Ischemic bowel secondary to ventriculoperitoneal shunt knotting: a surprisingly rare complication of ventriculoperitoneal shunting. Case report

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Small-bowel obstruction (SBO) and volvulus as a result of ventriculoperitoneal shunting are a rare phenomenon, especially when resulting in bowel necrosis. The authors report the rare event of SBO, bowel strangulation, and necrosis in a pediatric patient after the abdominal catheter became knotted around his small bowel, and they provide a comprehensive review of the literature. The authors argue that shunt configuration is an important consideration for a patient presenting with SBO, and that symptomatic shunt knotting is a reason for surgical correction.

<https://thejns.org/doi/abs/10.3171/2019.11.PEDS19138>

KEYWORDS small-bowel obstruction; SBO; ischemic bowel; VPS; knot; ventriculoperitoneal shunt complication; hydrocephalus

VENTRICULOPERITONEAL shunts (VPSs) are the most common form of CSF diversion system in use, and thousands are placed in the United States each year. Common complications include infection, malfunction, and over- and underdrainage, as well as abdominal complications including peritonitis, adhesions, herniation, migration, bowel perforation, pseudocyst formation, and small-bowel obstruction (SBO) or volvulus. SBO and volvulus as a complication of VPS remain an extremely rare phenomenon, with only 23 cases reported in the literature; moreover, SBO and volvulus associated with bowel gangrene and necrosis as a complication of VPS remain exceedingly rare, with only 8 cases reported. We report the case of a 5-year-old boy presenting in extremis who was found to have SBO with necrotic bowel secondary to an abdominal VPS knot, and we have also performed a systematic review of the literature. We demonstrate the presence of a knot on shunt radiographs obtained over several years, which should be noted even in an asymptomatic patient. VPS configuration should be well noted on shunt radiographs, and the presence of a suspicious knot or coil should lower the threshold for operative intervention if the patient is clinically symptomatic.

Methods

The case was reviewed by the IRB committee and a release form for imaging and photography was signed by the underage patient's healthcare proxy. Imaging was obtained and annotated from the internal PACS. A photograph of the intraoperative findings was obtained. The literature review was completed using PubMed.gov and Google Scholar including the following search terms: "abdominal complications of ventriculoperitoneal shunt," "small bowel obstruction from ventriculoperitoneal shunt," and "volvulus from ventriculoperitoneal shunt." Any article related to SBO or volvulus as a result of VPS was included as well as reported knot/coil formation of any kind.

Case Report

History

We present the case of a 5-year-old boy who presented with a 1-day history of worsening lethargy and emesis. He had a past medical history significant for grade 4 intraventricular hemorrhage in infancy with a VPS status post multiple revisions (the last of which was placed via minilaparotomy at another institution), as well as a history

ABBREVIATIONS SBO = small-bowel obstruction; VPS = ventriculoperitoneal shunt.

SUBMITTED March 11, 2019. **ACCEPTED** November 18, 2019.

INCLUDE WHEN CITING Published online January 31, 2020; DOI: 10.3171/2019.11.PEDS19138.

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FIG. 1. Abdominal radiograph obtained on presentation showing dilated loops of small bowel and concern for SBO. The VPS knot was visible in the patient's right midabdomen (red arrow).

FIG. 2. Intraoperative photograph of small bowel strangulated by the VPS catheter knot.

of Nissen fundoplication, pyloroplasty, G-tube placement, and vagal nerve stimulator placement for medically refractory epilepsy. The patient had presented 1 year previously with a bowel obstruction that had been successfully treated with conservative management.

Examination

On arrival to the emergency department the patient was found to have agonal respirations and received emergency intubation. He underwent an emergency CT head and shunt series to evaluate for shunt failure, but the shunt was found to be functioning appropriately with no increase in ventricular size. On examination the patient was noted to have a markedly distended but compressible abdomen, and the shunt series abdominal film was significant for severely dilated bowel and concern for SBO (Fig. 1). On laboratory examination the patient was found to have an acute kidney injury and a profound leukocytosis of 53,700/ μ l with a procalcitonin of 8.23 ng/ml. A CT abdomen-pelvis sequence was performed, which showed a large amount of free fluid with severely dilated bowel with focal areas of decompression and no obvious transition point.

Operation

The patient was taken on an emergency basis to the operating room for an exploratory laparotomy. On inspection of the bowel it was noted that several loops of small bowel had been tied into a bouquet configuration by the VPS

catheter, which was found to be tightly knotted (Fig. 2). Approximately 60 cm of small bowel was noted to be necrotic and was ultimately resected. The patient's VPS was externalized, and he was left in discontinuity with plans to return to the operating room for closure the following day. At his second operation, another 15 cm of terminal ileum was found to be necrotic and nonviable and he underwent an ileocectomy with an ileocolonic anastomosis.

Postoperative Course

The patient was continued on broad-spectrum antibiotics in the interim and his shunt was reinternalized in the abdomen (since there was no active infection or history of nonabsorption) on postoperative day 12. He had an otherwise uncomplicated hospital course and was discharged home on hospital day 16.

Discussion

Thousands of VPSs are placed every year in the United States,²⁰ but bowel obstruction secondary to strangulation by the shunt catheter remains a rare complication. Shunt complications can include infection, proximal malfunction, disconnected tubing, distal clogging, shortening, and over- and underdrainage. Abdominal complications can include postoperative peritonitis, adhesions, herniation, migration, bowel perforation, pseudocyst formation, and bowel obstruction or volvulus.⁵ SBO from shunted hydrocephalus remains a rare phenomenon, with 23 cases

reported in the literature.^{1–3,5,6,9–12,14,17,18,21–27,29,31} SBO or volvulus secondary to VPS that involves bowel strangulation or necrosis is an even more rare phenomenon, with only 8 cases reported in the literature.^{12,15,22,25,26,29,31} Knot or coil formation in the distal portion of the VPS has been reported^{4–10,12–19,22–28,30} mainly as an iatrogenic event after an attempt at externalizing the abdominal shunt tubing.^{12,22,25,26} In 1 case, after meeting resistance while removing the distal catheter and finding it to be knotted in the abdomen, Sanan et al. were able to relieve the knot by using intraoperative fluoroscopy and an angiographic guidewire.²² Proposed mechanisms for spontaneous knot formation include crowding of the intraabdominal contents (such as in pregnancy), small diameter and high elasticity of the tubing, intense bowel peristalsis, or excessive intraperitoneal catheter length,^{6,7,15,16} although some authors believe that spontaneous intraabdominal knot formation is not possible.¹⁰ Table 1 includes a comprehensive list of all reported bowel obstruction/volvulus and knot/coil formation cases and their relationship to one another as well as whether this was a reportedly iatrogenic event.

In our case we believe there are several possible explanations for how this event may have happened. Proposed mechanisms (described above) for knot formation include iatrogenic events such as pulling the distal portion and leaving this in place when resistance is met. We believe that this is unlikely to be the case because the shunt in this patient is in complete continuity based on the imaging, with no retained catheter fragments indicative of this complication. Other proposed risk factors include crowding of the intraabdominal contents (such as in pregnancy), small diameter and high elasticity of the tubing, intense bowel peristalsis, or excessive intraperitoneal catheter length. The patient did have several prior abdominal surgeries including multiple shunt revisions, Nissen fundoplication, pyloroplasty, and G-tube placement. Prior surgeries can lead to abdominal adhesions, which effectively compartmentalize the abdomen and leads to worse crowding of the intraabdominal contents. The last revision was when the patient was 2 years old, and because it is common practice to place adequate catheter length for the patient to grow, excessive length may have been left during that time. It is possible that a combination of excessive catheter length, crowding of the abdominal contents, and peristalsis over several years could have led to the catheter becoming knotted. It could have also been slightly coiled when placed originally, and that could have been exacerbated over several years of growth and peristalsis.

The best way of preventing the shunt knot/coil formation would be to take anticipatory steps toward avoiding the aforementioned risk factors such as direct visualization of placement of the abdominal catheter or by routinely obtaining a postoperative shunt series. For example, in a patient with multiple prior abdominal surgeries, it would be reasonable to perform shunt revision in conjunction with the general surgery team and to place the distal catheter via direct laparoscopic observation. Given our patient's history, it may have been reasonable for the last revision to have been done in this way rather than via minilaparotomy. Other ways of preventing a knot in the acute setting of shunt revision would be to take active

steps in troubleshooting the operation if resistance is met. For example, if resistance is met when trying to remove the catheter it would be reasonable to call an immediate intraoperative general surgery consultation for assistance or to take a radiograph and observe the configuration. If the radiograph looks concerning for knotting, then it may be reasonable to convert the abdominal operation to laparoscopic or open surgery, or to try unknotting the catheter under fluoroscopic guidance as described by Sanan et al. This is done by taking a stiff angiographic guidewire and slowly introducing it into the lumen in an attempt to uncoil the catheter and allow it to be removed by gentle traction.²²

On review of the imaging from the patient's previous hospital visits it was apparent that the knot in his peritoneal catheter had been there for some time, even 3 years prior on a shunt series obtained after breakthrough seizures (Figs. 3 and 4). To the best of our knowledge, our case report is the only one documented with serial imaging concerning for impending bowel strangulation. This makes the argument that continuity and location of the shunt catheter is not the only important consideration in a shunt series, but also the configuration. In general, we believe that imaging should be obtained if a patient demonstrates signs of shunt failure or bowel obstruction or perioperatively. We do not advocate for routine serial imaging; we would obtain serial imaging only if the patient is symptomatic and following shunt revision, to provide a new baseline and ensure that the shunt is contiguous, in the correct cavity, and has a normal configuration. Bowel obstruction with a concerning shunt configuration should lower the threshold for operative intervention, even in the absence of an acute abdomen. It would have been reasonable to electively correct the knotted distal catheter during the first episode of SBO 1 year previously if it had been recognized at that time. We believe a knot or coil with evidence of SBO or volvulus warrants shunt exploration and revision because it may prevent a potentially life-threatening event in the future. An algorithm to manage a discovered knot/coil would be to assess the patient's symptoms: If asymptomatic, observation should be used. If there are signs of shunt failure, the cranial component should be explored with testing of proximal and distal flow. If there is proximal failure, the proximal catheter should be replaced. If there is distal failure, or if the patient presents with symptoms of bowel obstruction, then exploration of the abdominal component should be performed with replacement or revision of the distal catheter.

Conclusions

Fortunately, bowel obstruction and subsequent ischemia/necrosis secondary to VPS catheter strangulation remain an extremely rare phenomenon. We advocate that bowel obstruction with an abdominal VPS catheter should prompt assessment of the configuration of the catheter, particularly looking for the presence of any knotting or coiling on imaging. The presence of a knot or coil in the setting of bowel obstruction is an indication for surgical correction because it could potentially prevent a future intraabdominal catastrophe.

TABLE 1. Comprehensive literature review of SBO/volvulus and VPS knot/coil formation

Authors & Year	Pt Age, Sex	Pathology	Presentation	Volvulus/ SBO	Necrosis	Knot/ Coil	lat	Treatment
Ameah & Nmadu, 2000	8 mos, NR	NR	Vomiting/abdominal distension	Yes	NR	No	No	Reduction of volvulus/repositioning tube
Arora et al., 2014	Infant, F	Meningomyelocele/hydro	Abdominal distension/vomiting	Yes	No	No	No	Lysis of adhesions
Bal et al., 1999	11 mos, M	Hydro/meningomyelocele	SBO	Yes	No	No	No	De-rotation/perforation closure
Borek et al., 2012	3 yrs, M	Posttraumatic hydro	Hydro/shunt failure	No	No	Yes	No	Replacement of system
Burhan et al., 2018	9 mos, F	Congenital hydro/meningomyelocele	NR/abdominal pseudocyst	Yes	NR	No	No	VPS externalized/ileostomy
Chopra et al., 2004	4 yrs, F	Posterior fossa tumor	NR	NR	NR	Yes	NR	VPS revision
Eftekhari & Hunn, 2008	25 yrs, F	Hydro/bithalamic glioma	Hydro/shunt failure	No	No	Yes	NR	Distal portion replaced
Espósito et al., 2003	3 yrs, M	IVH/hydro	Shunt failure/abdominal cyst	No	No	Yes	No	Collection evacuated/untangled knot
Espósito et al., 1998	NR	NR	SBO	Yes	No	Yes	No	Unrolled and replaced
Fekete et al., 2013	NR	NR	SBO	Yes	No	Yes	No	Catheter removed laparoscopically
	4 mos, F	Premature/hydro/IVH	Vomiting/tense fontanelle/shunt failure	No	No	Yes	Yes	Cut knot, left catheter
Grosfeld et al., 1974	10 mos, M	Postmeningitic hydro	SBO	Yes	No	No	No	Catheter removed
Hlavín et al., 1990	3 yrs, M	Lumbar myelomeningocele	Infection/externalization attempt/ SBO	Yes	Yes	Yes	Yes	Externalization/re-anastomosis/VA shunt
Kiat et al., 2018	32 yrs, F	Neonatal meningitis/hydro/recurrent SBO	SBO	Yes	No	Yes	No	Shunt dissected out & shortened
Lo et al., 2013	10 yrs, F	Cerebral palsy	Hydro/shunt failure	No	No	Yes	No	Shunt system replaced
Martínez Fernández et al., 2016	24 yrs, F	Cerebellar medulloblastoma	SBO	Yes	Yes	Yes	No	Anastomosis
Mohammed et al., 2011	14 yrs, M	X-linked congenital hydro	Shunt failure	No	No	Yes	NR	Replacement of system
Mohindra & Sharma, 2012	10 yrs, M	Crouzon's disease/congenital hydro	Asymptomatic/chest boil	No	No	Yes	No	Antibiotics
Murtagh et al., 1980	16 mos, F	Extraventricular obstructive hydro	SBO	Yes	No	Yes	No	Takedown of adhesions
Naim-Ur-Rahman et al., 1996	3 yrs, M	Hydro	Abdominal distension/pain	Yes	No	Yes	Yes	Repositioning into abdomen
Sakoda et al., 1971	5 wks, M	Lumbar myelomeningocele/hydro	SBO/volvulus	Yes	No	No	No	Volvulus reduced/catheter removed
Sanan et al., 1995	1 yr, F	Subdural peritoneal shunt	SBO	Yes	Yes	Yes	Yes	Anastomosis
	7 mos, M	Myelomeningocele/hydro/CSF collection	Shunt failure	No	No	Yes	Yes	Knot untied w/ angiowire
Shakir et al., 2018	16 yrs, M	Cerebral palsy/hydro/ileostomy	Decreased ileostomy output	Yes	No	Yes	No	Lysis of adhesions/volvulus reduced
Sims & Chamberlain, 1976	14 wks, M	Prematurity/communicating hydro	SBO/infection	Yes	No	Yes	No	Lysis of adhesions/externalization/VA shunt
Starreveld et al., 1998	8 wks, F	Dandy-Walker	Infection/SBO	Yes	Yes	Yes	Yes	Anastomosis/antibiotics/ventricular taps/VA shunt
Tan et al., 2014	3 mos, F	Chiari II/myelomeningocele	Infection/SBO	Yes	Yes	Yes	Yes	Ileostomy/VA shunt
Toshifumi & Tatsu, 2001	63 yrs, M	Posttraumatic hydro	SBO	Yes	No	Yes	No	Unknotting of catheter
Ul-Haq et al., 2013	8 yrs, M	Congenital hydro	Infection/hydro/shunt failure	No	No	Yes	No	Replacement of system
Wilson & Bertan, 1966	8 mos, M	Communicating hydro	SBO	Yes	Yes	No	No	Ileostomy/colostomy

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TABLE 1. Comprehensive literature review of SBO/volvulus and VPS knot/coil formation

Authors & Year	Pt Age, Sex	Pathology	Presentation	Volvulus/ SBO	Necrosis	Knot/ Coil	lat	Treatment
Woerdeman & Hanlo, 2006	1 wk, M	LS meningomyelocele/Chiari II	Shunt failure	No	No	Yes	No	Tubing untangled
Zhao et al., 2015	4 mos, M	Posterior fossa medulloblastoma	Vomiting/abdominal distension	Yes	Yes	No	No	Laparotomy
	6 yrs, M	3rd ventricular teratoma	Vomiting/abdominal pain	Yes	Yes	No	No	Conservative management, then laparotomy

Hydro = hydrocephalus; lat = iatrogenic; IVH = intraventricular hemorrhage; LS = lumbosacral; NR = not recorded; Pt = patient; VA = ventriculoatrial.

FIG. 3. 3D reconstructions of CT scans from the patient's sentinel bowel obstruction 1 year prior to current presentation (2017; left) and from the patient's most recent admission (2018; right). The knot in the catheter is highlighted with arrows.

FIG. 4. Shunt series and abdominal radiographs spanning 3 years demonstrating a persistent knot in the VPS catheter. Knot is highlighted with arrows. A: Shunt series (8/5/15). B: Shunt series (3/21/17). C: Abdominal radiograph evaluating for obstruction (8/24/17). D: Abdominal radiograph obtained during patient's current admission (8/24/17).

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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

Conception and design: all authors. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Szweczyk. Administrative/technical/material support: Adamo, Whyte. Study supervision: Adamo, Whyte.

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