

Chapter 1

Embryology of the Umbilicus and Associated Newborn Defects

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1.1 Introduction

The embryology of the umbilicus and the developmental basis for surgical abnormalities has been well described for more than 100 years. Umbilical hernias, abdominal wall defects, umbilical polyps and drainage, and omphalomesenteric remnants are well described [1].

Treatment of some umbilical disorders, such as umbilical granulomas with silver nitrate, has changed little over the last century. In the early 1900s, umbilical hernia repair was a challenging procedure. Spontaneous closure of these hernias and preservation of the appearance of the natural umbilicus were recognized. Today, umbilical hernia repair is one of the most common procedures performed by pediatric surgeons. The umbilicus is currently a common site for port entry for laparoscopic surgery.

Newborns with umbilical disorders usually present with an abdominal wall opening, drainage, a mass, or combination of findings. Most umbilical disorders result from failure of normal embryologic or physiologic processes. Unusual umbilical anatomy, such as a single umbilical artery or abnormal position of the umbilicus, may be associated with other congenital anomalies or syndromes [2, 3]. Omphalocele and gastroschisis are common major abdominal wall defects associated with the umbilicus [4]. Ectopia cordis and bladder exstrophy are other major abdominal wall defects and are beyond the scope of this chapter [5].

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The frequencies of the many different umbilical disorders vary. Umbilical infections are now identified in fewer than 1% of hospitalized newborns.

Umbilical hernias are commonly identified in early infancy, however, most spontaneously close [6]. No sex predilection is noted. The incidence at age 1 year ranges from 2 to 15%. Incidence is increased in infants who are black and in infants with low birth weight, Down syndrome, trisomy 13, trisomy 18, or Beckwith–Wiedemann syndrome [7].

1.2 Embryology

The development of the anterior abdominal wall depends on differential growth of embryonic tissues (Fig. 1.1) [8]. As the embryo grows, the yolk sac is divided into an intra-coelomic portion and an extra-coelomic portion. The intra-coelomic

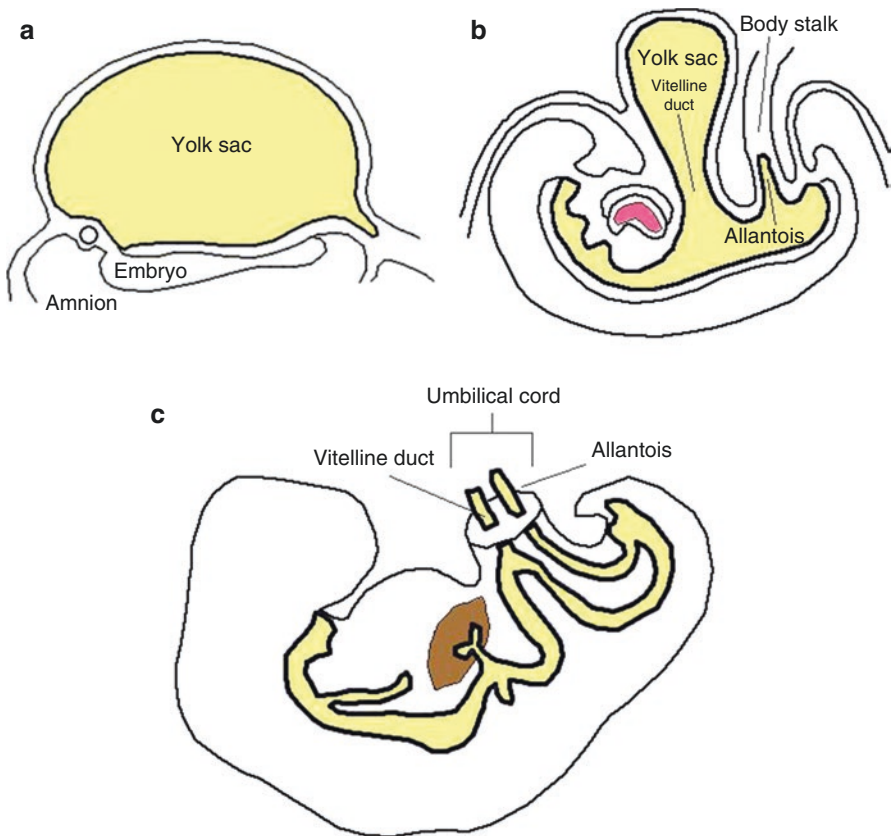


Fig. 1.1 The developing umbilical cord. (a) Embryonic disk: At this stage, the ventral surface of the fetus is in contact with the yolk sac. (b) The yolk sac narrows as the fetus grows and folds. The intra-coelomic yolk sac forms the intestine and communicates with the extra-coelomic yolk sac through the vitelline duct. The vitelline duct is also referred to as the omphalomesenteric duct and the yolk stalk. The allantois has begun to grow into the body stalk. (c) The yolk and body stalks fuse to become the umbilical cord (Image used with permission from Minkes et al. [28])

portion becomes the primitive alimentary canal and communicates with the extra-coelomic portion through the vitelline duct, also known as the omphalomesenteric duct. This communication is lost at 5–7 weeks' gestation. Persistence of part or all of this connection results in omphalomesenteric anomalies.

In the third week, the yolk sac develops a diverticulum, the allantois, which grows into the body stalk. As the distal hindgut and the urogenital sinus separate, the developing bladder remains connected to the allantois through a connection called the urachus [9]. Persistence of the urachus or urachal tissue leads to urachal remnants. Subsequently, the yolk and body stalks fuse to become the umbilical cord. Development of the abdominal wall narrows the umbilical ring, which should close before birth. Persistence of the ring results in an umbilical hernia.

Failure of the normal obliterative processes of the vitelline duct and the urachus leads to abnormal communications or cysts. Retention of components of the umbilical cord can also produce a mass or drainage.

A patent umbilical ring at birth is responsible for most umbilical hernias. The umbilical opening is usually inferiorly reinforced by the attachments of the median umbilical ligament (the obliterated urachus) and the paired medial umbilical ligaments (the obliterated umbilical arteries) and is more weakly superiorly reinforced by the round ligament (the obliterated umbilical vein) (Figs. 1.2 and 1.3).

Richet fascia, derived from the transversalis fascia, covers the ring. The peritoneum covers the innermost portion of the ring. Variability in the attachment of the ligaments and the covering by Richet fascia may predispose some children to developing umbilical hernias. This fascia may completely or partially cover the umbilical ring or be completely absent. These variations predispose to umbilical hernia formation and are responsible for the range of defects encountered clinically. It is important to note that many children undergo spontaneous closure in the first few years of life. The pressure exerted on the umbilical skin, even when only a small umbilical defect is present, can result in marked stretching of the skin and a proboscis appearance.

Masses of the umbilicus may be related to lesions of the skin, embryologic remnants, or an umbilical hernia. Masses associated with the skin include dermoid cysts, hemangiomas, and inclusion cysts. Umbilical drainage is associated with granulomas and embryologic remnants. Failure of the normal physiologic processes results in postnatal disease. The umbilical cord separates from the umbilicus between 1 and 8 weeks postnatally [10, 11]. Topical antimicrobials are applied after delivery, followed by isopropyl alcohol until the cord separates. Delayed separation of the cord may signify an underlying immune disorder [12].

1.3 Umbilical Granuloma

Umbilical granuloma is a common finding resulting from persistent granulation tissue after cord separation. Umbilical granulomas appear as 1 mm to 1 cm or more pink friable tissue at the base of the umbilicus. They produce variable amounts of drainage that can irritate the surrounding skin. Histologically, this tissue is composed of fibroblasts and capillaries. Most respond to silver nitrate, and larger lesions

Fig. 1.2 Umbilical region viewed from the posterior surface of the abdominal wall of an infant with the umbilical cord attached. *UA* umbilical artery, *UV* umbilical vein, *RL*, round ligament (obliterated umbilical vein), *UR* umbilical ring, *UL* umbilical ligament, medial (obliterated umbilical arteries), median (obliterated urachus). Note fascial covering of surface and umbilical ring (Image used with permission from Minkes et al. [28])

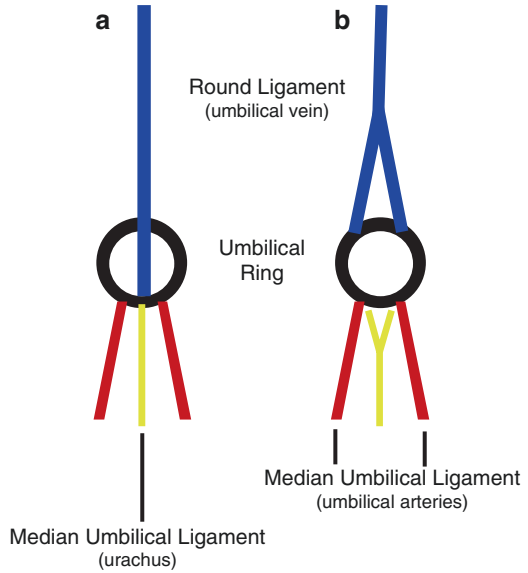
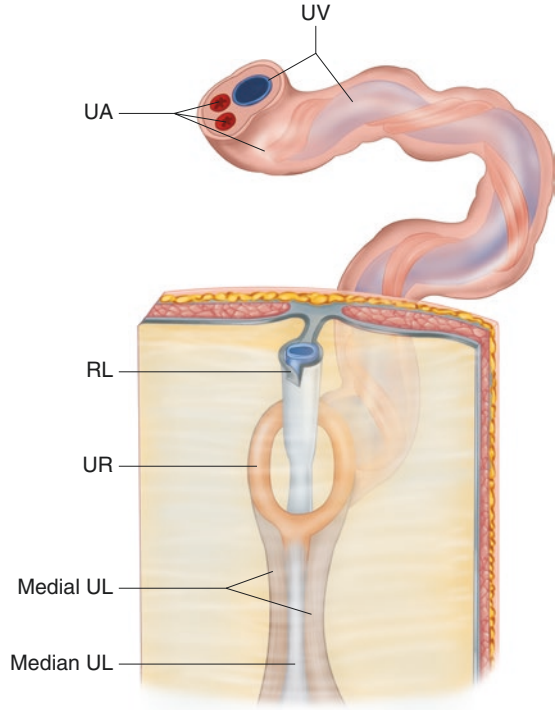


Fig. 1.3 Variations in the umbilical ring structure. (a) Usual configuration of the round ligament and urachus. (b) Less common configuration that can result in weakness at the umbilical ring (Image used with permission from Minkes et al. [28])

can often be safely excised with scissors in the office setting [13]. Persistent or recurrent granulation may suggest an umbilical polyp and warrant further investigation or umbilical exploration. An umbilical polyp is brighter red than a granuloma and represents retained intestinal or gastric or urogenital tissue from the omphalomesenteric duct or urachus [1, 13–15].

1.4 Umbilical Infections

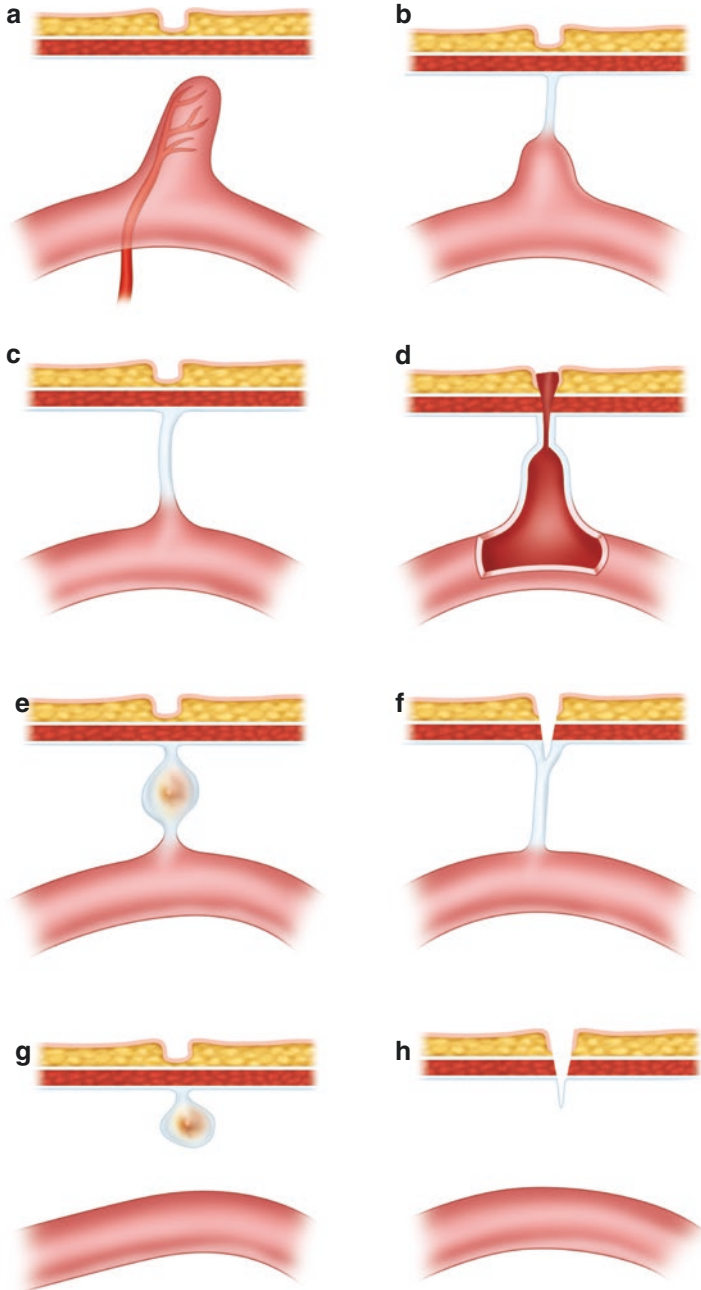
Umbilical infections can occur because of an embryologic remnant or poor hygiene. Newborns or infants with omphalitis may present with purulent umbilical discharge or periumbilical cellulitis. Current aseptic practices and the routine use of antimicrobials on the umbilical cord have reduced the incidence of umbilical infection to less than 1%. Umbilical infections can be life threatening in the newborn period. Patients with umbilical infections can present with drainage from the umbilicus, swelling, and redness. Cellulitis may rapidly progress and lead to necrotizing fasciitis. Necrotizing fasciitis is characterized by abdominal distention, tachycardia, purpura, leukocytosis, and other signs of sepsis that may not respond antibiotic therapy. A simple appearing cellulitis may become severe within hours and progress to necrotizing fasciitis and generalized sepsis.

Traditionally, gram-positive organisms, such as *Staphylococcus aureus* and *Streptococcus pyogenes*, were most commonly identified. Gram-negative and polymicrobial infections are seen today, especially in rapidly progressing cellulitis and necrotizing fasciitis. For necrotizing fasciitis and gangrene of the umbilical skin, emergency surgical debridement is required and can be life-saving.

1.5 Omphalomesenteric and Urachal Remnants

The presentation of omphalomesenteric and urachal remnants represent persistence of all or portions of the omphalomesenteric (vitelline) duct. They can result in fistulas, sinus tracts, cysts, congenital bands, and mucosal remnants [16]. Omphalomesenteric remnants and urachal remnants require surgical excision. The precise diagnosis is often not confirmed until surgery is performed, and the anatomy of the umbilicus is established. The presentation depends on the specific type of defect (Fig. 1.4). If a communication persists from the terminal ileum to the umbilicus, intestinal contents or stool can be observed leaking from the umbilicus. Prolapse of the intestine through an omphalomesenteric fistula can also be observed (Fig. 1.5). The drainage from a fistula that does not communicate with the ileum varies, as it may be clear, bloody, or purulent. Cystic remnants may become infected and manifest with pain and swelling.

The developing bladder remains connected to the allantois through the urachus, and remnants of this connection include a patent urachus, urachal sinus, and urachal cysts. Umbilical polyps can also be observed in association with a urachal remnant. The presentation of urachal remnants also varies. Clear drainage from the umbilicus



Patent Omphalomesenteric Duct

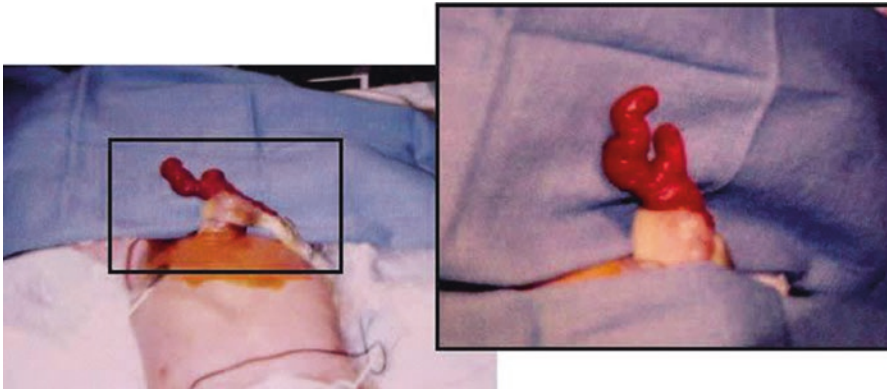


Fig. 1.5 (Left and Right) Newborn with intestinal prolapse through a patent omphalomesenteric duct. Both the proximal and distal limbs of the intestine have prolapsed. The umbilicus was explored, the bowel was easily reduced, and the patent duct was excised. The child was discharged from the hospital 2 days later (Image used with permission from Minkes et al. [28])

is characteristic of a urachal fistula. Drainage of urine from the umbilicus may suggest bladder outlet obstruction and warrants further investigation.

A urachal cyst is usually discovered as a painful mass between the umbilicus and suprapubic area when it becomes infected. Pain and retraction of the umbilicus during urination may suggest a urachal anomaly [17]. A urachal sinus manifests with drainage that can be clear or purulent and occurs through the umbilicus or midline skin below the umbilicus (Fig. 1.6).

1.6 Umbilical Hernia

Umbilical hernias result from persistence of a patent umbilical ring (Fig. 1.7). Most small umbilical hernias close spontaneously, but many require surgical repair [6, 7, 18]. Patients with umbilical hernias present early in life with bulging at the umbilicus. The swelling is most prominent when the infant or child is crying or straining. Umbilical hernias are usually asymptomatic and rarely cause pain but may become incarcerated



Fig. 1.4 Omphalomesenteric duct remnants. (a) Meckel's diverticulum. Note feeding vessel. (b) Meckel's diverticulum attached to posterior surface of anterior abdominal wall by a fibrous cord. (c) Fibrous cord attaching ileum to abdominal wall. (d) Intestinal-umbilical fistula. Intestinal mucosa extends to skin surface. (e) Omphalomesenteric cyst arising in a fibrous cord. The cyst may contain intestinal or gastric mucosa. (f) Umbilical sinus ending in a fibrous cord attaching to the ileum. (g, h) Omphalomesenteric cyst and sinus without intestinal attachments (Image used with permission from Minkes et al. [28])

Fig. 1.6 Urachal sinus with purulent drainage in midline below the umbilicus (*black arrow*). A laparoscope was placed in the supraumbilical crease (*red arrow*) for mobilization of the internal portion of the urachal remnant as depicted in the next image (Image used with permission with permission from Minkes et al. [28])

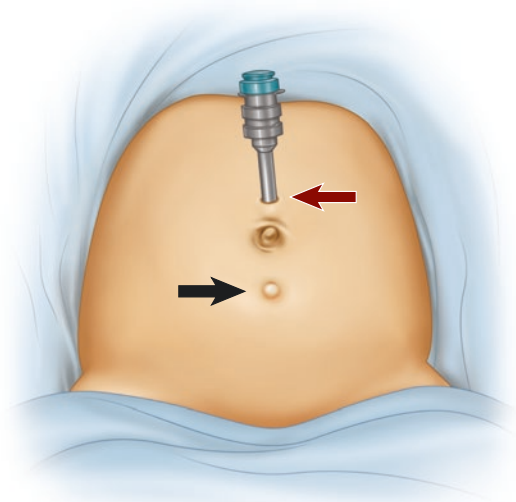


Fig. 1.7 Preoperative patient demonstrating umbilical hernia with redundant skin (Image used with permission from Minkes et al. [28])



especially later in life. The skin can become severely [18–20] stretched, which may be alarming to parents and physicians. Parents often mention that the child plays with the redundant skin.

The diameter of the umbilical ring defect is predictive of spontaneous closure. The length of the protruding skin is not prognostically significant. Umbilical hernias with ring diameters less than 1 cm are more likely to spontaneously close than those with ring diameters more than 1.5 cm. Surgery is indicated for all symptomatic umbilical hernias. Incarceration, strangulation, skin erosion, and bowel perforation are indications for immediate surgery. Similarly, patients presenting with pain should be repaired on an elective basis.

Asymptomatic umbilical hernias can be safely monitored until the child is aged 4–5 years to allow spontaneous closure, especially if the ring defect is small.

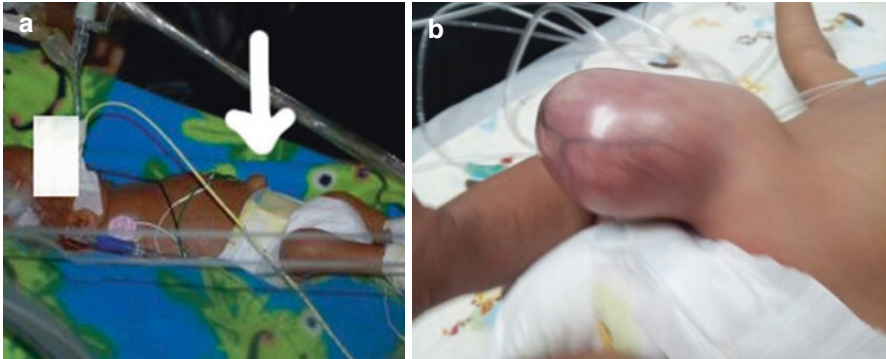


Fig. 1.8 (a) Small umbilical hernia (*arrow*) in premature infant. (b) Progressed into large symptomatic hernia. Note skin erythema that eventually resulted in skin ulceration

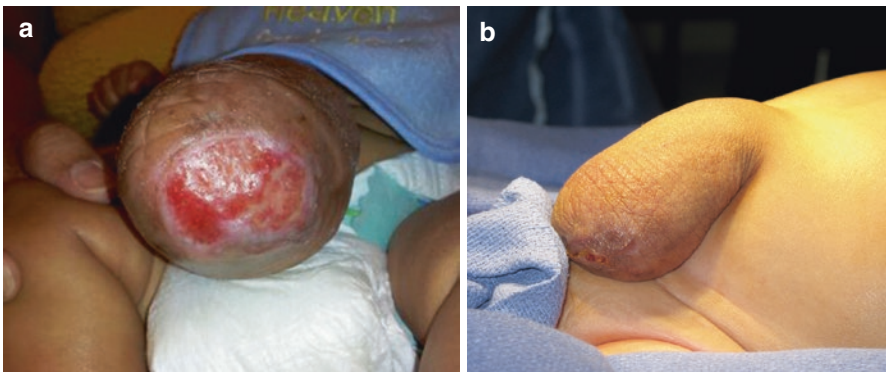


Fig. 1.9 (a, b) Skin ulceration and presentation at surgery. The hernia was no longer reducible

Because umbilical hernias with larger defects (i.e., >1.5 cm) are unlikely to close spontaneously, surgery can be performed at an earlier age.

Similarly, closing umbilical hernias with large ring defects is reasonable in younger children if the child is having a general anesthetic for another procedure, such as an inguinal hernia repair. It is also reasonable to consider surgery in younger children who have a large protrusion of the umbilical skin that is causing distress to the parents.

Incarceration, strangulation, bowel obstruction, erosion of the overlying skin (Figs. 1.8 and 1.9), and bowel perforation are rare events in infants and small children. The risk of incarceration increases significantly in adults with umbilical hernias.

1.7 Imaging

Radiography is not indicated in most children with umbilical disorders. Umbilical hernias are diagnosed by means of physical examination.

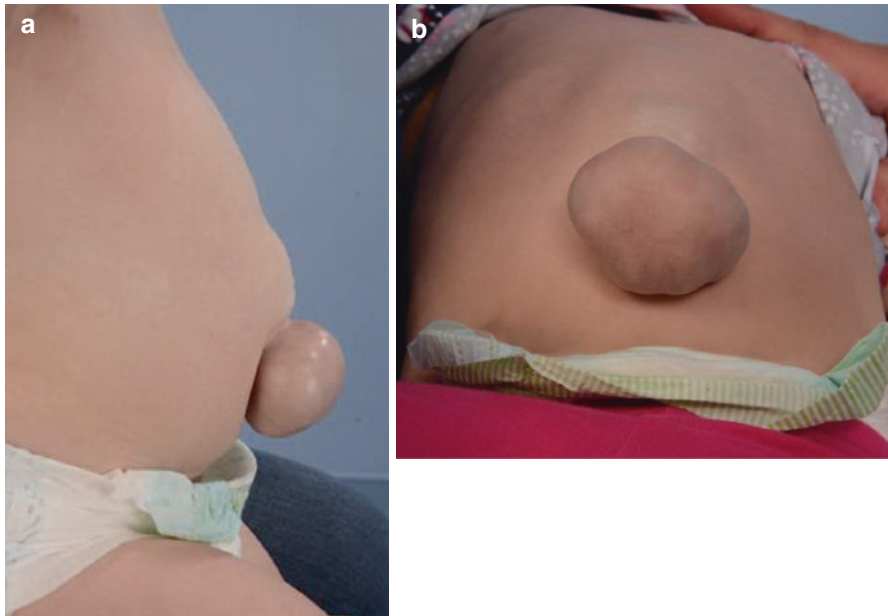


Fig. 1.10 (a, b) Complex abdominal wall defect with umbilical and associated ventral defect

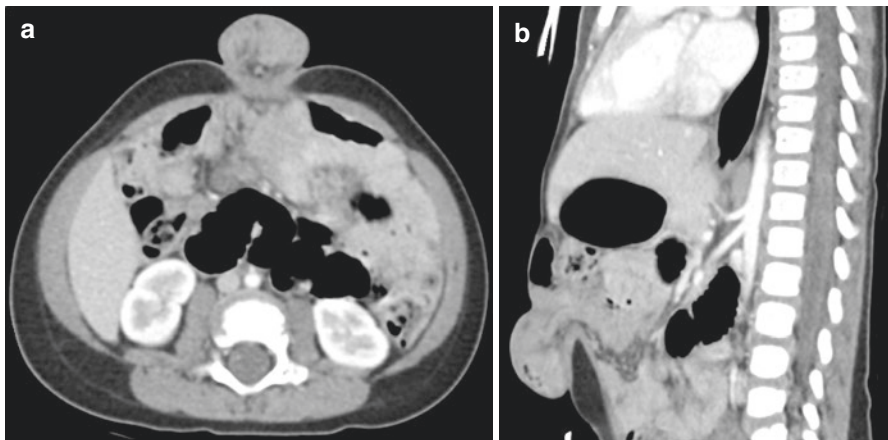


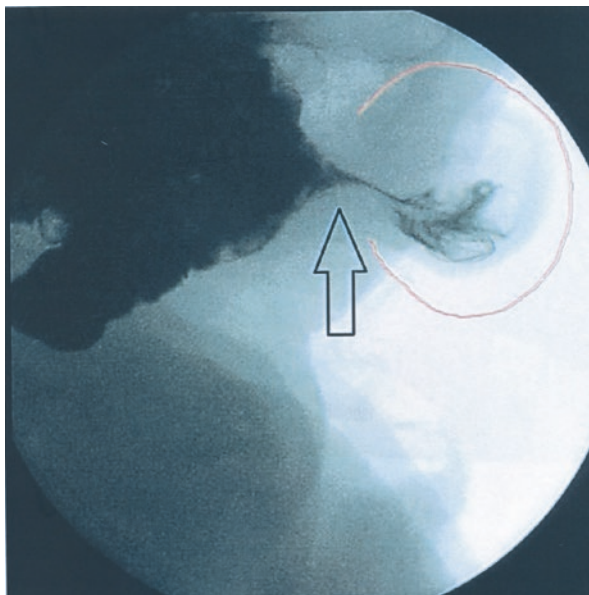
Fig. 1.11 (a, b) Computed tomography defining complex abdominal wall defect

Fistulography or sinography may be performed if a definitive opening is observed within the umbilicus. Fistulography can be performed by injecting water-soluble contrast medium into the opening at the base of the umbilicus. If the track is blind-ended, the child has a sinus; if it enters the intestine or bladder, a fistula is present.

Ultrasonography is helpful when a mass is present. Ultrasonography may be useful in identifying cysts of the umbilicus. Evaluating for a urachal cyst is useful; this cyst most commonly appears as a mass between the umbilicus and suprapubic area.

Computed tomography may be useful for surgical preparation for more complex abdominal wall defects (Figs. 1.10 and 1.11)

Fig. 1.12 Upper gastrointestinal contrast study showing incidental umbilical hernia in an infant. *Red line* outlines the umbilical hernia. The *arrow* shows contrast flowing into the intestine within the umbilical hernia. The umbilical hernia was easily reducible, and no intervention based on this study was performed (Image used with permission from Minkes et al. [28])



Plain radiography may be useful in children with omphalitis. Air in the subcutaneous tissue or muscle planes is an ominous sign. An umbilical hernia may be incidentally observed on an upper gastrointestinal tract contrast study with small-bowel follow-through (Fig. 1.12).

Cystography or cystoscopy may be indicated to identify bladder outlet obstruction in children draining frank urine from a urachal fistula. However, studies suggest that, in most cases, history and ultrasonography are sufficient for the diagnosis [21].

1.8 Technique

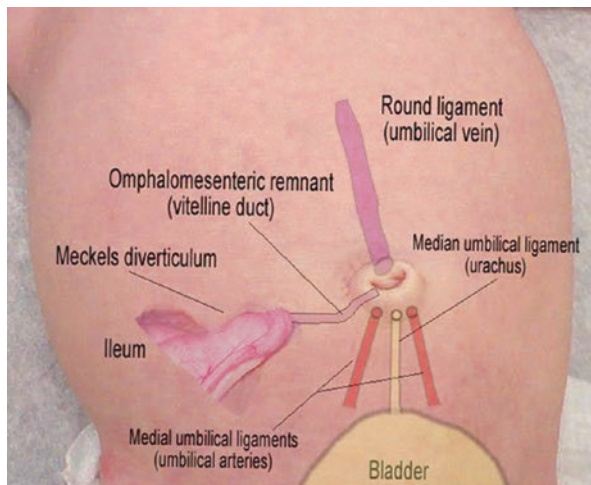
Surgical therapy is the mainstay of treatment for the following umbilical conditions:

1. Large persistent umbilical granulomas
2. Umbilical polyps
3. Omphalomesenteric remnants
4. Urachal remnants
5. Umbilical gangrene and necrotizing fasciitis
6. Umbilical hernias that are symptomatic or do not spontaneously close

Often, surgery on the umbilicus is performed for a mass or drainage without a specific preoperative diagnosis. Surgical principles include identification of all structures of the umbilicus, excision of urachal or omphalomesenteric remnants, closure of the umbilical ring, and preservation of the natural appearance of the umbilicus.

Laparoscopy is a valuable adjunct to open umbilical exploration [22]. It allows identification of both normal and abnormal structures. The laparoscopic approach can be used to remove urachal remnants, as well as omphalomesenteric abnormalities.

Fig. 1.13 Anatomic relationship between the umbilicus and its embryologic attachments (Image used with permission from Minkes et al. [28])



If acute infection with an abscess is present, surgical drainage is carried out in the operating room or by means of interventional radiology. In most cases, definitive surgical resection of the underlying lesion is needed several weeks following the initial infection.

Wide surgical debridement of the umbilicus and abdominal wall can be life-saving in patients with necrotizing umbilical infections [23–25].

Most umbilical procedures can be performed on an elective basis, and surgery should be scheduled when the child is in his or her usual state of health. The exceptions are infants with necrotizing infections and those with stool draining from the umbilicus, indicative of an enteric-umbilical fistula.

Mechanical bowel preparation is not needed. Perioperative antibiotics are used for sinuses and fistulas. Antibiotics are not needed for umbilical hernia repairs.

General anesthesia is used. The child is placed supine. The abdomen should be widely prepared with antiseptic solution and draped in standard fashion. Omphalomesenteric and urachal remnants can be approached directly through the umbilicus or through an incision in the infraumbilical or supraumbilical crease. Larger children may need additional or larger incisions. Laparoscopy can be performed through the umbilical incision.

During exploration of the umbilicus, an attempt is made to identify all anatomic structures (Fig. 1.13). A history of infection and resultant inflammation or scar tissue can complicate identification of normal and abnormal structures. Dissection of the umbilicus off the fascia is often useful and can provide access for a small port to perform laparoscopy. The umbilical vein (round ligament), the umbilical arteries (medial umbilical ligaments), and the urachus (median umbilical ligament) can often be identified.

A patent vitelline duct must be traced to its origin and divided. If a Meckel's diverticulum is present (see the image below), it is excised. Similarly, the urachus should be traced to its origin and divided. Broad-based connections of the urachus and bladder are closed in two layers.

Fig. 1.14 Laparoscopic removal of urachal cyst (*U*). *L* indicates the left medial umbilical ligament. *R* indicates the right medial umbilical remnant. *B* indicates the bladder. The distal attachment to the bladder is being grasped (Image used with permission from Minkes et al. [28])

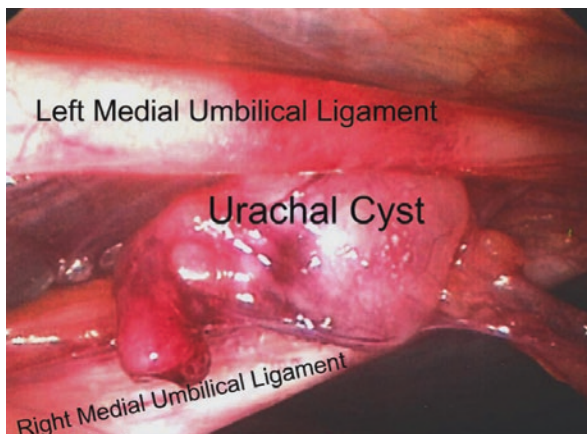
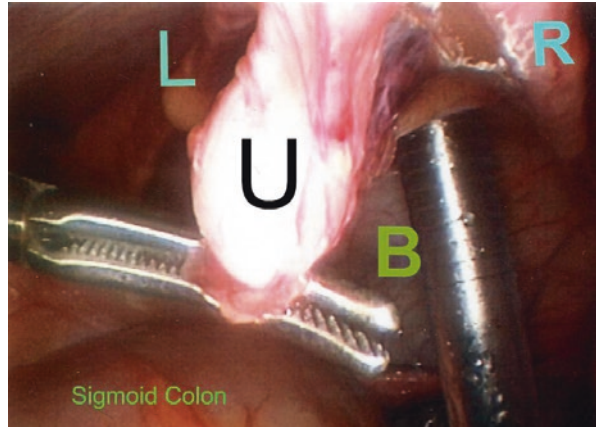


Fig. 1.15 Laparoscopic removal of urachal cyst. View is from left lower abdomen port. The umbilicus is on the right and the bladder on the left. The attachments of the urachal cyst to the bladder and the umbilicus have been clipped (not shown) and divided. Note the convergence of the right and left medial umbilical ligaments as they approach the umbilical ring on the right (Image used with permission with permission from Minkes et al. [28])

Laparoscopy is a useful adjunct, especially when there is a confirmed preoperative diagnosis or when no anomaly is found during the exploration (Figs. 1.14, 1.15, 1.16, 1.17, and 1.18) [5]. Additional ports can be placed to remove identified urachal or omphalomesenteric structures. Removed structures are sent to pathology for histologic examination.

The umbilical fascia is closed with interrupted or running suture. The umbilical skin is then closed. Attempts should be made to create a natural-appearing umbilicus. Antibiotic ointment and a light dressing can be applied to the incision. If an abscess is identified preoperatively or found intraoperatively, an incision-and-drainage procedure is indicated. Definitive resection should be delayed.

Fig. 1.16 Laparoscopically removed urachal cyst and its attachments (Image used with permission from Minkes et al. [28])

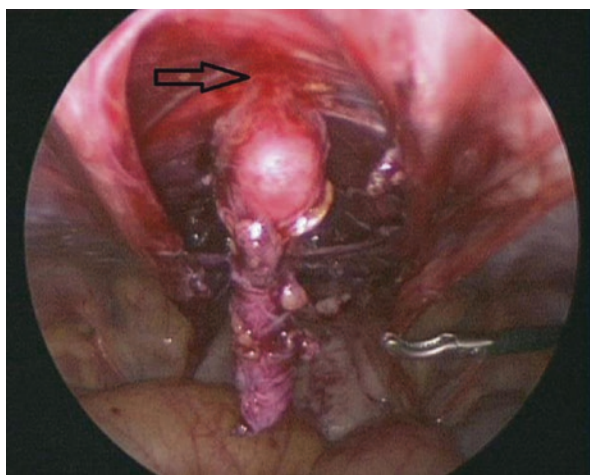


Fig. 1.17 Urachal cyst mobilized by the laparoscopic approach. *Arrow* demonstrates sinus communication through abdominal wall and skin 3 cm inferior to the umbilicus (Image used with permission from Minkes et al. [28])

Umbilical hernias are approached through an incision in the infraumbilical or supraumbilical crease. Dissection is carried down to the level of the fascia. The hernia sac is identified at its base and encircled. Contents from the hernia sac should be reduced. The sac is then disconnected from its attachment with the umbilicus.

Care is taken to avoid injury to contents within the hernia sac and to the umbilical skin. Opening the anterior surface of the sac may help to avoid injury to the bowel. The sac is resected down to the level of the fascia. The umbilical fascia is closed with interrupted or running absorbable suture. For large or recurrent hernias,

Fig. 1.18 External mobilization of urachal sinus through abdominal wall incision 3 cm inferior to umbilicus. Patient presented with recurrent drainage and infection from sinus. The internal portion was mobilized laparoscopically (Image used with permission from Minkes et al. [28])

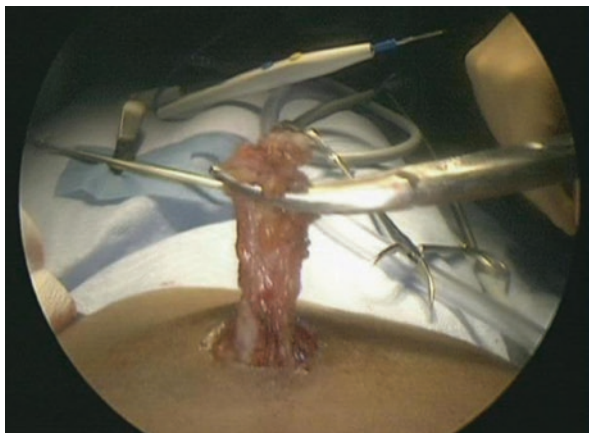


Fig. 1.19 Neoumbilicus following umbilicoplasty (Image used with permission from with permission from Minkes et al. [28])



nonabsorbable suture is used by many surgeons. The wound should be inspected and meticulous hemostasis achieved.

The umbilicus is tacked down to the fascia with an interrupted suture. The subcutaneous tissue is reapproximated with a few interrupted sutures, and the skin is closed with a subcuticular suture. Bupivacaine can be injected for postoperative analgesia. The skin is cleaned, and Steri-Strips are applied. A pressure dressing may be used for large hernias to prevent a postoperative hematoma or seroma.

Routine umbilicoplasty (Fig. 1.19), the removal of excessive umbilical skin, is generally not needed. In most cases, a redundant umbilicus appears more natural than a neoumbilicus. Several techniques can be used for extremely protuberant umbilical hernias (Figs. 1.20, 1.21, 1.22, and 1.23) [26, 27]. A simple technique is to invert the umbilicus over a finger so that the undersurface is exposed. The skin is

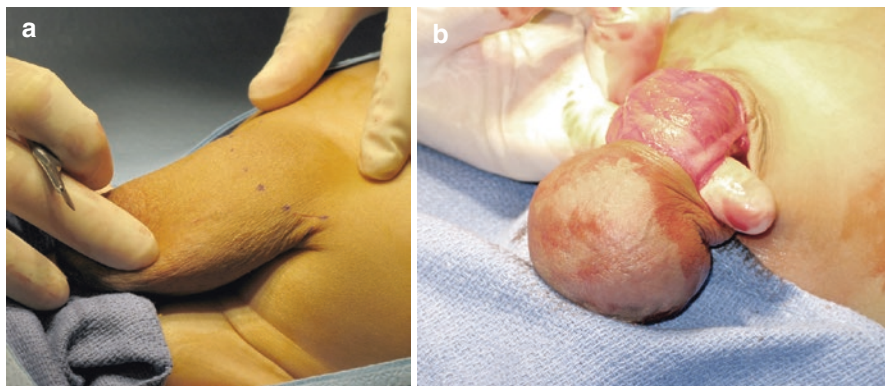


Fig. 1.20 Large proboscis umbilical hernia after healed ulcer of skin. (a) Incision line has been marked. (b) Hernia sac with nonreducible bowel has been encircled

Fig. 1.21 Small intestine adherent to hernia sac. The bowel was dissected free and safely returned to the peritoneal cavity

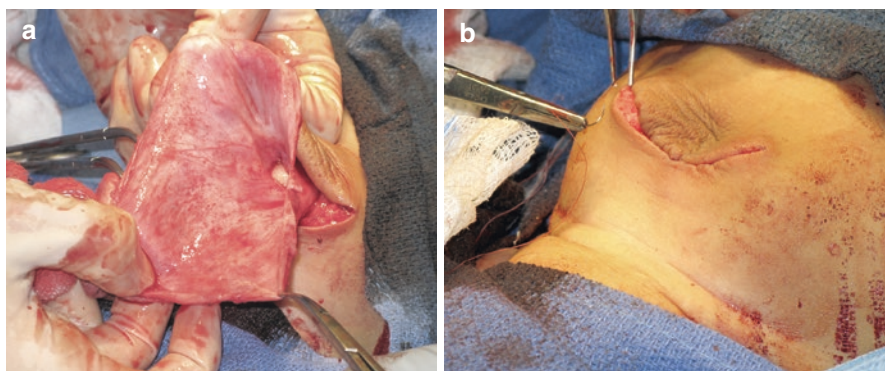
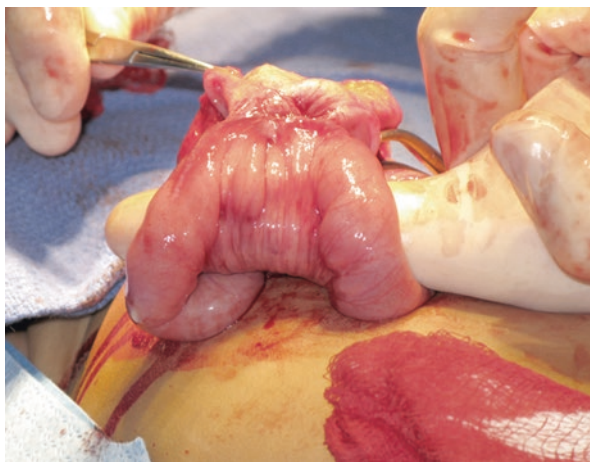


Fig. 1.22 (a) Hernia sac after bowel was reduced. (b) The sac was resected, and the fascia was closed with interrupted suture. Lower image shows initial closure

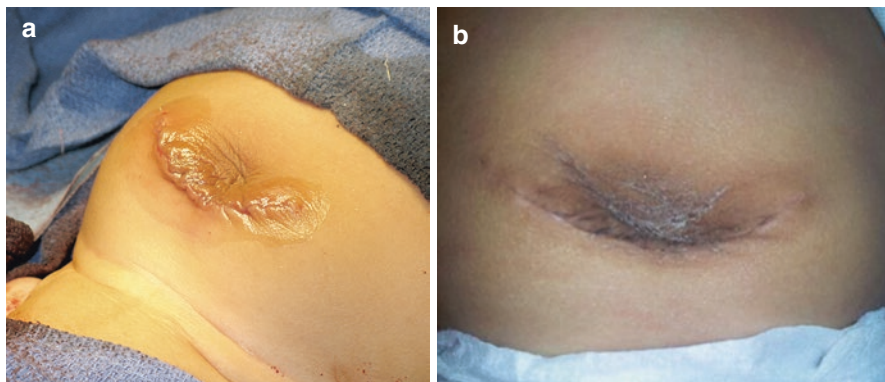


Fig. 1.23 (a) Completed closure. (b) One month after surgery

then incised circumferentially so that a 1–2-cm rim of umbilicus remains. The umbilical skin defect is reapproximated from within the umbilicus and tacked down to the fascia.

1.9 Conclusions

During development, the embryonic disk is in contact with the yolk sac anteriorly. As the embryo grows and differential growth of tissues leads to the folding appearance of the embryo, the ventral attachment of the yolk sac narrows. The intra-coelomic portion of the yolk sac becomes the primitive alimentary canal and attaches to the extra-coelomic portion through the vitelline duct. The allantois buds from the hindgut and grows into the body stalk. The yolk stalk and the body stalk eventually fuse to become the umbilical cord.

As the abdominal wall forms, the umbilical ring is narrowed. The vitelline and umbilical vessels, vitelline duct, and allantois should be absent in the umbilicus at term. Residual tissue leads to remnants that require surgical intervention. During exploration for a sinus or fistula, all structures, including the round ligament, median, and medial umbilical ligaments, must be identified. Umbilical hernias are common, and many will close spontaneously.

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