renal complications.[7]
- Our case was complicated by multi-modal shunt failure requiring third-line revision to a ventriculoatrial shunt. The reuse of the left-sided shunt catheter consequently required the left-sided approach and navigation of more difficult anatomy. Technical innovation with the use of the guidewire as reference for catheter length contributed to the success of the VAS placement.

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