CHAPTER 64
VITELLINE DUCT ANOMALIES

Bankole S. Rouma
Kokila Lakhoo

Introduction

Vitelline duct or omphalomesenteric duct anomalies are secondary to the persistence of the embryonic vitelline duct, which normally obliterates by weeks 5–9 of intrauterine life. These anomalies occur in approximately 2% of the population and may remain silent throughout life, or may present incidentally sometimes with an intraabdominal complication. Although Meckel's diverticulum is the most common vitelline duct anomaly (Figure 64.1(G)), a patent vitelline duct (Figure 64.1(A)) is the most common symptomatic presentation in developing countries.[1]

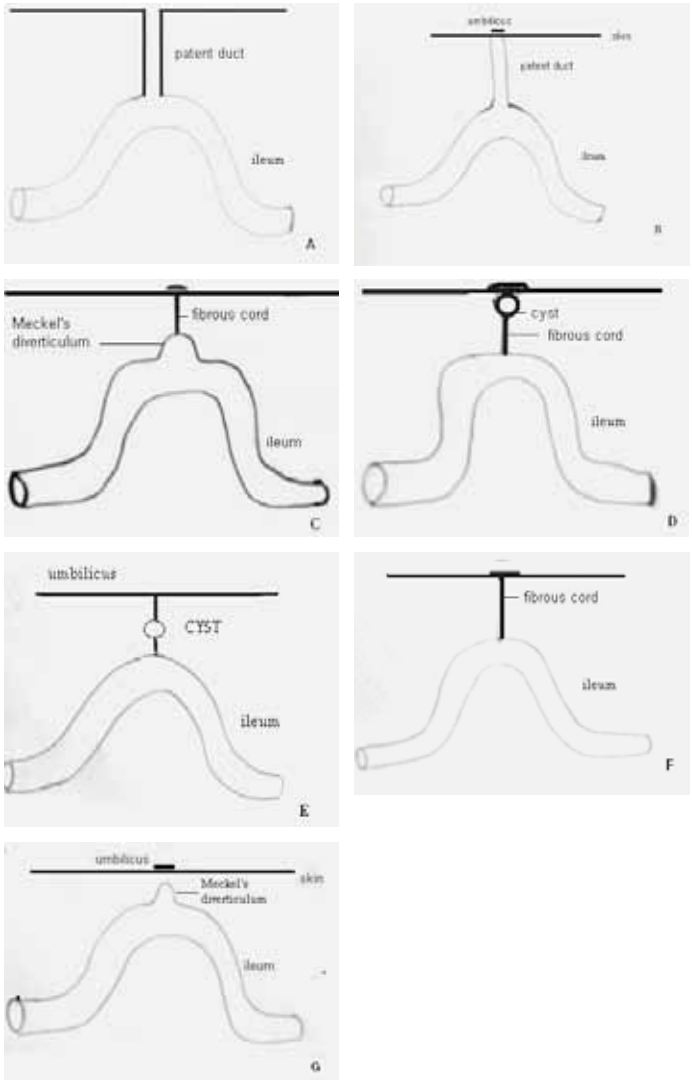


Figure 64.1: Remnants of the omphalomesenteric duct: (A) patent vitelline duct; (B) patent vitelline duct covered by skin; (C) Meckel's diverticulum with fibrous cord; (D) cyst with fibrous cord; (E) cyst; (F) fibrous cord; (G) Meckel's diverticulum.

Demographics

The most frequent malformation is Meckel's diverticulum, with an incidence of 2–3% of the population, but it is one of the most unlikely to cause symptoms. About 4% of children with a Meckel's diverticulum develop symptoms, and more than 60% of those who develop symptoms are younger than 2 years of age.[2–5] The male-to-female complication rate ratio is about 3:1.[3]

Embryology

During week 3 of gestation, the midgut is open into the yolk sac, which does not grow as rapidly as the rest of the embryo. Subsequently, by week 5, the connection with the yolk sac becomes narrowed and is then termed a yolk stalk, vitelline duct, or omphalomesenteric duct. Normally, the vitelline duct disappears by gestational week 9, just before the midgut returns to the abdomen. Persistence of some portion of the vitelline duct results in a number of congenital anomalies, of which Meckel's diverticulum is the most common. This anomaly is variable in length and location, but most often it is observed as a 1–5 cm intestinal diverticulum projecting from the antimesenteric wall of the ileum within 100 cm of the caecum. It possesses all three layers of the intestinal wall and has its own blood supply. The connection in a patent vitelline duct is usually to the ileum, but less commonly may be to the appendix or colon.[1] In other cases, part of the vitelline duct within the abdominal wall persists, forming an open omphalomesenteric fistula, an enterocyst, or a fibrous band connecting the small bowel to the umbilicus.[2–7]

Pathophysiology

Vitelline duct malformations comprise a wide spectrum of anatomic structures, depending on the degree of involution of the vitelline duct. The most common anomaly is Meckel's diverticulum, described as being 60 cm from the ileocaecal valve, 2 cm in diameter, 3 cm in length, and not attached to the abdominal wall. Most complications of these abnormalities are related to ectopic tissue (gastric, pancreatic, colonic, endometriosis, or hepatobiliary).[7]

Ectopic gastric tissue usually causes bleeding from ulceration of the adjacent ileal mucosa. The ileal mucosa is not equipped to buffer the acid produced by the ectopic gastric mucosa and thus is prone to ulceration. The site of the ulceration is most often at the junction of the normal ileal mucosa and the ectopic gastric mucosa. Some studies have shown a very low colonisation rate with *Helicobacter pylori* in children with ulcerative bleeding of Meckel's diverticulum.[3]

Intestinal obstruction may be caused by a Meckel's diverticulum attached to the umbilicus by a fibrous cord or by a fibrous cord between the ileum and the umbilicus. This may lead to a volvulus around the fibrous cord. A persistent vitelline artery, an end artery from the superior mesenteric artery, may cause obstruction and volvulus. Bowel obstruction can also occur by intussusception with the diverticulum as a lead point or by herniation or prolapse of the bowel through a patent omphalomesenteric fistula (with a characteristic "ram's horn" appearance).[5] Obstruction may be caused by phytobezoar.[6,7]

Like the appendix, a Meckel's diverticulum can become inflamed when the lumen is obstructed, resulting in decreased mucosal perfusion, tissue acidosis, and bacterial invasion of the wall. This can lead to progressive inflammation, with tissue gangrene and perforation. It is possible that the gastric or pancreatic mucosa contributes to the luminal obstruction, or the gastric mucosa can lead to ileal mucosal ulceration first, which facilitates bacterial invasion. Rarely, foreign bodies and parasites may be trapped within the diverticulum and cause obstruction of the diverticulum as does an enterolith.[7,8] Diverticular torsion leads to secondary ischaemia and inflammatory change.[7]

Anomalies of the omphalomesenteric duct can result in umbilical drainage from granulation tissue. Other anomalies include a duct extending to the umbilicus but covered with skin (Figure 64.1(B)); diverticulum attached to the umbilicus with a fibrous cord (Figure 64.1(C)), Littre's hernia, and intraabdominal cystic mass (Figure 64.1(D,E)).

Some tumours can be found in ectopic tissues, such as nesidioblastosis in ectopic pancreas tissue of a Meckel's diverticulum or tumours such as carcinoid, leiomyoma, neurofibroma, and angioma.[6–9]

Associated congenital anomalies include cardiac defects, congenital diaphragmatic hernia, duodenal atresia, esophageal atresia, imperforate anus, gastroschisis, malrotation, omphalocele, Hirschsprung's disease, and Down syndrome.

Clinical Presentation

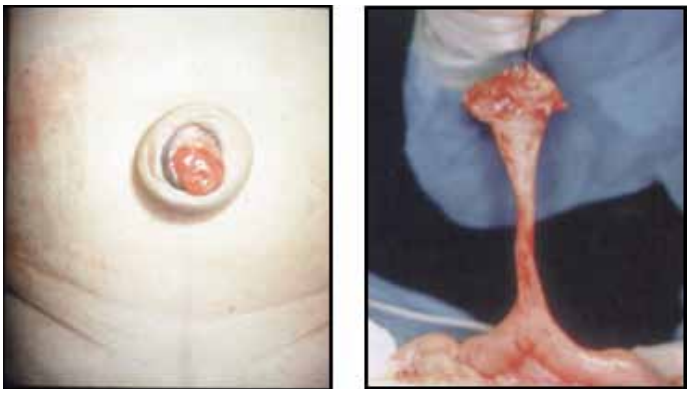
The clinical presentation of vitelline duct abnormalities is variable and depends on the configuration of the remnant of the vitelline duct and whether it contains ectopic gastric or pancreatic tissues. In developed countries, the main forms of presentation are haemorrhage in 40–60%, obstruction in 25%, diverticulitis in 10–20%, and umbilical drainage.[3–5]

The classic presentation is an older infant or young child with painless rectal bleeding. This usually consists of a large volume of bright red bleeding but can occasionally also present as dark, tarry stools in small amounts. The bleeding is often massive and frequently requires transfusion. Melena may be episodic and usually ceases without treatment; sometimes the melena is insidious and not appreciated by the family. In a young child with haemoglobin positive stools and a chronic iron deficiency anaemia, the diagnosis of Meckel's diverticulum should be considered.

Intestinal obstruction, usually due to intussusception, is the most typical presentation in newborns and infants. The symptoms include crampy abdominal pain, bilious vomiting, currant-jelly stools, and abdominal distention. Intestinal obstruction may also be caused by a volvulus or arterial band. Because the volvulus usually involves the distal small bowel and the obstruction is most often a closed loop, there may be little emesis until late in the course. The sequelae of intestinal ischaemia, such as acidosis, peritonitis, and shock, may occur first, and can be fatal in infants.

Patients with Meckel's diverticulitis often have symptoms that resemble appendicitis. They are usually older children. Periumbilical pain is the first symptom. They usually do not have the same amount or intensity of vomiting and nausea as do children with appendicitis. On physical examination, their point of maximal tenderness may migrate across the abdomen as the child moves. About the same percentage of patients with diverticulitis will present with perforation. A perforated Meckel's diverticulum is potentially more serious than a perforated appendix because the former is more difficult to wall off due to its more mobile position. This may explain why perforated diverticulitis is more likely to result in diffuse peritonitis and pneumoperitoneum detectable on abdominal radiographs. For this reason, it is imperative to search carefully for a perforated Meckel's diverticulum as the cause of peritonitis when no inflamed appendix is discovered at appendectomy.

Other types of symptomatic omphalomesenteric duct malformations can result in umbilical drainage as well. The quantity and character of the drainage may indicate the origin of the lesion. Clear drainage or yellowish drainage signifies a probable urachal anomaly, whereas



(A) (B)

Figure 64.2: (A) Patent vitelline duct; (B) vitello-intestinal communication.

an omphalomesenteric duct remnant manifests as faeculent drainage (Figure 64.2). The most common umbilical lesion is an umbilical granuloma, which secretes a mucoid material. If the drainage persists despite cauterization of the presumed granuloma with silver nitrate, or if the drainage is copious, imaging studies are indicated. Prolapse of the ileum into the duct at the anterior abdominal wall presents as a discoloured, mucosa-covered mass situated at the umbilicus.

Diagnosis

Diagnosis of a symptomatic vitelline duct malformation is dependent on the anatomic configuration and its presentation, signs, and symptoms. History and physical examination are important for the diagnosis. Some abnormalities are evident on physical examination (faecal fistula, prolapse of ileum through a patent duct, and umbilical granulation tissue with a small fistula). A fistulogram may be necessary to identify the part of the intestine involved preoperatively.

A complete description of the quality and frequency of the bloody stools is necessary in patients with rectal bleeding. Rectal examination and lower endoscopy is useful to identify other causes of lower bleeding (polyps and rectal tears). The test of choice for a bleeding Meckel's diverticulum is a technetium-99m pertechnetate isotope scan (Meckel scan), which preferentially concentrates the isotope in ectopic gastric mucosa. The specificity of scintigraphy is 95%, but the sensitivity is 85%.[7] A negative scan result does not, however, exclude a bleeding Meckel's diverticulum. Capsule endoscopy has proven to be of diagnostic value in some cases of bleeding Meckel's diverticulum, but the reports are very few. These tests are rarely available in developing countries. The best diagnostic test may be a laparotomy to visually look for a Meckel's diverticulum in children with unexplained rectal bleeding.

If obstruction from either intussusception or volvulus is suspected, plain x-rays may reveal dilated bowel loops and multiple air-fluid levels. An air enema or upper gastrointestinal study with small bowel follow-through is suggestive. Ultrasonography remains fairly reliable to diagnose intussusception.

A sinogram will exclude intestinal communication in umbilical sinuses, and abdominal ultrasonography should localise a cyst.[1] Inflammatory symptoms are similar to those of appendicitis and are diagnosed clinically.

Treatment

Symptomatic children with omphalomesenteric duct remnants should be resuscitated before intervention. Those with significant haemorrhage should be transfused. Patients with obstructive symptoms should be resuscitated as rapidly as possible to obviate the need for ischaemic bowel resection. The incision chosen varies with the symptoms and the age of the patient. Children with faeculent umbilical drainage (see Figure 64.2) or prolapse of the omphalomesenteric duct remnant can be explored by a small infraumbilical incision.

Children with Meckel's diverticulitis or a bleeding Meckel's diverticulum are operated on by using a transverse appendectomy incision with medial extension if necessary. Patients with suspected intestinal obstruction should be explored through a generous laparotomy incision.

An open diverticulectomy includes the following steps:

1. A transverse appendectomy incision or subumbilical incision is made.
2. The caecum and ileum are identified.
3. The ileum is followed proximally to find Meckel's diverticulum, approximately 60 cm from the ileocaecal valve.
4. The diverticulum with the ileum are delivered into the wound.
5. The diverticulum is excised with the adjacent ileum and primary ileal end-to-end anastomosis is fashioned.

In developed countries, some surgeons use linear staplers applied to the base of the anomaly, allowing complete amputation of the diverticulum without narrowing the lumen of the ileum. When ectopic gastric or pancreatic tissues are present near the base of the diverticulum, or if the base is wide, inflamed, or perforated, resection of the involved ileum is required with an end-to-end anastomosis.[2–5,10] If perforation has occurred, thorough peritoneal toileting is done after segmental ileal resection. The use of laparoscopy for resection of Meckel's diverticula has been reported by many authors.[11]

Controversy exists about what should be done when a Meckel's diverticulum is encountered during a laparotomy for unrelated symptoms. The debate focuses on the probability of the Meckel's diverticulum becoming symptomatic in the future weighed against the possibility of complications associated with resection.[2,4,5,7,10–13] Lesions with palpable ectopic mucosa (the consistency of gastric or pancreatic tissue differs sharply from that of ileal, jejunal, or colonic mucosal lining), a prominent vitelline artery, a fibrous vitelline artery remnant, evidence of inflammation, or a narrow base may all increase the chance of bleeding, obstruction, or diverticulitis and should be resected when encountered. In patients who have abdominal pain, it is prudent to resect a discovered diverticulum or any lesion with attachments to the umbilicus (to prevent ileal volvulus). Some authors suggest that resection of asymptomatic vitelline remnants in early childhood is reasonable at the time of laparotomy for other conditions.[10–13] In developing countries incidental Meckel's diverticulum should be removed in children to prevent later complications. If the diverticulum is left in place, it is imperative to alert the patient's family and the primary care physician about the presence of the lesion and its possible symptoms.

Postoperative Complications

Postoperative complications are generally the same as that of other operations: bleeding, infection, intraabdominal abscess formation, wound dehiscence, incisional hernia, and postoperative adhesive intestinal obstruction.

Evidence-Based Research

The study presented in Table 64.1 is a systematic review that addresses the management of incidentally detected Meckel's diverticulum.

Table 64.1: Evidence-based research.

Title	Incidentally detected Meckel diverticulum: to resect or not to resect?
Authors	Zani A, Eaton S, Rees CM, Pierro A
Institution	Department of Paediatric Surgery, Institute of Child Health, London, England
Reference	Ann Surg 2008; 247(2):276–281
Problem	The management of incidentally detected Meckel's diverticulum (MD) remains controversial.
Intervention	The aims of this paper were to establish the prevalence of MD, and the morbidity and mortality due to MD.
Comparison/ control (quality of evidence)	The prevalence of MD is 1.2%, and historical mortality of MD was 0.01%. The current mortality from MD is 0.001%. The number of MD resections per year per 100,000 population decreased significantly after the paediatric age range ($P < 0.001$). Resection of incidentally detected MD has a significantly higher postoperative complication rate than leaving it in situ ($P < 0.0001$). The long-term outcome of patients with incidentally detected MD left in situ showed no complications. To prevent one death from MD, 758 patients would require incidentally detected MD resection.
Outcome/ effect	The prevalence of MD is 1.2%, and historical mortality of MD was 0.01%. The current mortality from MD is 0.001%. The number of MD resections per year per 100,000 population decreased significantly after the paediatric age range ($P < 0.001$). Resection of incidentally detected MD has a significantly higher postoperative complication rate than leaving it in situ ($P < 0.0001$). The long-term outcome of patients with incidentally detected MD left in situ showed no complications. To prevent one death from MD, 758 patients would require incidentally detected MD resection.
Historical significance/ comments	MD is present in 1.2% of the population, it is a very rare cause of mortality, and it is primarily a disease of the young. Leaving an incidentally detected MD in situ reduces the risk of postoperative complications without increasing late complications. A large number of MD resections would need to be performed to prevent one death from MD. The above evidence does not support the resection of incidentally detected MD, in developed countries.

Key Summary Points

1. A patent vitelline duct with umbilical faecal drainage is the most symptomatic presentation of vitelline duct anomalies in developing countries.
2. In developed countries, the main forms of presentation are haemorrhage in 40–60%, obstruction in 25%, diverticulitis in 10–20%, and umbilical drainage.
3. The most common umbilical lesion is an umbilical granuloma, which secretes a mucoid material.
4. If the umbilical drainage persists despite cauterization of the presumed granuloma with silver nitrate, or if the drainage is copious, imaging studies are indicated.
5. In African children, an incidental Meckel's diverticulum must be resected because of the difficulties to rapidly access paediatric surgical heath facilities in case of complications.
6. Resection of asymptomatic vitelline remnants in early childhood at the time of laparotomy or laparoscopy for other conditions is indicated.
7. When ectopic gastric or pancreatic tissues are present near the base of the diverticulum, or if this base is wide, inflamed, or perforated, resection of the involved ileum is required with an end-to-end anastomosis.
8. If the indication of diverticulectomy is bleeding, then segmental ileal resection should be performed.

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