Ventriculoperitoneal shunt with a rare twist: small-bowel ischemia and necrosis secondary to knotting of peritoneal catheter

Case report

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Small-bowel ischemia and necrosis due to knotting of the peritoneal catheter is an extremely rare complication related to a ventriculoperitoneal shunt (VPS). A 3-month-old girl, with a history of Chiari II malformation and myelomeningocele (MM) after undergoing right occipital VPS insertion and MM repair at birth, presented to the emergency department with a high-grade fever. Examination of a CSF sample obtained via shunt tap raised suspicion for the presence of infection. Antibiotic therapy was initiated, and subsequently the VPS was removed and an external ventricular drain was placed. Intraoperatively, as attempts at pulling the distal catheter from the scalp incision were met with resistance, the distal catheter was cut and left in the abdomen while the remainder of the shunt system was successfully removed. While the patient was awaiting definitive shunt revision surgery to replace the VPS, she developed abdominal distension due to small-bowel obstruction. An emergency exploratory laparotomy revealed a knot in the distal catheter looping around and strangulating the distal ileum, causing small-bowel ischemia and necrosis in addition to the obstruction. A small-bowel resection with ileostomy was performed, with subsequent placement of ventriculoatrial shunt for treatment of hydrocephalus. The authors report this exceedingly rare clinical scenario to highlight the fact that any retained distal catheter must be carefully managed with immediate abdominal exploration to remove the distal catheter to avoid bowel necrosis as pulling of a knotted peritoneal catheter may strangulate the bowel and cause ischemia, with significant clinical morbidity and possible mortality.

Key words • ventriculoperitoneal shunt • shunt complication • knot • shunt malfunction • bowel obstruction • bowel necrosis • hydrocephalus • Chiari

Ventriculoperitoneal shunt (VPS) placement is one of the most common procedures performed by neurosurgeons throughout the world. Ever since the initial shunt system was developed in the 1950s, there have been major advances in the technology. Nevertheless, the neurosurgical community is still plagued by significant shunt-related complications such as infection, proximal or distal catheter occlusion, valve malfunction, and pseudocyst formation. A recent study from a large national database showed a 30-day complication rate of 12.9% and a 1-year complication rate of 28.8%,1 with many other historical studies reporting complication rates up to 59% at 1 year.8,12 The most common VPS complications include shunt malfunction and infection. Other unusual complications such as distal catheter migration into the heart, scrotum, or bowel have also been reported.5,7,11,19 Knotting of the distal catheter is very rare but generally present with a shunt block and subsequent malfunction, which often only require simple distal revisions.5,6,9,10,13-16,18,20-22 Small-bowel necrosis due to knotting of the peritoneal catheter is an exceedingly rare complication of a VPS, with only 1 previously described case in the literature.18 We report only the second case of bowel necrosis due to knotting of peritoneal catheter, and we provide a comprehensive review of the literature while considering the clinical relevance of this rare complication.

Case Report

History and Presentation. A 3-month-old girl with a history of Chiari II malformation and myelomeningocele...
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(MM) after undergoing right occipital VPS placement and MM repair at birth presented to the emergency department with a high-grade fever. Examination of a CSF sample obtained via shunt tap was positive for Gram-negative rods, raising suspicion for shunt infection. Computed tomography of the head showed stable ventricle size; a shunt series was ordered but was not completed in the emergency department.

Management. The patient was started on broad-spectrum antibiotics while awaiting culture results. She was taken to the operating room immediately for VPS removal and external ventricular drain placement. Intraoperatively, attempts at pulling out the peritoneal catheter were met with great resistance. At the time, it was thought that the peritoneal catheter was trapped due to intraabdominal adhesions; thus, the peritoneal catheter was cut with the distal part retained in the peritoneal cavity. The rest of the shunt system was removed, and the external ventricular drain was placed. Postoperative abdominal radiography showed knotting of the retained peritoneal catheter in the right lower quadrant (Fig. 1). Because the final CSF cultures were negative and the patient was doing well initially after surgery, the retained abdominal catheter was left in place; the plan was to remove it when a new VPS was being inserted after the child had medically stabilized. However, the patient developed clinical and radiographic signs of small-bowel obstruction over the next few days (Fig. 2). She was taken to the operating room for exploratory laparotomy, which revealed knotting of the peritoneal catheter around distal ileum that was causing bowel ischemia and necrosis. Resection of the necrotic bowel and ileostomy were performed.

Postoperative Course. The patient was in stable condition, with improved clinical status after surgery. Postoperative abdominal radiography showed complete resolution of bowel obstruction and distention (Fig. 3). A ventriculoatrial shunt was subsequently placed and the patient was eventually discharged to home in stable condition.

Discussion

There are about 36,000 shunt-related procedures performed in the United States each year.1 Spontaneous formation of a knot of the distal VPS catheter in the abdomen is an unusual complication; only 12 cases have been reported in the literature to date.4,6,9,10,13–16,18,20–22 Bowel ischemia and necrosis due to strangulation of intestines by the knotted peritoneal catheter can occur, and this complication is exceedingly rare, with only 1 case previously reported in the literature.18 The case reported here represents only the second occurrence of bowel necrosis caused by a knotted distal shunt catheter. Previously reported cases and associated complications are summarized in Table 1.

The extremely low incidence of such a complication makes it a difficult phenomenon to apprehend, and the exact mechanism of spontaneous knotting of the peritoneal catheter remains a matter of speculation. Some authors have hypothesized that longer catheter length, smaller diameter, greater elasticity, and a crowded intraabdominal...
space are potential risk factors for spontaneous formation of a knot in the peritoneal catheter.\(^5,6,5\) Others have suggested that vigorous intestinal peristalsis could also contribute to spontaneous knotting.\(^2,5\) Raymer and Smith performed an interesting physical experiment by placing a piece of string in a rotating cubic box, and they found that the probability of spontaneous knot formation increased with longer string length, increased flexibility, increased total number of rotations, slower rotational speed, and larger cube volume.\(^7\) Although this physical experiment does not totally simulate the physiological milieu of shunt catheter in the peritoneal cavity, it does provide some insight into the plausible mechanism of spontaneous knot formation. Perhaps a more physiologically comparable phenomenon is spontaneous knotting of the umbilical cord during pregnancy, which has a reported incidence of 1.22%.\(^3\) Bakas et al. reviewed several large retrospective studies and concluded that long umbilical cords, male embryos, and multiparity (larger uterine volume) were correlated with umbilical cord knot formation.\(^2\) The physical experiment and physiological phenomenon described above both seem to suggest that excessively long catheter length in the abdomen probably plays an important role in knot formation. Interestingly, 11 of the 13 known cases (including the present case) of peritoneal catheter knotting occurred in the pediatric patients, in whom a longer relative length of catheter in the peritoneal cavity might be present due to their small physical size. It is also interesting to note that 1 of the 2 adult patients was a 25-year-old pregnant woman, highlighting the fact that a crowded peritoneal space and increased peristalsis may also contribute to knot formation.\(^2,2\) Though not proven, consideration may be given to shortening the length of the distal catheter in very young infants to reduce this complication.

A majority (8 of 13) of patients with peritoneal catheter knots presented with shunt malfunction along with one case in which the knot was an incidental finding (Table 1). The remaining 4 cases had bowel obstruction as part of the symptoms, and bowel necrosis only occurred in 2 patients (including the present case). Both cases of bowel ischemia and necrosis occurred after initial unsuccessful attempts to remove the peritoneal catheters, and retrieval of the retained catheters was done in a delayed fashion. It is likely that the excessive length of distal catheter was looping around a segment of bowel, and pulling of distal catheter during attempted removal actually tightened the loop, resulting in knot formation and strangulation of the bowel. Delayed recognition of the problem and lack of prompt removal of the retained catheter eventually led to bowel necrosis. Therefore, it is prudent to carefully study preoperative shunt series to check for any existing loop or knot formed by the peritoneal catheter before shunt revision procedures. If resistance is met during distal catheter removal, one should refrain from using excessive pulling force, since pulling may tighten an existing knot/loop and strangulate the bowel. Prompt general surgery consultation should be obtained in such circumstances to remove the distal catheter to prevent more serious complications such as bowel necrosis. In the setting of a suspected infection, prompt removal of retained catheter is even more important for eradicating a potential nidus and facilitating the clearance of infection.

### Conclusions

Bowel ischemia and necrosis secondary to peritoneal catheter knotting is a rare but serious potential complication that all neurosurgeons should bear in mind. Any retained distal catheter must be carefully investigated and an intraoperative general surgery consult should be strongly considered. Excessive pulling of a looped/knotted peritoneal catheter may strangulate the bowel. The value of a shunt series should not be underestimated as it can provide a clue to the presence of such a complic-
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tion. Immediate general surgery consultation should be obtained when difficult distal catheter extraction is encountered.

Disclosure

Dr. Moftakhar is a consultant for Covidien and Penumbra.

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References


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