Enterocutaneous fistula in the setting of ventriculoperitoneal shunt extrusion through the skin and perforation through the small bowel

Case report

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The authors report a case of a 2-year-old boy with postinfectious hydrocephalus, managed with a ventriculoperitoneal (VP) shunt and complicated by shunt extrusion through the cranial skin. The shunt was removed due to concern for infection, and the child was found to have an enterocutaneous fistula (ECF) communicating along the shunt track between the small bowel and a clavicular sinus. Self-closure of the ECF was anticipated. Thus, the fistula was managed expectantly with dressing changes of the clavicular sinus, while the patient’s malnutrition was managed in accordance with World Health Organization protocols. The presentation, prognosis and management of ECFs, including the likelihood of self-resolution and the role of expectant management, are discussed. Additionally, proposed mechanisms of ECF formation in the setting of a VP shunt are discussed, with an emphasis on the roles of infection and malnutrition.

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KEY WORDS • ventriculoperitoneal shunt • hydrocephalus • enterocutaneous fistula • distal catheter complication

Ventriculoperitoneal (VP) shunting is the standard treatment for hydrocephalus in industrialized countries and in the developing world. Although the perioperative risk of a shunt placement operation is low, the 2-year failure rate is reported to be as high as 50%.6 Abdominal complications occur in as many as 10%–30% of cases of VP shunting, while bowel perforation is much rarer, occurring in 0.1%–1.0% of cases.10 We report a unique case of a pediatric postinfectious hydrocephalus patient with an enterocutaneous fistula (ECF), an abnormal communication between bowel and skin, in the setting of small-bowel perforation by a VP shunt catheter and in association with extrusion of the shunt catheter through the cranial skin. Our case represents only the second reported instance of ECF in the setting of VP shunting. We describe the presentation and prognosis of ECFs in the developed world and in the developing world, and compare our case to a prior report of an ECF in a British adult with a VP shunt. Additionally, we review strategies of ECF management and discuss our approach. Finally, we theorize on the mechanism of ECF development in our case.

Case Report

A 2-year-old boy with a prior medical history of hydrocephalus, seizures, and malnutrition presented to Kijabe Hospital in Kenya with a 2-month history of an exposed right frontal VP shunt. The child initially presented at 1 year of age with a 7-month history of head enlargement, 11-month history of seizures, congenital blindness, developmental delay, malnutrition, and anemia. On examination, the child’s occipitofrontal circumference (OFC) had been 50 cm, and his weight was 9.8 kg. The fontanelle was convex and firm. Ultrasonography of the child’s head demonstrated enlargement of the lateral and third ventricles, in addition to septations. Endoscopic cyst fenestration and right VP shunt placement were performed.

At the time of this second presentation, the mother reported seeing shunt tubing in the right frontal region for the previous 2 months. The child had a 1-week history of subjective fever, without vomiting, diarrhea, irritability, or seizures. He had long-standing difficulty swallowing and poor appetite, resulting in severe malnutrition. On examination, the child was afebrile, with an OFC of 56 cm and weight of 7 kg. The fontanelle was concave and soft. A small area of exposed shunt tubing measuring less than 2 cm was visible on the right frontal scalp. Additionally, the child was found to have an ulceration measuring less than 1 cm in the right clavicular region and associated with minimal drainage. A palpable nonfluctuant mass above the umbilicus was also identified. Cerebrospinal fluid analysis did not demonstrate leukocytosis but did show a decreased glucose level.

The child was admitted to the pediatric neurosurgery service. On the second hospital day, the shunt was removed in its entirety through the right frontal area where it could

Abbreviations used in this paper: ECF = enterocutaneous fistula; OFC = occipitofrontal circumference; VP = ventriculoperitoneal; WHO = World Health Organization.
Enterocutaneous fistula secondary to a VP shunt

be seen. The distal portion of the abdominal catheter was bile stained. Within minutes of shunt removal, bilious discharge was observed from the wound in the right clavicular region. The wound continued to drain bilious material, and on the 4th postoperative day the child underwent a fistulographic examination under fluoroscopy with catheterization of the clavicular sinus. The fistulogram demonstrated an ECF connecting the clavicular sinus to a loop of small bowel along the previous shunt tract (see Figs. 1 and 2). A loop of bowel attached at the epigastrum was also demonstrated, consistent with the palpable mass found above the umbilicus (see Fig. 1). Although the precise segment of the small bowel where the ECF originated was unclear, it was thought to be duodenal given the fistula’s low output. The ECF was managed with dressing changes of the clavicular sinus in anticipation of the fistula’s self-resolution. Additionally, the child met World Health Organization (WHO) criteria for severe wasting. Nutritional support was provided in accordance with WHO guidelines for management of severe malnutrition, with F75 formula, gentamicin, penicillin, chloramphenicol, metronidazole, multivitamin, folic acid, vitamin A, calcium, and zinc. The hospitalization was complicated by pneumonia, *Citrobacter* urinary tract infection, and seizures. The child was discharged home after a 19-day hospitalization with persistent small-volume drainage from the clavicular sinus. He had no signs or symptoms of hydrocephalus at the time of discharge, and no additional interventions for hydrocephalus were undertaken. The fistula closed spontaneously 1 month after discharge. Nine months later, the child had not returned for follow-up but was said to be doing well with no increase in head size being evident to his mother.

**Discussion**

Enterocutaneous fistulas remain a challenging problem today, with mortality rates ranging from 10% to 30%, with death often due to the complications of sepsis, malnutrition, and electrolyte abnormalities. This range suggests a higher mortality than the 16%–18% rate reported for patients with bowel perforation by VP shunt without an ECF. Overall, 75%–85% of ECFs are postoperative (rather than spontaneous), and they are usually associated with an operation for infectious causes in the developing world and with malignancy, inflammatory bowel disease, or adhesiolysis in the developed world. Although our case does not align with these etiologies, the present report of a Kenyan child with postinfectious hydrocephalus is more reflective of the infectious etiology seen in the developing world. A case of ECF in a UK-residing adult with a history of myelomeningocele repair and numerous VP shunt revisions was reported by McGrogan et al. This 39-year-old man presented with months-long history of an abdominal sinus and a suprascapular sinus, without any signs of shunt malfunction or evidence of systemic infection. His original VP shunt was placed shortly after birth, and it required 18 revisions prior to presentation, resulting in the patient having 3 shunt catheters in situ, including 2 VP shunts. The patient was found to have an ECF of small bowel communicating with the abdominal sinus. The patient was treated with dressing changes and nutritional support, and the fistula closed spontaneously 1 month after discharge. Nine months later, the child had not returned for follow-up but was said to be doing well with no increase in head size being evident to his mother.
sinus. A well-defined lumen “in keeping with” the track of the nonfunctional abdominal shunt catheter was demonstrated, with the catheter perforating the small bowel. The similar configuration of ECF in our patient and in this previously reported case is notable in light of the significant differences in patient demographics and medical histories. Additionally, both cases are associated with catheter perforation of the small bowel, which is significantly rarer than colonic perforation by a VP shunt.11

Reports of ECF spontaneous closure rates, which are reflective of a good outcome, range from 19% to 92%, with the variability attributed to differences in patient populations and fistula characteristics.9 In a Nigerian case series of 54 pediatric patients with ECF, the authors reported a 53.7% spontaneous closure rate and a 24.1% mortality rate.14 Predictive factors for spontaneous fistula closure and/or reduced mortality include surgical (rather than spontaneous) etiology, healthy surrounding bowel, no associated abscess cavity, fistula tract greater than 2 cm, enteral defect less than 1 cm and without discontinuity, fistula output less than 500 ml per day, and no comorbidities. Additionally, markers of good nutritional status and the absence of sepsis have been correlated with lower mortality.7,9 In our case, the postsurgical nature of the fistula, the longer fistula tract, and the low output would suggest a better outcome. However, the child’s several comorbidities and severe wasting are consistent with a direr prognosis.

Given that many ECFs close spontaneously, delay in surgical closure of up to 2 months is often warranted, especially as it allows for the correction of metabolic and nutritional deficiencies.9 Spontaneous closure after 4–6 weeks is considered unlikely. In accord with current management strategies for ECF, the initial approach to our patient focused on improvement of nutritional status, electrolyte repletion, control of fistula drainage, as well as local skin care and protection.7 In our case, the case reported by McGrogan et al.10 did not immediately come to neurosurgical attention and was initially managed by general surgeons with daily dressings and the laying open of the sinuses. Once the abdominal ECF in association with the VP shunt track was identified, the patient underwent exploration of the fistula with removal of the offending nonfunctional catheter, curettage of the track, and packing of the wound. Both of this patient’s fistulas were reported to begin to close spontaneously after the procedure.10 In both our case and in the case of McGrogan et al, definitive surgical therapy for ECF involving bowel resection and anastomosis was deferred. Parenteral nutrition, somatostatin, and octreotide were not used in either case. Although these approaches may occasionally be seen in the management of ECF, they are rarely available in developing countries.7,9

We theorize that the creation of the ECF in our case followed from the initial event of shunt catheter extrusion through the cranial skin. Infection and inflammation then promoted the perforation of the bowel. Bile tracking around the shunt catheter led to skin excoriation where bile could ascend from the initial event of shunt extrusion. This promoted the formation of the clavicular sinus. Finally, the removal of the shunt effectively increased the diameter of the fistula, reducing the resistance along the shunt track and resulting in increased flow of bowel contents to the clavicular sinus.

### Table 1: Previous reports of VP shunt extrusion and protrusion through the skin by location of skin defect

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Location of Skin Defect</th>
<th>Age at Extrusion</th>
<th>Age at Placement</th>
<th>Diagnosis</th>
<th>Op Brand/Type</th>
<th>Time to Extrusion From Last Op</th>
<th>Prior Revision</th>
<th>Prior Extrusion, VPS</th>
<th>Indication of Systemic Infection</th>
<th>Tip Extrusion</th>
<th>Location of Systemic Infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aras et al., 2013</td>
<td>abdominal, operated MMC area</td>
<td>21 mos. F</td>
<td>4 mos</td>
<td>HC, MMC</td>
<td>Chhabra slit-n-spring</td>
<td>9 mos</td>
<td>no</td>
<td>NR</td>
<td>no</td>
<td>yes</td>
<td>no</td>
</tr>
<tr>
<td>DeSousa &amp; Worth, 1979</td>
<td>abdominal incision</td>
<td>57 yrs. M</td>
<td>NPH</td>
<td>54 yrs</td>
<td>Holter medium-pressure valve &amp; tubing</td>
<td>3 yrs</td>
<td>no</td>
<td>NR</td>
<td>no</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td>Kankoja et al., 2008</td>
<td>cervical (2 cases)</td>
<td>3–6 mos. NR</td>
<td>5 mos</td>
<td>HC, SB w/lumbosacral defects</td>
<td>Chhabra slit-n-spring System</td>
<td>3–6 mos</td>
<td>yes</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td>Borkar et al., 2008</td>
<td>17 yrs. F</td>
<td>3–6 mos. M</td>
<td>3 mos</td>
<td>obstructive HC, CPA mass defects</td>
<td>Chhabra slit-n-spring System</td>
<td>6 mos</td>
<td>yes</td>
<td>no</td>
<td>yes</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td>Kankoja et al., 2008</td>
<td>15 mos. F</td>
<td>3–6 mos. M</td>
<td>3–6 mos</td>
<td>HC, SB w/lumbosacral defects</td>
<td>Chhabra slit-n-spring System</td>
<td>9 mos</td>
<td>yes</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>yes</td>
</tr>
<tr>
<td>Odebode, 2007</td>
<td>posterior auricular subcutaneous</td>
<td>&lt;1 mos</td>
<td>7 mos</td>
<td>F</td>
<td>Ch, MMC</td>
<td>no</td>
<td>NR</td>
<td>no</td>
<td>NR</td>
<td>NR</td>
<td>no</td>
</tr>
<tr>
<td>Vural et al., 2008</td>
<td>sacrococcygeal</td>
<td>7 mos. F</td>
<td>7 mos</td>
<td>HC, MMC</td>
<td>Holter medium-pressure hydrocephalus</td>
<td>no</td>
<td>NR</td>
<td>no</td>
<td>no</td>
<td>no</td>
<td>yes</td>
</tr>
</tbody>
</table>

* CPA = cerebellopontine angle; HC = hydrocephalus; MMC = myelomeningocele; NR = not reported; SB = spina bifida; VPS = VP shunt.
The initial extrusion of the shunt catheter through the cranial skin may have been a result of tissue fragility due to unrecognized infection with a contribution from malnutrition. Several cases of shunt catheter extrusion through the skin are summarized in Table 1, although there are likely many more cases that have not been reported. Additionally, a report of colonic perforation by a VP shunt catheter mentions a history of repaired shunt protrusion through the abdominal skin having taken place sometime prior to the presentation and with unclear involvement of the catheter tip. Two older reports also describe extrusion through the chest wall and extrusion through a neck incision, with unclear involvement of the catheter tip in both cases. Interestingly, our case of small-bowel perforation and the previously reported case of jejunal perforation both presented with a segment of the catheter eroding through the skin without tip involvement, possibly suggestive of a descending infectious etiology contributing to the perforation. The catheter erosion through the skin was likely facilitated by skin and connective tissue fragility due to malnutrition, exacerbated by the patient's neurological status (i.e., difficulty swallowing) and low resource availability for nutritional restitution.

Several mechanisms of bowel perforation by VP shunt catheters have been proposed. Shunt infection was suggested to lead to bowel perforation via a mechanism similar to the rejection of the catheter as an infected foreign body. Alternatively, irritation from the infected catheter or infected cerebrospinal fluid could lead to a low-grade inflammatory reaction, resulting in fibrous encapsulation. The encasing fibrosis may then have an anchoring effect on the tubing, leading to decubitus ulceration of the bowel wall and resulting in bowel perforation. In our case, bowel weakness due to severe wasting is likely to have further facilitated perforation by the catheter. The formation of the clavicular sinus was then promoted by the migration of bile upward outside the catheter, with possible contribution from the tracking of low-grade infection and inflammation along an area of subcutaneous weakness.

Encasing fibrosis, which has an anchoring effect on the tubing with resultant decubitus ulceration of the bowel wall and eventual bowel perforation, has been described as one of the possible mechanisms of bowel perforation.

**Disclosures**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors.

**References**


