47 Omphalomesenteric Duct and Urachal Remnants

Nada Sudhakaran and Bruce Okoye

Abstract

The umbilical cord remnant usually separates in the neonatal period and its persistence beyond the first couple of months is considered abnormal.

Umbilical abnormalities may present with failure of the umbilical cord to separate, omphalitis, mass lesions, or discharge. The commonest umbilical lesion in the neonate is an umbilical granuloma. Other abnormalities are umbilical polyps, omphalomesenteric duct and urachal remnants. It is essential to distinguish between these conditions in order to initiate appropriate treatment.

Keywords

Human embryology • Umbilical disorders • Meckel's diverticulum Urachal abnormalities

47.1 Introduction

The umbilical cord remnant usually separates in the neonatal period and its persistence beyond the first couple of months is considered abnormal [1].

Umbilical abnormalities may present with failure of the umbilical cord to separate, omphalitis, mass lesions, or discharge. The commonest umbilical lesion in the neonate is an umbilical granuloma [1, 2] Other abnormalities are umbilical polyps, omphalomesenteric duct and urachal remnants. It is essential to distinguish between these conditions in order to initiate appropriate treatment.

47.2 Omphalomesenteric Duct Remnant

47.2.1 History

Fabricius Hildanus was the first to report this congenital anomaly in 1598 [3, 4]. Morgagni further defined the anatomy and clinical presentation of Meckel's diverticulum [5–7].

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In 1809 Johann Friedrich Meckel, described the embryology and the clinical features of this condition. His study of 22 paediatric cadavers gave rise to his description of the various forms of omphalomesenteric duct remnants namely, omphalomesenteric fistulas, omphalomesenteric cysts, umbilical sinuses and mesodiverticular bands. Meckel deduced that these malformations arose from the incomplete obliteration of the omphalomesenteric duct [4, 8, 9].

47.2.2 Epidemiology

The commonest Omphalomesenteric duct (OMD) remnant is the Meckel's diverticulum (MD). MD is also the commonest congenital abnormality of the gastrointestinal tract. It is referred to as the disease of "2s": It occurs in 2% of the population, arises 2 feet from the ileocoecal valve (adults), is about 2 in. long, about 2 cm in diameter, symptoms are often seen before the age of 2 and males are reported to be twice more likely to present with clinical symptoms [10, 11].

Meckels diverticulum is sporadic, but its presence is reportedly increased in children with Hirschsprung's disease, Down syndrome, esophageal atresia, duodenal atresia, malrotation, and congenital cardiac abnormalities [1].

47.2.3 Embryology

The yolk sac, is an extra embryonic extension from the primitive mid gut. This is formed by the 4th week of gestation. As the cranial and caudal body of the embryo folds, the neck of the yolk sac narrows. The lateral edges of the embryonic disk then start to fuse in the midline. The ectoderm covers the entire embryo, except where the yolk sac and connecting stalk emerge.

By the 6th week of gestation, the yolk sac is narrowed to a slim stalk now, known as the vitelline duct, omphalomesenteric duct or the yolk stalk [12] (Fig. 47.1).

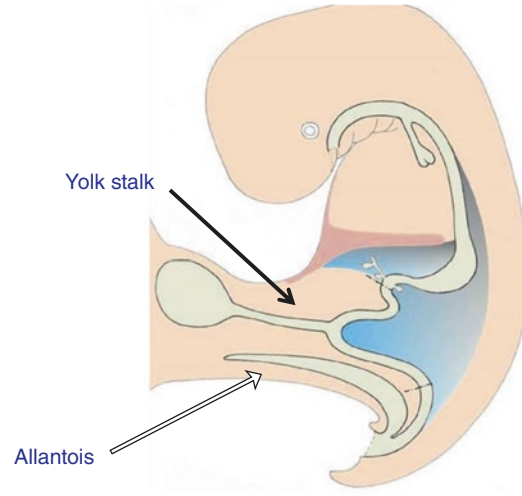

Fig. 47.1 Diagram of a fetus. *Black arrow*: Yolk stalk attached to the yolk sac on the left and the developing midgut on the right. *White arrow*: The allantois, which later becomes the urachus

The yolk stalk is thought to provide nutrients from the yolk sac to the developing embryo [13] After the 6th week, the yolk sac and yolk stalk disappear, along with the vitelline arteries [14]. Failure of this regression, creates the various forms of omphalomesenteric remnants. Although a number of other OMD anomalies can occur, MD is by far the most common.

As the yolk sac is continuous with the developing intestine, it contains all the layers of the intestinal wall as does a MD. Approximately 55–70% of MD contain ectopic tissue, usually either heterotopic gastric or pancreatic mucosa [15, 16]. The exact cause for this ectopic tissue in the diverticulum is unknown [17–19]. There have been suggestions that small buds of the pancreas are left on the foregut prior to its fusion, which then moves with the elongation of the gut onto the MD [20, 21]. There have been case reports of other ectopic tissues such as, colonic, duodenal, jejunal, hepatic, and endometrial, however these cases are rare and are isolated findings [22–25].

47.2.4 Anatomy

Meckels diverticulum is usually located on the antemesenteric border of the ileum [26]. It contains all five layers of the small intestine and is supplied by the vitelline artery (Fig. 47.2). This feature distinguishes it from a duplication cyst. The vitelline artery arises directly off the aorta [13, 27–30]. In addition to the mesenteric location of the ileum, MD has also been reported involving the proximal jejunum and the rectum [31]. The MD may be free (74%) or attached (26%) by fibrous bands to the umbilicus [32].

47.2.5 Clinical Presentation

Omphalomesenteric duct (OMD) remnants present clinically with a complication at an incidence of 4—6% [26, 33]. It has been noted that this incidence decreases with age. Clinical presentation is very varied and is related to the degree and pattern of patency or obliteration of the OMD. This may range from a completely patent omphalomesenteric duct at the umbilicus communicating with the bowel to a variety of lesser remnants, including the MD.

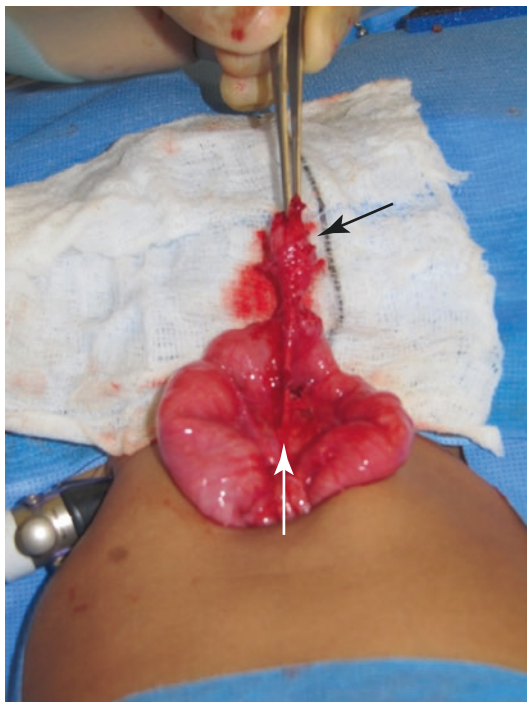

Fig. 47.2 *Black arrow*: Meckel's diverticulum; *White arrow*: vitelline artery arising from the mesentery, supplying the MD

Omphalomesenteric duct remnants may present with the persistent discharge of bowel content or mucus from the umbilicus, intussusception, prolapse of ileum at the umbilicus, intestinal obstruction, melena, anaemia and peritonitis [2].

Symptoms occur most frequently during childhood years (especially in the first 2 years of life) [34] The commonest modes of presentation are obstruction (30%), bleeding (27%), intussusception (19%), omphalitis (1%), and others (23%) [11, 35]. In the neonatal period MD may present with perforation, intussusception, ileal volvulus and less commonly, bleeding [2, 11].

Bowel obstruction is usually due to a mesodiverticular band, which is a fibrous remnant of the vitelline artery. This band, extending from the mesentery into the diverticulum, may trap a portion of the bowel [4, 36, 37]. In addition, volvulus of the bowel may occur around a persistent vitelline duct or band which connects the diverticulum to the umbilicus [16]. This may lead to bowel obstruction, perforation and peritonitis. Less commonly, an axial torsion of the MD may also occur. This occurs around its base when it is attached to either the umbilicus or ileal mesentery [26, 38]. Perforation of the MD may occur due to distal intestinal obstruction such as with Hirschsprungs disease or distal atresia [39].

Gastrointestinal bleeding is an important clinical presentation of MD. The incidence of bleeding in childhood has been recorded as high as 70% [4]. Bleeding occurs due to the presence of gastric or pancreatic tissue within the MD. Gastric tissue tends to be the more prevalent of the two, seen in 60–65% of cases, with pancreatic tissue seen in 5% of cases [15, 16]. The acidic secretions of the gastric tissues and alkaline secretions from the pancreatic tissues cause ulcerations to the adjacent normal ileal mucosa at the base of the MD, often upstream. This ultimately leads to the early detection of the diverticulum and may

explain why it is most commonly found in children [22, 25]. The bleeding is often bright red fresh bleeding if large in volume or may be darker in colour. Melaena is unusual. The painless bleeding can be catastrophic, sometimes requiring urgent blood transfusion [4].

Ileo-ileal intussusception results when the MD, an aperistaltic segment of ileum, is pushed into the adjacent ileum or when the MD falls into the bowel lumen becoming a lead point for the intussusception. The intussusception may progress into the colon becoming ileo-colic [4].

MD have also been reported within inguinal or umbilical herniae (Littres hernia) [36, 40].

A small proportion of OMD may present with omphalitis, often, due to an infected OMD cyst. These cysts are what remains when the umbilical and bowel margins of the OMD obliterates but the central portion remains patent. Inflammation within an MD may occur but is unusual in the neonatal period. In addition, OMDs may present as an umbilical sinus, an unconnected collection of ectopic mucosa of ileal or gastric origin or pancreatic tissue at the umbilicus [10].

47.2.6 Management

Management varies with clinical presentation. Most importantly, following acute presentation such as bleeding, bowel obstruction, intusscusception or peritonitis, adequate rescusitation is the key priority. Crystalloid, blood products and antibiotics should be administered as needed with insertion of a naso gastric tube to aid bowel decompression and prevent pulmonary aspiration.

In some cases, such as with bowel volvulus, obstruction or peritonitis, the diagnosis will only be made following emergency laparotomy or laparoscopy. Surgery in cases of peritonitis or suspected bowel ischaemia must not be delayed in an attempt to obtain a precise diagnosis. Differential diagnoses in such cases will include the full spectrum of possible causes of acute abdomen or obstruction in the neonate such as malrotation, intussusception or bowel atresia.

In a stable child with an uncertain diagnosis further investigation may include plain radiography, ultrasonography, or imaging of the small and large bowel through contrast follow through or enema.

Obvious umbilical lesions with prolapsed intestinal mucosa would require surgical resection. If the baby presents with omphalitis or a mass under the umbilicus, then an ultrasound scan can be done prior to surgery. If the diagnosis is still unclear following radiological investigations in a stable child, diagnostic laparoscopy may be useful. Laparoscopy is increasingly used in both the diagnosis and treatment of MD. The diverticulum can be exteriorized via a periumbilical incision allowing either diverticulectomy or segmental resection and reanastomosis [41–45].

A "well" baby presenting with significant rectal bleeding will require a Meckel's scan. This scan utilizes Tc99 sodium pertechnetate given intravenously. The presence of ectopic gastric mucosa is highlighted by scintigraphy (Fig. 47.3).

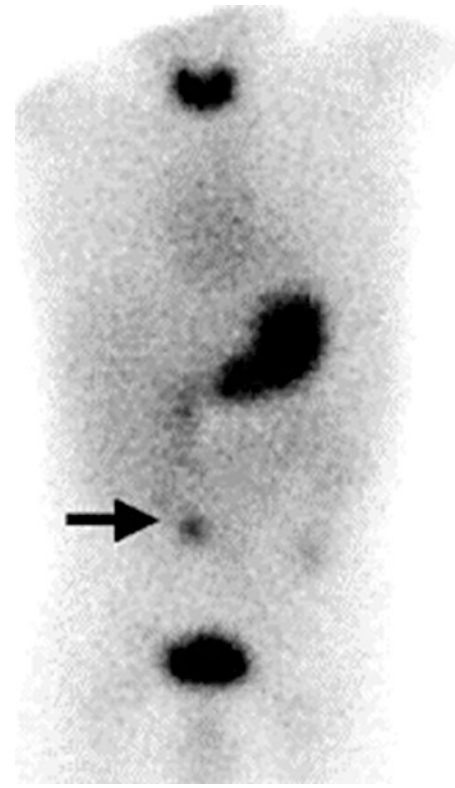

Fig. 47.3 Meckels Scan using Tc99 sodium pertechnetate, the arrow showing an area of ectopic gastric mucosa. Also highlighted are the thyroid gland, stomach and the urinary bladder

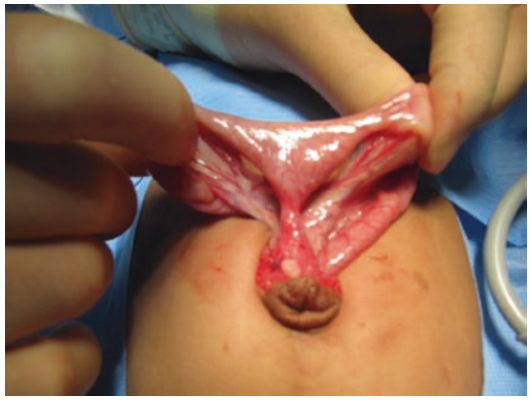

Fig. 47.4 Meckel's diverticulum attached to the umbilicus

The isotope has a high affinity for parietal cells of gastric mucosa. The residual isotope is concentrated in the urinary bladder. A positive scan shows abnormal uptake of the isotope outside the stomach and urinary bladder. The Meckel's scan has a reported sensitivity of 25–92% [41, 46–50]. Prescribing pentagastrin, histamine-2 (H-2) blockers and glucagon, have been reported to increase the diagnostic yield of the Meckel's study [33, 51]. In view of the wide variation in the sensitivity of the meckles scan, consideration should be given to early laparoscopy or laparotomy in children with suspicious clinical presentation (Fig. 47.4).

MD presenting with intestinal bleeding should have a segmental ileal resection along with the MD as the bleeding is often form ulcerated ileal mucosa adjacent to the MD. In addition, the heterotropic tissues may be found at the base of the MD. Hence a simple diverticulectomy is insufficient. A wedge excision or a segmental ileal resection would ensure complete resection of abnormal tissue [33].

47.2.7 Incidental Finding of Meckel's Diverticulum

Meckels diverticulum may be found incidentally during laparotomy or laparoscopy. There is varying opinion regarding the need to resect the MD in this situation. It has been suggested that diverticuli less than 2 cm in length, with no heterotopic palpable mucosa, constitute a lower risk group [52]. There are concerns that resecting an MD in a "clean" operation potentially converts it into a "dirty" or contaminated one. In addition, it is argued that the risk of the MD becoming symptomatic is small and that resection could result in a longer hospital stay with a risk of anastamotic leaks bowel obstruction, or infection [17, 53].

Proponents of resection suggest that the morbidity or mortality of the primary procedure may not be increased and that the palpable characteristics of the diverticulum may be unreliable [54, 55]. Tumors have rarely been reported within MD. These may be benign, such as lipoma, neuromuscular and vascular hamartomas, or malignant with carcinoids making up the majority of such cases [4, 56].

Two large studies looking at 50 years of data have shown an approximately 6% risk of complications arising from MD. Diverticulectomy performed in the presence of complications carries an operative mortality and morbidity of approximately 2% and 12% respectively [51, 57]. However, this risk must be weighed against the risks of complications from an incidental resection, a morbidity figure of around 1—2% [51, 57].

47.3 Urachal Abnormalities

The urachus is a fibrous, midline, tubular structure that extends from the dome of the bladder to the umbilicus. It represents an incomplete regression of the allantois. Urachal remnants may be completely asymptomatic but can also cause significant morbidity.

47.3.1 Epidemiology

Urachal remnants are considered rare. In pediatric autopsy studies, an incidence of 1 in 7610 cases for patent urachus and 1 in 5000 cases for urachal cysts has been documented [58]. However the incidence of symptomatic presentation with a urachal remnant is significantly smaller with the most common abnormality being urachal cysts [59].

In one review of 56 children with urachal abnormalities, about half were identified incidentally [60]. Babies with umbilical discharge and a patent urachus usually present at birth while non discharging anomalies usually present before 5 years of age [61, 62].

47.3.2 Embryology

The allantois is a finger like projection, connected to the cloaca of the primitive hindgut. The cloaca separates to form the urogenital sinus anteriorly and the rectum posteriorly [63–66] (Fig. 47.1).

The fetal bladder descends from the umbilicus into the pelvis around the fourth or fifth month of gestation. The allantois, which is attached to the dome of the bladder, stretches and progressively narrows down. It forms an epithelialized fibromuscular tube, the urachus. The urachus obliterates by fibrosis and forms the median umbilical ligament by about the 4th or 5th month of gestation [66, 67]. The precise aetiology of urachal anomalies remains undefined, however its presence has historically been attributed to bladder outlet obstruction. This "pop-off" anatomic theory is not well supported in the literature. One study reports up to 14% of patients with urachal abnormalities had evidence of bladder outlet obstruction, this finding is disputed in larger series [68, 69]. Urachal remnants can present as Umbilicourachal sinus (an incomplete tract from the umbilicus) or a complete one (patent urachus), urachal cysts or a vesicourachal diverticulum (Fig. 47.5). The most common abnormality, urachal cyst, can occur anywhere between the bladder and umbilicus and mostly occur in the distal third of the urachus. Vesicourachal diverticuli are rare, consisting of outpouchings of the bladder at the insertion of the urachus [69]. Other genitourinary conditions such as vesico-ureteric reflux, hypospadias and crossed renal ectopia are associated with urachal anomalies [70, 71].

47.3.3 Anatomy

The urachal remnant remains as a fibrous band lying in the retropubic, preperitoneal, perivesical space between the transversalis fascia and the parietal peritoneum, extending from the dome of the bladder to the umbilicus [65, 67] (Fig. 47.6). Its length varies from 3 to 10 cm and from 8 to

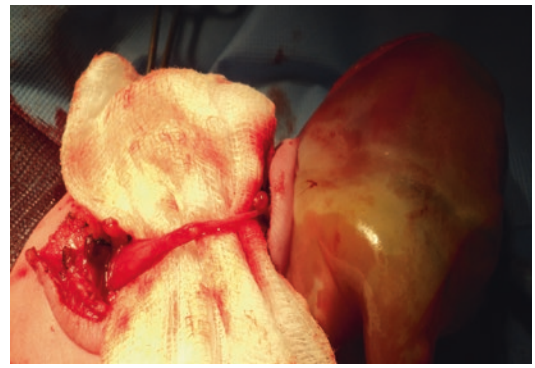

Fig. 47.6 Patent urachus attached to the bladder on the left, extending into an omphalocoele sac

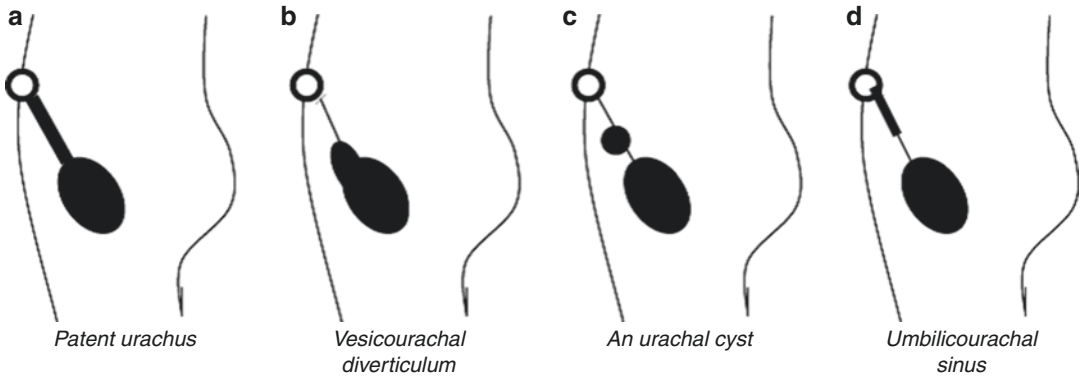

Fig. 47.5 Different forms of urachal remnants

10 mm in diameter [72, 73] Coexistence of a vitelline and urachal remnant is also uncommon, although reported [74].

47.3.4 Clinical Presentation

A patency of the embryologic urachal remnant after birth may give rise to various clinical problems. These include umbilical discharge, local infection, lower abdominal pain, and urinary tract infection. They may also be asymptomatic and be discovered incidentally [61, 62]. In a recent series, with 56 patients, the presentation was as follows: umbilical discharge (43%), umbilical infection (43%), and palpable cysts or masses associated with pain (14%). In this same group of patients, further investigations revealed that 14% had an associated genitourinary abnormality including vesico-ureteric reflux, a duplicated collecting system, hypospadias, meatal stenosis, bladder diverticulum, periurethral polyp, and renal dysplasia [69]. A patent urachus is estimated to account for about 10–15% of urachal anomalies [75]. These may sometimes present as a pseudocyst of the cord in the antenatal/fetal period [76].

As urachal remnants are rare, the literature on presentation in the neonatal period consists largely of case reports. These include prolapse of the urinary bladder, giant umbilical cord or with an omphalocoele [77–80]. There is a report of urinary ascites following trauma to a urachal remnant during umbilical artery catheterisation [81]. A subtle clinical sign of the presence of urachal remnant is the retraction of the umbilicus during voiding. This is often associated with pain [82]. Some urachal cysts are identified on ultrasound scan for another indication [83].

In an older child or young adult, infected urachal cysts can present with signs mimicking appendicitis, this is often an unsuspected finding at operation [84, 85].

47.3.5 Management

Clinical management depends on the mode of presentation. If the baby presents with an infected urachal cyst or urinary tract infection, it is important to treat the acute condition, with antibiotics, and appropriate fluid resuscitation. Ultrasound scan will assist with confirming the diagnosis and planning definitive treatment [86]. The presence of an abscess is traditionally managed in two stages: initially with antibiotics and drainage (either surgical or via interventional radiology), followed by delayed resection once the infection has resolved [86–89].

A micturating cysto-urethrogram (MCUG) may not always provide the diagnosis of a urachal remnant, especially if there is no connection with the bladder. However it may be informative in patients with a patent urachus and in whom a posterior urethral valves are a consideration [61, 69, 90]. A recent large study of 66 children with urachal remnant, from the Mayo clinic showed that of those who had a MCUG, 71% had grade 3 or less of vesico ureteric reflux and 12% had grade 4/5 reflux [91].

There have been reports of a patent urachus closing in the early newborn period. Some centres would advocate following some of these asymptomatic urachal remnants with serial ultrasound scans and conservative management [60, 75, 92]. However the long term risks of leaving these urachal remnants are stone formation and malignancy. The risk of future cancer in urachal remnants is well recognized. Urachal cancers account for 1–10% of adult bladder cancers, with a 10-year disease-free survival of about 50% [93, 94]. There have also been reports of cancers arising from urachal remnants in adolescence [95]. Urachal cancers are usually adenocarcinomas, although transitional cell, squamous cell and sarcomas have been reported [96–98].

For these reasons, the treatment of choice should be surgical resection. The tract along with the cyst and a small cuff of bladder at the insertion are removed. Mucosa should not be left at the umbilicus because of the concern that the urachal remnant may harbour a future carcinoma. This procedure can be performed by either open techniques or via laparoscopy. An open procedure may be performed via a curvilinear umbilical incision in infants. A transverse incision midway between the umbilicus and the pubis provides better exposure in older children [43–45, 99–101].

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