Symptomatic vitelline duct anomalies in children

EMMAUEL A. AMEH
PHILIP M. MSHELBWALA
MOHAMMED M. DAUDA
LIADI SABIU
PAUL T. NMADU

Division of Paediatric Surgery, Department of Surgery, Ahmadu Bello University Teaching Hospital, Zaria, Nigeria

Summary
A wide variety of anomalies may occur as a result of the vitelline duct (VD) failing to obliterate completely. Most reports on symptomatic VD focus on Meckel’s diverticulum, while other anomalies are given little attention. A retrospective review was conducted at our institution. According to the records 18 symptomatic children with VD anomalies were seen over 22 years, including 10 boys and 8 girls aged 11 days - 14 years (median 7.5 months). Twelve patients aged below 10 years (median 28 days) had patent vitelline ducts (PVDs), 3 children aged 13 months, 13 years and 14 years respectively had Meckel’s diverticulum (MD), presenting as inflammation, tapeworm incarceration and volvulus respectively. Two patients, both 8 years old, had umbilical sinus, and a 3-year-old had a vitelline cyst. Only 1 patient with PVD had an associated anomaly (intestinal malrotation). The diagnosis of PVD was obvious clinically, but in 1 patient the fistula was demonstrated by fistulogram. The diagnosis of MD was intraoperative in all 3 patients. Treatment was by various types of resection for PVD and MD and excision for umbilical sinuses and cysts. One patient with PVD developed postoperative intestinal obstruction from adhesions, requiring re-laparotomy and adhesiolysis. Two patients with PVD died from sepsis and anaesthetic-related complications, respectively. Although MD is the most commonly VD anomaly, PVD is the most common symptomatic presentation in our environment.

Results
The modes of presentation and treatments were as follows:

Patent vitelline duct (PVD)
Twelve children, age 11 days – 9 years, presented with PVD. Seven were neonates, 3 were aged 3 months and below, and the remaining 2 were aged 2 years and 9 years respectively. The main presenting feature was intermittent or persistent discharge of bowel content from an opening in the umbilicus. The discharge usually started soon after the umbilical stump fell off during the neonatal period. In 1 neonate there was prolapse of gangrenous intestinal mucosa through the opening. In 8 patients the fistula was in the ileum, in 1 case it was in the appendix, and in 2 patients it was in the ascending colon. A fistulogram was done in 1 patient with a fistula in the ascending colon, confirming the fistula site before surgery. The site of the fistula could not be ascertained in 1 neonate who died before surgery. Only 1 patient with PVD had an associated anomaly (intestinal malrotation). The diagnosis of PVD was obvious clinically, but in 1 patient the fistula was demonstrated by fistulogram. The diagnosis of MD was intraoperative in all 3 patients. Treatment was by various types of resection for PVD and MD and excision for umbilical sinuses and cysts. One patient with PVD developed postoperative intestinal obstruction from adhesions, requiring re-laparotomy and adhesiolysis. Two patients with PVD died from sepsis and anaesthetic-related complications, respectively. Although MD is the most commonly VD anomaly, PVD is the most common symptomatic presentation in our environment.

Meckel’s diverticulum (MD)
Three children presented with complicated MD. One, a 13-month-old boy, had acute abdominal pain and tenderness, vomiting and passage of bloody stools; intussusception was suspected but an inflamed MD was found at surgery. A 13-year-old girl with acute right lower quadrant pain, suspected to have appendicitis, was found at surgery to have an inflamed MD with a tapeworm trapped in it. The third child, a 14-year-old girl, presented with intestinal obstruction caused by a volvulus of a gangrenous loop of ileum around a fibrous band attached to the abdominal wall, and a MD. Treatment in the 3 patients was by wedge resection of the MD, and resection of the gangrenous loop of ileum in the third patient.
Umbilical sinus
Two children, both aged 8 years, had recurrent discharge of mucoid fluid from the umbilicus dating back to the neonatal period. There was no connection to the intestine. Treatment was by excision of the sinus via an infraumbilical incision in 1 and minilaparotomy in the other.

Umbilical cyst
A 3-year-old boy presented with a cystic left flank mass (15 cm × 10 cm), which was found at surgery to be attached to the inner aspect of the umbilicus by a fibrous band. The cyst and fibrous band were excised at laparotomy.

Morbidity and mortality
One child treated for PVD developed adhesive intestinal obstruction necessitating laparotomy and adhesiolysis. There were 2 deaths – a 26-day-old baby with PVD died from sepsis (the focus was omphalitis before the umbilical stump fell off) before surgery, and a 2-month-old baby with PVD died without recovering from anaesthesia.

Histopathological examination
No ectopic gastric or pancreatic tissue was seen at histological examination of the excised VD remnant in any patient.

Discussion
During development of the gastrointestinal tract a wide range of anomalies may occur in relation to the obliteration of the vitellointestinal duct. Knowledge of the embryology of the vitellointestinal duct is necessary for a good understanding of the various anomalies and their management. The embryology of these anomalies has been well described.

Presentation
MD is alleged to be the most common pathology among symptomatic children with VD anomalies. Several complications are known to cause symptoms, including intestinal obstruction from intussusception or volvulus, bleeding and inflammation, and very rarely foreign bodies and parasites may be trapped within the diverticulum. Most complications of MD occur in patients under the age of 10 years, but bleeding usually occurs in those younger than 5 years. There are only a few reports of other modes of presentation of VD anomalies other than MD. In one report of 217 cases of VD anomalies, 84 (40%) were asymptomatic, mostly MD, with an average age of 2.4 years; 4 neonates had PVD. In the current report, PVD was the mode of presentation in 10 of the 16 patients (62.5%), most of them neonates or young infants. The connection in PVD is usually to the ileum, but less commonly to the appendix or colon as in this report. Other modes of presentation such as umbilical sinus and cysts were less common in this report. It may well be that symptomatic Meckel’s diverticulum is missed in our environment because many children with abdominal pain are undiagnosed; they are frequently treated empirically for malaria and parasitic infestations, which are more common problems in our environment.

Evaluation
Although the clinical picture may be obvious in patients with PVD, a fistulogram may be necessary to identify the part of the intestine involved preoperatively. Bleeding MD may require radionuclide scanning for localisation. A sinogram will exclude intestinal communication in umbilical sinus and abdominal ultrasonography should localise a cyst.

Treatment
When connected to the ileum, a PVD may have a wide lumen or be a high-output fistula. This may lead to fluid and electrolyte deficits, especially in neonates, and treatment should be expeditious. The treatment of a PVD is wedge or segmental resection. The treatment for MD is similar as well as the resection of any associated gangrenous bowel due to intestinal obstruction. When the diagnosis of MD is made preoperatively, laparoscopic excision has been reported. The excision of umbilical sinus and cyst should include any fibrous attachments to the intestine to avoid intestinal obstruction in the future. The role of surgery in asymptomatic patients (incidental findings) with MD remains controversial. However, as most symptomatic patients are young children, elective resection of the VD remnant should be performed if found coincidentally.

In this review 2 patients with PVD died; however, morbidity and mortality can be avoided by prompt treatment. In conclusion, although MD is the commonest VD anomaly reported, PVD is the most common symptomatic presentation in our environment. The lack of information in patients with incidental finding of MD makes it difficult to appreciate the overall incidence of these anomalies in our environment.

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REFERENCES