



Stephen J. Fenton

The umbilical cord originates from a connecting stalk that attaches the early embryo to the developing placenta. During the fourth week of gestation, the embryo begins to fold, from the cranial and caudal regions as well as laterally. The coalescence of these fourfolds (cranial, caudal, lateral) transforms the flat trilaminar embryonic disk into a cylindrical C-shaped fetus with body cavities. As a result, the connecting stalk develops into the umbilical cord that arises from the central portion of the anterior embryonic abdominal wall, containing the umbilical arteries (normally two), umbilical veins (the right one involutes), and the allantois. The abdominal wall at the base of the umbilical cord is not closed but instead forms a ring. This opening in the linea alba serves as a passageway for the umbilical structures.

The allantois is a diverticulum from the caudal end of the yolk sac. It remains very small and is involved with early embryonic exchange of waste and bladder development. As the bladder enlarges, the allantois becomes the urachus, a tube between the umbilicus and the bladder. It typically obliterates but maintains a connection to the umbilicus seen as the median umbilical ligament. For a brief period of time, the omphalomesenteric (vitelline) duct, a connection between the yolk sac and the terminal ileum of the midgut, also traverses through the umbilical cord. This usually involutes between the fifth and seventh weeks of gestation. Additionally, the opening at the umbilical ring allows for the physiologic herniation of the midgut between the sixth and tenth gestational weeks allowing for the midgut to elongate prior to reducing back into the abdominal cavity around the tenth week.

It is relatively common for parents to bring their child to a pediatric surgeon because of a problem within the umbilicus. The disorders seen within the clinic are almost always a direct result of development and take on the form of a mass, drainage or infection.

Umbilical Mass

Umbilical Hernia

By far the most common umbilical mass referred for pediatric surgical evaluation is an umbilical hernia. The fascial opening at the umbilical ring should spontaneously close and incomplete closure results in a hernia with projection of the peritoneum through the ring creating a hernia sac. Umbilical hernias are most commonly found on initial examination of the newborn and most parents state that their child has had the hernia since birth. It becomes more apparent with increased abdominal pressure such as during crying, straining while stooling, and with increased activity. Most are able to be reduced with very little pressure and the fascial edges can be easily palpated deep to the skin. The amount of redundant skin does not correlate with the actual size of the hernia defect itself.

The great majority of umbilical hernias are asymptomatic. Even in what appears to be a large hernia in an infant will become much smaller over 6–12 months and will rarely cause a problem. Since most will close spontaneously, it is standard practice to wait until the child is 4–5 years of age before recommending surgical repair. The size of the fascial defect determines the likelihood of spontaneous closure and a hernia with a fascial defect larger than 1.5 cm is less likely to close without intervention (Fig. 83.1). Some parents and referring pediatricians need reassurance that delaying repair will cause no harm to the child. Occasionally, parents will describe episodes of incarceration requiring manual reduction, either by them or by a physician. These children should be considered for early repair. Although true incarceration is extraordinarily rare, parents should be informed of the signs and symptoms and recommended to seek immediate medical attention should they occur.

Umbilical hernia repair is a very safe and generally straightforward operation and can usually be completed quickly under general anesthesia. An infra-umbilical incision is preferred; however, some prefer a supra-umbilical

S. J. Fenton (✉)
University of Utah School of Medicine, Salt Lake City, UT, USA
e-mail: stephen.fenton@hsc.utah.edu



Fig. 83.1 Proboscis umbilical hernia. Large proboscis umbilical hernia (courtesy of Michael D. Rollins)

incision if the hernia is on the more superior side of the umbilicus or if repair of a second epigastric hernia just superior to the umbilicus will also be performed. The hernia sac should be dissected circumferentially and either carefully separated from the dermis or amputated, leaving a disk of sac on the undersurface of the umbilical skin. The fascia can then be closed transversely or longitudinally according to surgeon's preference. I prefer to place all sutures first being careful to visualize the needle tip and avoid injury to underlying bowel and then tie them in succession. Suture preference (monofilament or braided) and closure approach (interrupted or running) are a matter of surgeon's choice with favorable outcomes for all. The umbilicus is then reconstructed by tacking the base down to the fascia, at or just below the fascial repair. I like to do this at two points, ensuring both a good bite of fascia and umbilical dermis, taking care to not buttonhole the skin. The skin is closed, and the incision dressed according to the surgeon's desire.

In children who have a large amount of redundant overlying skin (proboscoid hernia) an umbilicoplasty might be required. There are a variety of techniques to recreate the umbilicus and one should utilize the technique with which they feel most comfortable. My preferred technique is to trim some of the excess skin. An absorbable braided stitch is then placed through the fascia in what will be the center of the umbilicus. It is then sewn intradermally around the edges of the wound in a running purse-string fashion, carefully tightening the suture along the way. Once around the entire wound, the suture is then placed back through the fascia next

to the first fascial bite and tied. This typically results in an eversion of the skin edges around the center of the neo-umbilicus.

Umbilical Granuloma

Separation of the dry umbilical cord stump should occur within the first week of life and is considered delayed when it persists for longer than 3 weeks. Delayed separation can be associated with an underlying immunodeficiency disorder, infection, or urachal abnormality and should prompt further evaluation. Once the cord has separated, there is generally a base of granulation tissue that will undergo epithelialization within several days. When new skin growth fails to occur, the result is a small mass of granulation tissue, often referred to as a granuloma. It is typically a soft, reddish-pink, friable mass up to 10 mm in size. Some can be sessile but are often pedunculated (Fig. 83.2). After a urachal or vitelline duct remnant has been ruled out by a detailed history (drainage of urine or succus?) and examination (complex, mucosa-like structure?) most granulomas can be treated with the application of silver nitrate. Care should be taken to apply the silver nitrate precisely on the granulation tissue and to protect the skin so as to avoid burning or staining of the surrounding healthy skin.

When seen by a pediatric surgeon, the child has usually already been treated several times with silver nitrate by the referring pediatrician. If after evaluation, the diagnosis

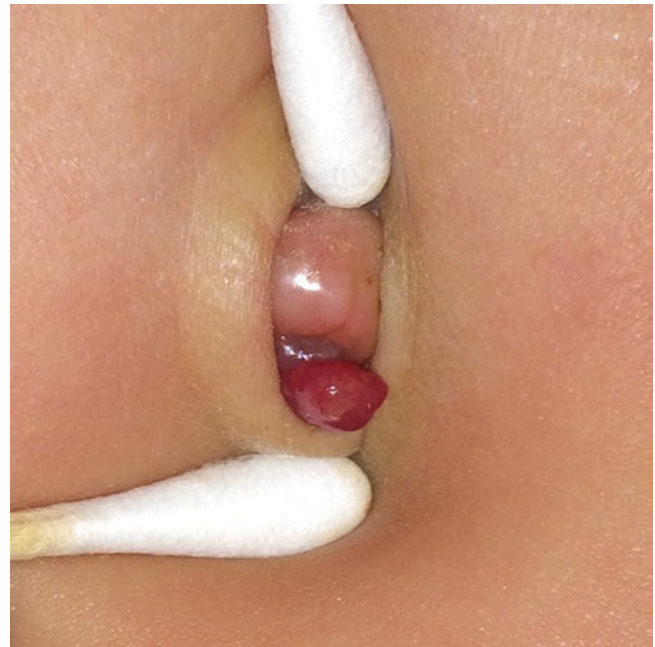


Fig. 83.2 Umbilical granuloma. A soft, pink, friable pedunculated mass typical of an umbilical granuloma

remains an umbilical granuloma, I usually remove it using a fine hemostat in the office followed by silver nitrate cauterization of the base and gentle pressure. The parents should be informed that this will not be painful to the infant and that some slight bleeding might occur but will resolve with a brief period of pressure. If it is truly pedunculated, suture ligation is described, however, I have found it quite challenging to place a suture around the base, down in a small hole, with a moving uncooperative infant. I have yet to have a child have significant hemorrhage from just pulling off the granuloma with a hemostat and applying pressure.

Umbilical Polyp

An umbilical polyp is a retained remnant of the omphalomesenteric duct or urachus and is comprised of either intestinal or urogenital mucosa, respectively. The polyp is distinguishable from a granuloma by its appearance, which is shiny, red, and smooth. They are not responsive to cauterization and require surgical excision along with the underlying remnant.

Umbilical Drainage

Most drainage from the umbilicus that occurs in the newborn results from healing of granulation tissue following separation of the umbilical cord stump and is rather short-lived. Persistence of drainage, especially after the newborn period, can be caused by a patent omphalomesenteric duct or patent urachal remnant. It is important to identify these problems and not treat them like simple granulation tissue.

Omphalomesenteric Duct Remnant

Incomplete involution of the omphalomesenteric (vitelline) duct can lead to a variety of disorders (umbilical polyp, omphalomesenteric duct cyst, patent omphalomesenteric duct, Meckel's diverticulum, omphalomesenteric fibrous band). Complete patency of the omphalomesenteric duct results in communication between the terminal ileum and the umbilicus. After separation of the umbilical cord, a stoma-like lesion appears with intermittent drainage of succus entericus. Further evaluation with ultrasound imaging is reasonable to look for a connection with underlying bowel. Surgical excision of the duct with closure of the enteric defect and repair of the umbilical hernia is required. The fistula tract should be traced back to the terminal ileum and

divided flush with the bowel using either a stapling device or sutured closure. Rarely, a segmental bowel resection is required.

Urachal Remnant

Failure of the urachus to completely obliterate will result in a variety of urachal anomalies (umbilical polyp, urachal cyst, patent urachus, bladder diverticulum). A patent urachus results in communication between the bladder and the umbilicus and is three times more likely to occur in boys. Drainage of urine is the most common symptom but recurrent urinary tract infections can also be seen. The persistent drainage of urine can result in an erythematous rash of the surrounding skin that is sometimes mistaken for omphalitis. US imaging is helpful in demonstrating the urachal remnant and should be obtained whenever a urachal anomaly is suspected. A patent urachus can result from distal urinary obstruction (posterior urethral valves) especially when discovered in the newborn period. Therefore, a voiding cystourethrogram (VCUG) should be obtained prior to surgical repair. Complete surgical excision including a cuff of bladder is recommended for repair and to prevent the risk of future urachal adenocarcinoma (Fig. 83.3). Most close the bladder in two layers with absorbable suture, which can usually be done completely extraperitoneally through an infra-umbilical incision. Bladder spasms sometimes occur following repair and can be treated with oxybutynin.

Laparoscopic resection of a urachal remnant is well described. I typically use this approach for older and larger sized children or when the diagnosis is in question since most can be done through a transumbilical approach in a relatively scarless manner. Three access sites are required, one in the epigastrium for visualization and two additional incisions on either side of the umbilicus for working instruments. Use of the camera through the epigastric trocar will allow for complete visualization of the abdominal side of the umbilicus as well as the entire urachal remnant to the bladder. Complete mobilization of the remnant should be performed using electrocautery and ligation as close to the bladder as possible using an endoloop. The remnant is then divided and removed from the abdomen (Fig. 83.4). This approach remains somewhat controversial, as the dictum has been to include a cuff of bladder along with the remnant to prevent future development of adenocarcinoma. However, the true risk of developing a malignancy in a urachal remnant is unknown. Given that spontaneous resolution of these remnants has been reported, a laparoscopic resection is a reasonable approach.

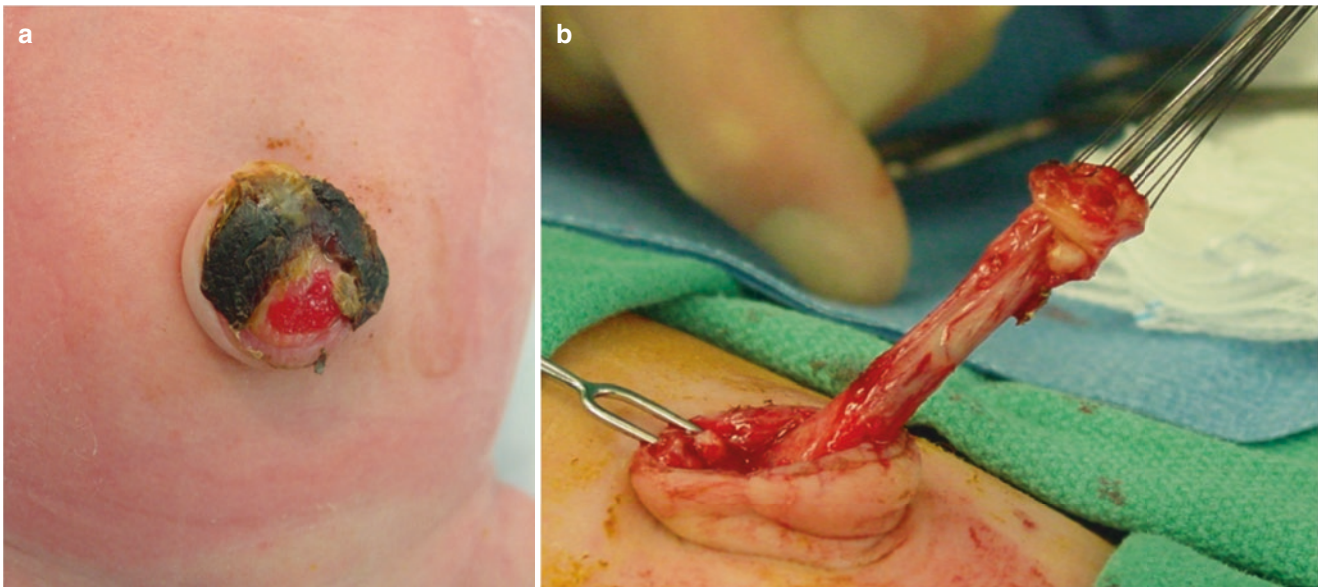


Fig. 83.3 Urachal remnant. (a) Umbilical opening of a patent urachus. (b) Removal of the urachal remnant through an infra-umbilical incision (courtesy of Earl C. Downey)

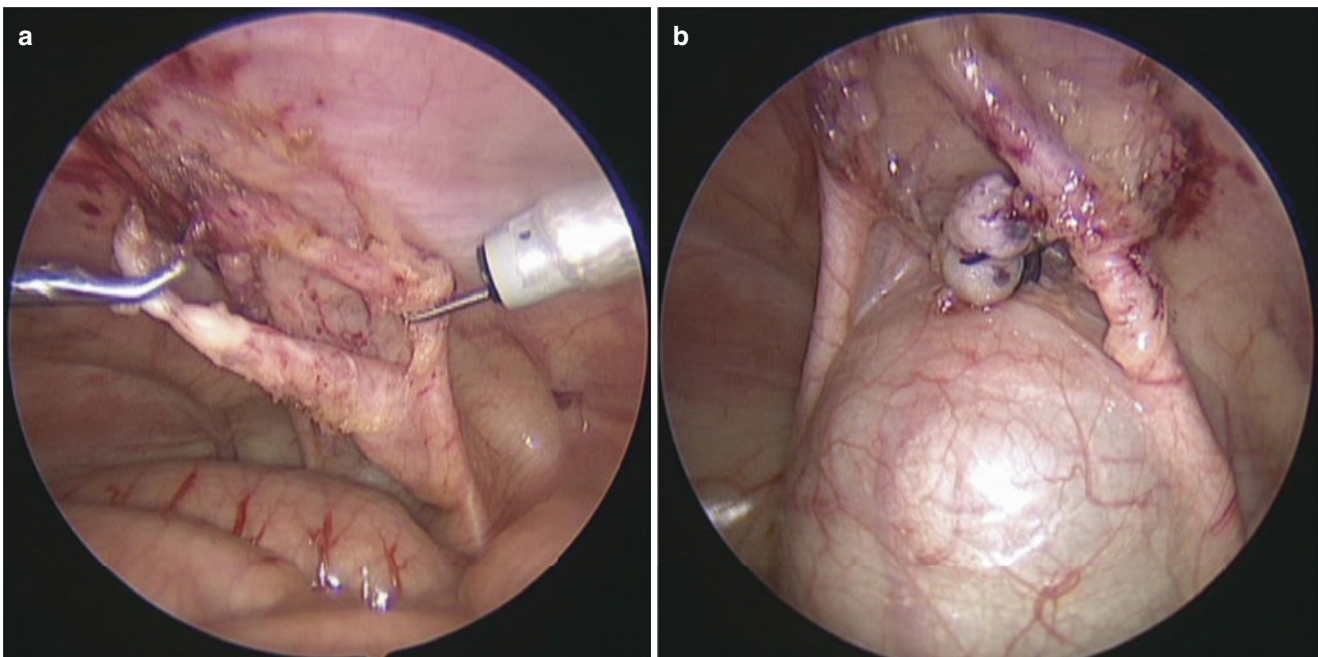


Fig. 83.4 Laparoscopic resection of urachal remnant. (a) Laparoscopic mobilization of the urachal remnant. (b) Ligation of the remnant along with a small cuff of bladder using an absorbable endoloop (courtesy of Michael D. Rollins)

Umbilical Infection

Umbilical infections are rare but can be very serious, even life-threatening. In a newborn, they should be evaluated immediately by a pediatric surgeon as they can be the nidus for a necrotizing process.

Infected Urachal Cyst

Urachal cysts are subfascial and located in the preperitoneal space between the umbilicus and bladder. They often remain asymptomatic and undiagnosed until infected and present as a tender, erythematous mass. Parents will often deny ever

having seen umbilical drainage and the child will otherwise be healthy. A US is adequate for diagnosis and a CT or MRI should only be obtained if the US is unclear. Initial treatment should be antibiotic therapy with incision and drainage as indicated. Complete surgical excision should be undertaken at least 4–6 weeks later to allow for adequate resolution of the inflammation surrounding the cyst. Like any true cyst, it is important to remove the cyst completely to prevent recurrence. Some literature suggests that following treatment, formal excision might not be required as the majority will resolve spontaneously. I would, however, recommend follow-up imaging for confirmation should this be the primary course of treatment.

Infected Omphalomesenteric Cyst

Infected omphalomesenteric cysts are much less common than urachal cysts. Like Meckel's diverticulitis, the diagnosis is rarely made except at exploratory laparotomy or laparoscopy. They are more likely to be intra-abdominal and present more like appendicitis or another infectious intra-abdominal processes. If the cyst is just posterior to the umbilicus, it can present like a urachal cyst, in which case antibiotics with or without incision and drainage is the best initial strategy, followed by delayed excision after complete resolution of the inflammatory process. Formal resection might require a laparotomy or periumbilical incision and attachments to the ileum need to be followed and completely excised.

Omphalitis

Omphalitis is an infection of the umbilicus and surrounding tissues and is primarily a disease of the newborn. It occurs in less than 1% of neonates in developed countries due to infection control techniques. It should be suspected when there is purulent drainage from the umbilical cord stump with surrounding erythema, induration, and tenderness. Risk factors include low birth weight, prolonged labor, prolonged rupture of membranes, and umbilical catheterization. The most common complication is sepsis, and the most feared complication is necrotizing fasciitis. Portal vein thrombosis and liver abscesses can also occur.

Most often the infection is polymicrobial; however, *Streptococcus*, *Staphylococcus*, and *Escherichia coli* are common pathogens. Intravenous administration of broad-spectrum antibiotics is the primary treatment along with close and frequent observation. Mortality rates are reported to be as high as 15%. In the rare instance that this infection progresses to necrotizing fasciitis, prompt and aggressive

surgical debridement of all necrotic tissue should be undertaken. Planned re-exploration should be performed as the necrosis is often progressive and several procedures are frequently required resulting in devastating wounds of the abdominal wall. This type of infection spreads rapidly and is associated with a mortality rate as high as 85%.

Editor's Comments

New pediatric surgical attendings might be surprised by the number and variety of umbilical disorders that present daily in outpatient practice. Umbilical granulomas can usually be treated with silver nitrate, but the pediatrician will have tried two or three times already. Pedunculated granulomas can be ligated after application of alcohol using a 000 braided absorbable suture. The granuloma usually falls off within 2 weeks. Surgical excision should rarely be necessary and sometimes means you are dealing with a duct remnant.

Umbilical hernias are very common and usually close spontaneously by the age of 4. Until then, the skin might gradually become more and more stretched out, causing alarm for parents who believe the hernia is enlarging when in fact the actual defect is getting smaller. Parents should be reassured that umbilical hernias rarely cause pain, are safe to observe and never actually enlarge. It might be helpful to have the parents feel the size of the fascial defect. It is reasonable to offer repair of large defects in children as young as 3, but repair under 2 is associated with a high rate of recurrence. Mesh reinforcement is unnecessary and the long-term risks are unknown. Transverse closure usually results in less tension and interrupted absorbable sutures, with figures-of-8 at the apices, provide a reliable closure. Tacking the central scar down to a point *inferior* to the fascial suture line results in a more natural appearance of the umbilicus. The pressure dressing is a pointless remnant of a bygone era and should be abandoned in favor of a precise hemostatic closure and cyanoacrylate glue.

Umbilicoplasty is reserved for those with absurdly excessive skin and parents should be warned that a small amount of redundant (not protuberant) skin is to be expected and will result in a better cosmetic result as the child grows into adulthood. One must decide whether umbilicoplasty is needed beforehand so that the proper incision can be made—infra-umbilical or through the central scar. Regardless of the technique, it is important to leave at least some excess skin. A simple purse-string is fine, but an alternative is to excise three 1–1.5 cm inverted triangles of skin equally spaced around the edge of the skin defect before placing the purse string.

Initial management of an infected urachal cyst includes antibiotics with incision and drainage if a sizable abscess is present. Delayed resection of the remnant in at least 4–6 weeks is much less morbid than attempting to resect a large, acutely inflamed cyst. Extraperitoneal resection of the remnant can usually be performed easily through a small infra-umbilical incision. Nevertheless, some prefer to do it laparoscopically or robotically, in which case the remnant is ligated rather than excised. Whether this is safe is unknown—time will tell if the historical directive that a cuff of bladder be excised is valid or yet another example of groundless dogma.

Further Reading

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