42 Omphalomesenteric Duct Remnants

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42.1 Introduction

The omphalomesenteric (or vitellointestinal) duct is an embryonic communication between the primitive yolk sac and the developing midgut. During normal development at the sixth week of embryogenesis, the midgut loop elongates and herniates into the umbilical cord. Within the 'physiological umbilical hernia', the midgut rotates 90° counterclockwise around the axis of the superior mesenteric artery. At the same time, as the midgut elongates, the lumen of the omphalomesenteric duct begins a process of obliteration. By the tenth week of early foetal development, the midgut returns to the abdominal cavity and the omphalomesenteric duct becomes a thin fibrous band, which undergoes resorption. Persistence of the duct leads to a spectrum of anomalies that can present clinically in the newborn period, infancy, or later childhood years.

42.2 Variant Pathology of Omphalomesenteric Duct Remnants

1. Meckel's diverticulum (Fig. 42.1). The ileal segment remnant of the duct remains patent and usually contains heterotopic gastric mucosal tissue. The diverticulum may be connected to the umbilicus by a fibrous band, if the obliterated duct fails to be fully resorbed.
2. Umbilical fistula (Fig. 42.2). Completely patent omphalomesenteric duct connects the ileal segment of the small intestine to the anterior abdominal wall.
3. Omphalomesenteric cyst (Fig. 42.3) develops when a segment in the midportion of the duct remains patent whilst each corresponding end portion of the tract obliterates.
4. A persistent fibrous cord (Fig. 42.4) connects the umbilicus to the small intestine where the duct here obliterates but is not fully resorbed.
5. Umbilical polyp (Fig. 42.5). A bright red nodule of sequestered ectopic gastrointestinal tissue may reside in the umbilical dimple.

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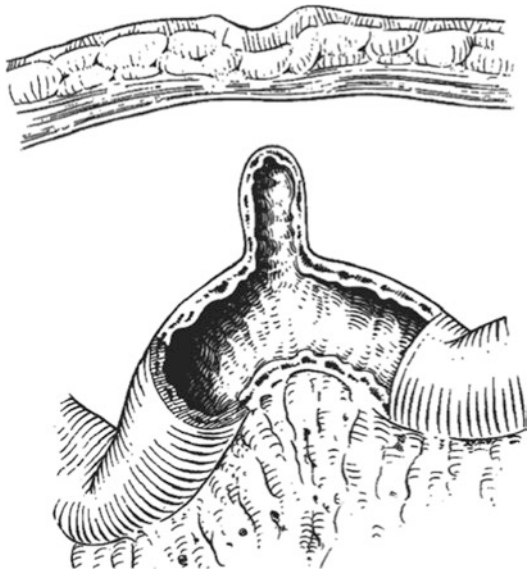

Fig. 42.1 Meckel's diverticulum

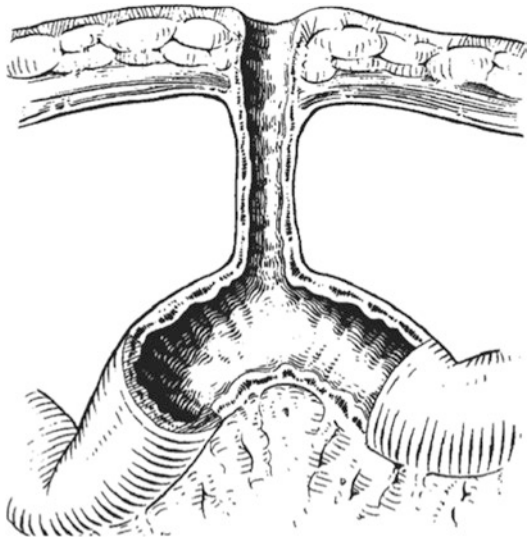

Fig. 42.2 Umbilical fistula

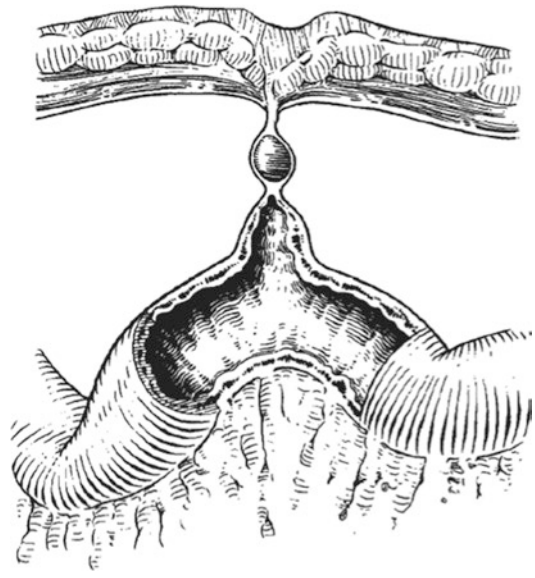

Fig. 42.3 Omphalomesenteric cyst

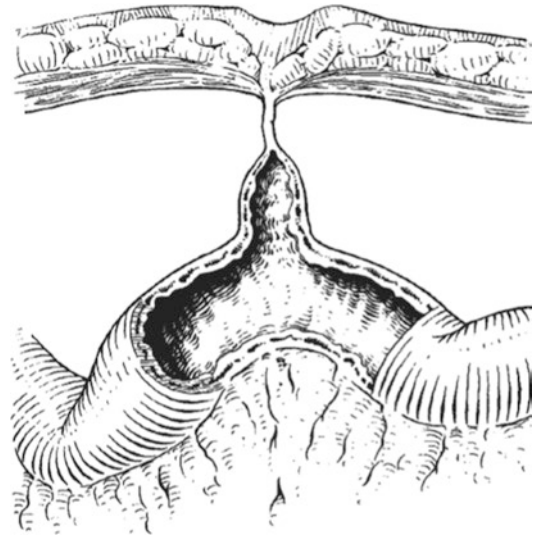

Fig. 42.4 Persistent fibrous cord

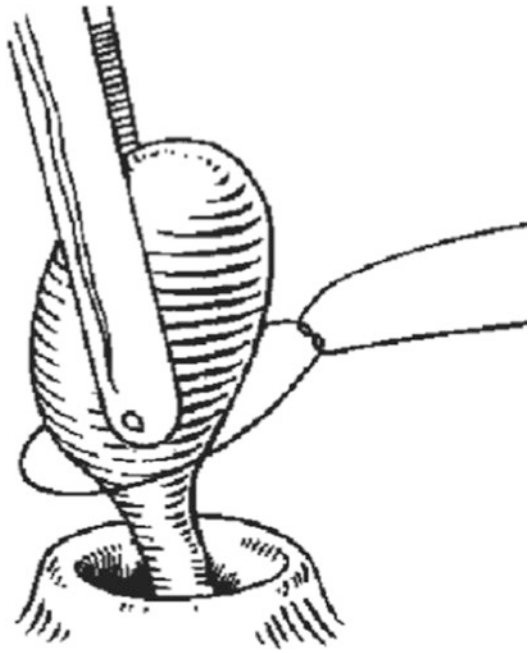

Fig. 42.5 Umbilical polyp

42.3 Meckel's Diverticulum

Originally described in 1809 by the German anatomist, Johann Friedrich Meckel (1781–1833), it is a true diverticulum composed of all three layers of the intestinal tract. Frequently, it contains heterotopic gastric, pancreatic and less commonly duodenal, colonic, or biliary mucosa.

Meckel's diverticulum is the most common of the omphalomesenteric duct anomalies encountered in clinical practice with a 0.3–2.9% prevalence (Zani et al. 2008; Hansen and Soreide 2018). It results from a patency of the intestinal segment of the duct, with or without a fibrous obliteration of the distal tract at the umbilicus. According to a recent systematic review by Hansen et al. (Hansen and Soreide 2018), the lesion is often located some 52 cm from the IC valve (range 7 cm–200 cm) on the antimesenteric border of the ileum. The diverticulum is nourished by a rich blood supply from the vitellointestinal vessels that lie within a fold of the gut mesentery.

42.4 Clinical Presentation

Regarding prevalence, Meckel's diverticulum is rare and often clinically silent. Symptomatic Meckel's diverticulum accounts for only 4–16% of all index cases (Zani et al. 2008; Park et al. 2005). It was estimated in two large published series that the lifetime probability of a Meckel's diverticulum becoming symptomatic is 4.2–6.4%. More than 75% of symptomatic MD occur in children younger than 10 years of age with a median age of some 3.5 years old (Keese et al. 2019). A lifetime risk of developing complication(s) from a Meckel's diverticulum is 6.4% (possibly higher in males vs females). The clinical presentation usually results from complications arising from the presence of the diverticulum, which include the following:

Lower gastrointestinal haemorrhage is the most common presentation in 25–56% of symptomatic cases (Keese et al. 2019; Menezes et al. 2008; Lohsiriwat et al. 2014) secondary to active bleeding resultant from peptic ulceration due to ectopic gastric mucosa. The incidence of gastric mucosa is estimated at 16–24% in asymptomatic case(s) and 24–71% in symptomatic Meckel's diverticulum associated with haemorrhage. Ectopic pancreatic tissue is found in some 12% of Meckel's diverticulum specimens (Hansen and Soreide 2018). The ulcer may be located in the diverticulum or adjacent ileum. Bleeding is usually profuse and painless, manifesting as bright red bloody stools with clots and physiological instability hypovolaemic shock. Rarely, bleeding may be occult in nature for weeks–months with patients featuring a chronic anaemia.

Abdominal pain may occur secondary to intestinal obstruction and diverticulitis.

Intestinal obstruction is the second commonest complication accounting for 14–46% of symptomatic cases (Hansen and Soreide 2018; Keese et al. 2019; Menezes et al. 2008). It is more often encountered in older children. Symptoms include bile- stained vomiting, abdominal distension and colicky pain. Findings at operation may

reveal intussusception, volvulus, or an internal visceral hernia from persistent internal attachment of the diverticulum to the umbilicus.

Diverticulitis is more commonly encountered in the adult surgical population compared to children (29% vs 20%) (Hansen and Soreide 2018), which may manifest as abdominal pain, fever and vomiting. Clinical presentation may be indistinguishable from that of acute appendicitis.

42.5 Investigations and Diagnosis

Imaging studies to aid definitive diagnosis should be tailored according to the varied clinical presentation.

Haemorrhage is the major complication of Meckel's diverticulum in paediatric population. Technetium-99 m pertechnetate scintigraphy (Fig. 42.6) is commonly employed to aid diagnosis. The gastric mucosa accumulates and secretes the pertechnetate isotope. After intravenous injection, a focal area of increased isotope activity is often apparent in the right lower abdominal quadrant within some 30 minutes. However, visualization may take up to1 hour if there are smaller amounts of heterotopic tissue. The sensitivity of Tc-99 m pertechnetate scintigraphy is estimated at approximately 85% with a specificity of 95%. These figures may decline with increasing age as seen in the poorer yield of positive studies in the adult surgical population. False-positive studies may occur when there is a gastric or small intestinal cystic or tubular duplication(s). False-negative reports are also seen with Meckel's diverticula that do not contain adequate amounts of heterotopic gastric mucosa to sufficiently concentrate the Tc-99 m isotope. A recent study from Canada (Vali et al. 2015) found 30% positivity on the second 'repeated' Tc-99 m scan after a first negative study. The study authors encourage better preparation of patients before repeating the isotope study to enhance the scan result. However, minimally invasive surgery is an option here to deploy or utilize an umbilical scar crease incision to access the abdomen in clinically suspicious cases with a negative scan.

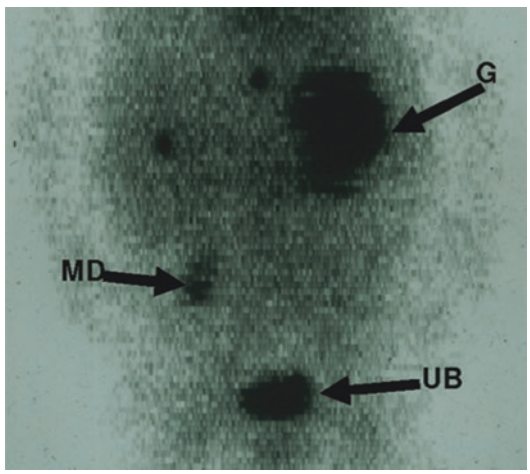

Fig. 42.6 Tc pertechnetate isotope scan showing increaseds uptake in a Meckel''s diverticulum (M) bearing gastric mucosa. Note the normal uptake in gastric mucosa (G) and excretion through the urinary bladder (UB)

Intestinal obstruction is readily diagnosed from plain film radiography. Ultrasonography may be of value in the further evaluation of children with a suspected Meckel's diverticulum and a negative Tc-99 m pertechnetate scintigraphy scan. Diverticula appear as round or tubular 'cyst-like' structures. Echogenic foci in the lumen of the diverticulum may represent enteroliths or inflammatory debris. Colour Doppler may also demonstrate anomalous vessels. CT scanning has been used in some centres. A recent report by Almadi et al. (Almadi and Aljohani 2020) interestingly showed complicated Meckel's diverticulum presenting with intestinal obstruction with Schistosoma parasites harbouring within the diverticulum.

42.6 Differential Diagnosis

Differential diagnosis includes appendicitis, bleeding peptic ulcer disease, inflammatory bowel disorders, or pelvic inflammatory disease, especially in teenage girls.

42.7 Management

The definitive treatment for symptomatic Meckel's diverticulum (Fig. 42.7) is open or laparoscopic surgical resection. This can be achieved either by diverticulectomy (Fig. 42.8a and b) or by a segmental-limited small bowel resection and then intestinal anastomosis (Fig. 42.9).

There has been ongoing debate about the merits of excision of Meckel's diverticulum when it is incidentally found at operation. A review from the Mayo Clinic, USA, in 2005, noted four features commonly associated with a symptomatic Meckel's (1) age < 50 years, (2) male sex, (3) a diverticulum exceeding more than 2 cm in length and (4) the presence of heterotopic tissue.

A subsequent review in 2018 by Slivova et al. (2018) and then Sinopidis et al. (2019) also found that the width of the Meckel's diverticulum base is thought to be a significant predictor for the presence of heterotopic mucosal tissue(s) and here recommend removing all incidental Meckel's diverticulum that also have any of the four features listed above (Park et al. 2005). It must be acknowledged, however, that during any operation, it is not always possible to clearly determine by inspection or palpation whether an incidentally found Meckel's diverticulum is at an increased risk of complications or not.

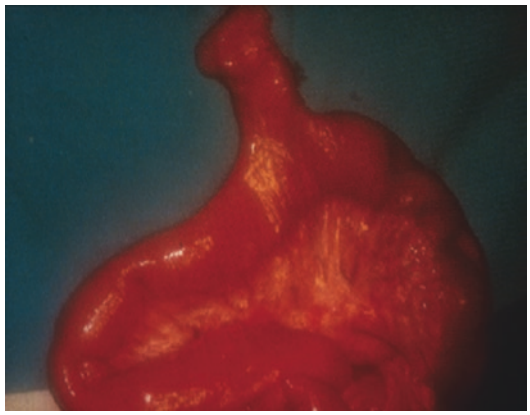

Fig. 42.7 Operative appearance of a Meckel's diverticulum

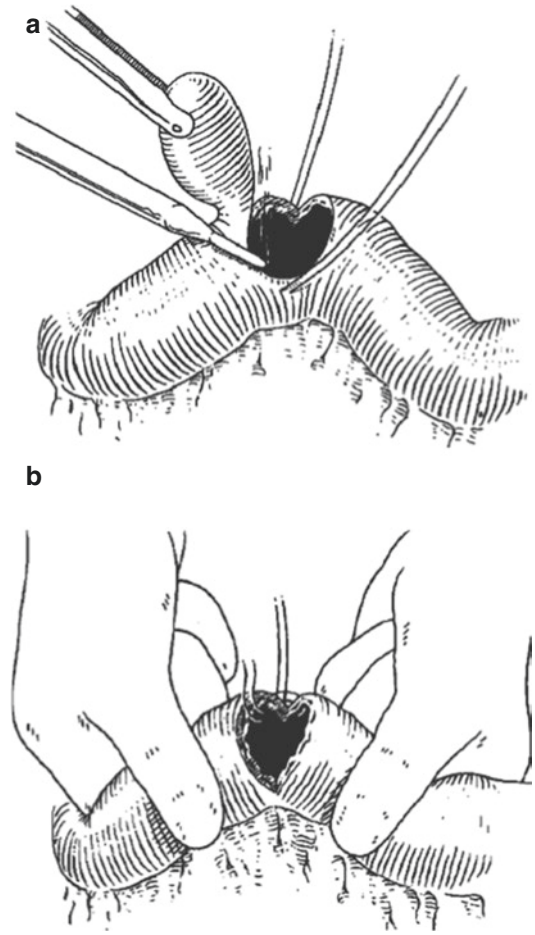

Fig. 42.8 Meckel's Diverticulectomy (**a** and **b**)

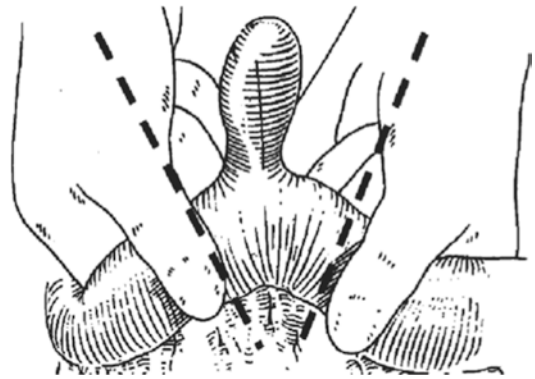

Fig. 42.9 Resection of a Meckel's diverticulum

A number of studies have advocated the resection of 'all' incidental Meckel's diverticulum based on the conclusion that incidental Meckel's diverticulectomy is not associated with added morbidity and mortality (Arnold and Pellicane 1997; Bani-Hani and Shatnawi 2004). Morbidity and mortality after operation for complicated diverticulitis was seen in up to 12% of cases, which is some 10% higher compared to incidental diverticulectomy (Cullen et al. 1994). A systematic review study in 2008 did not find compelling evidence to support routine excision of an asymptomatic Meckel's diverticulum as incidental 'unplanned' diverticulectomy was noted to have higher postoperative complications. Moreover, it was estimated by the study authors that some 758 patients would require incidentally detected Meckel's diverticulum to be resected to prevent a single fatality (Zani et al. 2008).

Of interest, several neoplasms have been reported arising in Meckel's diverticulum, which have clearly not received much attention in the paediatric surgical literature. These lesions include notably carcinoid tumours, GIST and signet ring cell intestinal adenocarcinomas. Amongst the tumours arising from Meckel's diverticulum, GIST tumours are the most common lesions, accounting for some 41% of cases with gut adenocarcinoma(s) the least common (Kabir et al. 2019). Sakpal et al. (2009) reported Krukenberg tumour metastases arising from a Meckel's diverticulum signet ring cell adenocarcinoma in a 56-year-old female with a family kindred cancer history. Berry et al. (2017) recorded another 48-year-old female with a Meckel's diverticulum harbouring GIST. Mora-Guzman et al. (2018) also described neuroendocrine tumours found in three patients with Meckel's diverticulum (Sakpal et al. 2009).

Overall, although tumours arising within Meckel's diverticulum are extremely rare, the benefits of incidental resection in 'asymptomatic' patients, with a strong cancer family history(s), may merit some special consideration (Berry et al. 2017; Mora-Guzman et al. 2018).

42.8 Morbidity

Early postoperative complications are estimated in around 10% patients following Meckel's resection, including anastomotic leak, stricture, adhesions and postoperative ileus. Late events may include intestinal obstruction from adhesions. Whilst enthusiasm for minimally invasive surgery in children has undoubtedly greatly advanced a large US national database study (Ezekian et al. 2019) claimed to report no differences in outcome metrics in patients having 'open operation' vs MIS for Meckel's. A study by Ruscher et al. (2011), however, had a shorter hospital stay in those having MIS resection.

42.9 Umbilico-Ileal Fistula (Fig. 42.10)

A persistent fistula here usually presents in the newborn period, with discharge of intestinal content from the umbilicus with periumbilical excoriation (Fig. 42.10). Investigations (if indicated) may include a contrast fistulogram (or ultrasound) to confirm aberrant anatomy. Management includes resection via a cosmetic umbilical skin crease incision, identification of the fistula tract and intestinal resection with anastomosis.

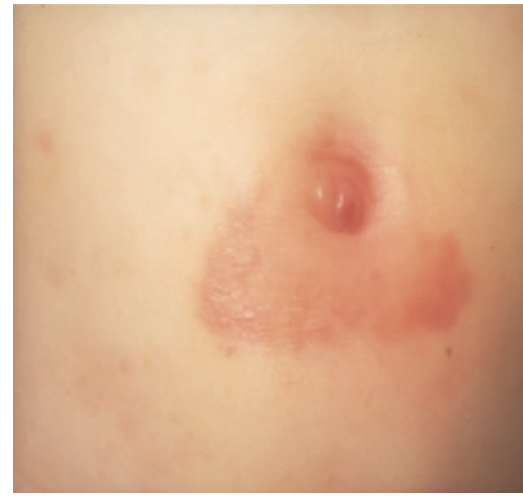

Fig. 42.10 Umbilical discharge from a patent vitellointestinal tract. Note the excoriation of the skin

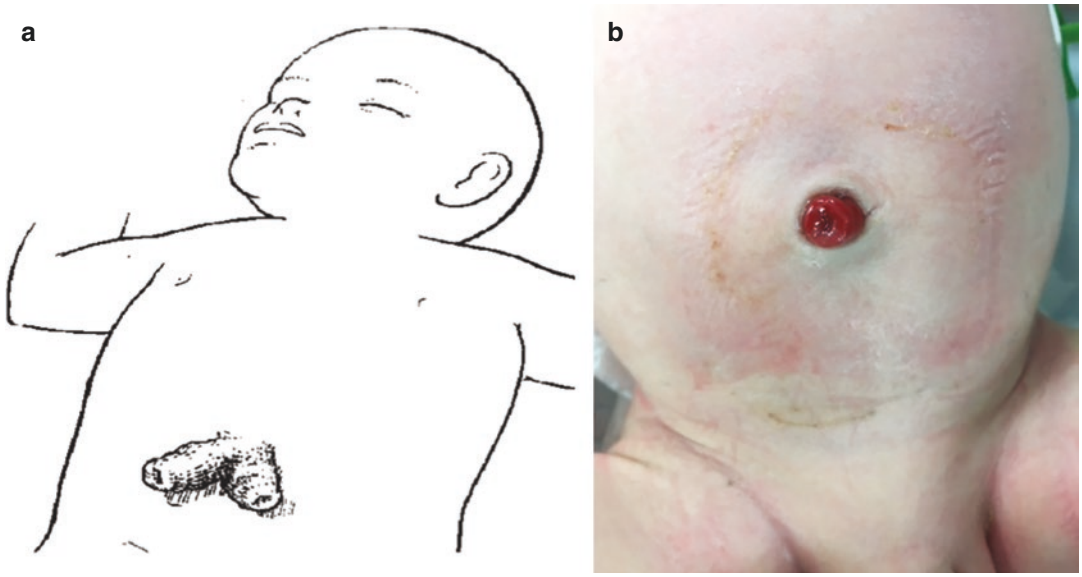

Fig. 42.11 (**a**) Prolapse of a patent's vitellointestinal tract with the classic ''double- horn'' anomaly; (**b**) shows newborn infant with a 'stoma-like' 'prolapsed vitello-intestinal mucosa'

Prolapse of a large patent vitellointestinal tract at the umbilicus presents as a characteristic 'double-horn' deformity with intestinal lumen clearly evident in the anomaly (Fig. 42.11a and b).

42.10 Umbilical Sinus

An umbilical sinus usually presents with a persistent serous or serosanguinous discharge from the navel area. When there is doubt about the nature of the discharge (often because of its intermittent nature), a radiology sinogram study may demonstrate the extent of the tract before formal resection.

42.11 Umbilical Cyst (Omphalomesenteric Cyst or Vitelline Cyst)

The cyst with the fibrous cord at either end of the tract can present with features of intestinal obstruction and is best managed by exploration and resection.

42.12 Persistent Fibrous Cord

Congenital fibrous bands are clinically significant as they may lead to intestinal obstruction or gut volvulus and must be resected when symptomatic.

42.13 Umbilical Polyp

Polyps related to the presence of an OMD remnant can occasionally present as a red nodular lesion in the umbilical dimple. They may contain tiny fragments of intestinal or gastric mucosa. One should also consider the more common diagnosis notably umbilical granuloma. Exploration of umbilical polyp lesions to exclude underlying omphalomesenteric duct anomalies is controversial. According to a recent useful study by Pacilli et al. (2007), exploration in these circumstances revealed 46% negative findings, i.e. no connection with the gastrointestinal intestinal tract, whilst another 54% of cases did not develop clinical symptoms after only simple polyp excision without exploring the abdominal cavity.

42.14 Conclusion

Omphalomesenteric duct remnants are fascinating developmental lesions that may present variantly at any age range though most notably will be encountered and managed by paediatric surgeons. It is therefore key that surgeons are wholly familiar with care pathways to expedite successful management and outcomes.

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