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Care of the umbilicus and management of umbilical disorders

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INTRODUCTION

At birth, the umbilical cord, which provided vascular flow between the fetus and placenta, is clamped and cut. Within the first week of life, the remnant umbilical cord stump separates from the neonate, creating the umbilicus (commonly referred to as the navel).

Infection, hernia, granulomas, and congenital anomalies can occur in the umbilicus and are more commonly seen in infancy.

The care of the umbilical cord after delivery and clinical problems associated with the umbilicus will be reviewed here. Prenatal diagnosis and management of umbilical cord abnormalities are discussed separately. (See "Umbilical cord abnormalities: Prenatal diagnosis and management".)

ANATOMY

Umbilical cord — The umbilical cord contains two arteries and one vein, which are surrounded and supported by gelatinous tissue called Wharton's jelly.

Thickness — At birth, the average diameter and circumference of the umbilical cord in a normal term infant is 1.5 and 3.6 cm, respectively [1,2]. Both thick and thin cords are associated with increased risk of significant pathology. (See 'Newborn examination' below.)

Length — The umbilical cord lengthens through gestation from a mean of 32 cm at 20 weeks gestation to 60 cm at term [3]. Within each gestational age group, the length is highly variable. As an example, in term infants born between 40 and 41 weeks gestation, the umbilical length varies from 35 to 80 cm.

Although the cord length has no effect on fetal blood flow, both long and short cords are associated with adverse effects (see "Gross examination of the placenta", section on 'Umbilical cord'):

- With longer umbilical cords, there is an increased risk of cord knots, nuchal cords (umbilical cord coiled around the neck of the fetus at the time of birth), cord prolapse, thrombi, and fetal demise.
- Short umbilical cords (<35 cm) have been associated with placental abruption and developmental abnormalities [4,5]. Short cords with no coiling are associated with poor fetal growth and decreased activity, which can occur in fetuses with severe neural and musculoskeletal abnormalities.

Coiling — Because the umbilical vessels are longer than the cord itself, twisting and bending of the vessels within the cord is common. Coiling of the umbilical cord is thought to protect the blood supply from mechanical disruption.

Although the mechanism of coiling is unknown, both excessive hypo- and hypercoiling are associated with adverse perinatal outcomes. This was illustrated in a review of 885 infants [6]. The following findings were noted:

- Infants with umbilical cord index (number of coils divided by the length of cord in cm) below the 10th percentile were more likely to be premature, have a trisomic genetic abnormality, or have a single umbilical artery (SUA). In addition, there was an increased risk of fetal death.
- Infants with umbilical cord index above the 90th percentile were more likely to have perinatal asphyxia, umbilical arterial pH below 7.05, trisomic genetic abnormality, SUA, or to be small for gestational age (SGA).

Umbilicus — The umbilicus is composed of three distinct anatomic areas ($\sqrt[M]{\text{figure 1}}$):

- Mamelon Area of central depression
- Cicatrix Dense scar, which marks the intersection of fetal intra- and extra-embryonic mesoderm
- Cushion Slightly raised margin around the mamelon and cicatrix

Variation of these three features result in more than 60 reported normal anatomical variants [7].

The umbilicus is located in the midline at the level of the iliac crest [8]. The umbilicus is lower than usual in patients with achondroplasia, displaced superiorly during pregnancy, and inferiorly by ascites (Tanyol's sign) [8].

Abnormal umbilical appearance or location may be a finding in a genetic disorder such as those illustrated by the following examples [9]:

- In Robinow's syndrome, a rare inherited disorder of short stature and macrocephaly with frontal bossing, the umbilicus is high, flat, and poorly epithelialized.
- In Axenfeld-Rieger syndrome, a rare genetic disorder that includes malformations of the anterior chamber of the eye and teeth, the umbilicus is broad and prominent with a large stalk and redundant skin. (See "Overview of glaucoma in infants and children", section on 'Anterior segment dysgenesis'.)
- In Aarskog-Scott syndrome, a disorder of multiple limb and genital abnormalities with short stature, the umbilicus can be either flat with radiating branches of the cicatrix or a deep longitudinally oriented ovoid depression.

EMBRYOLOGY

An appreciation of the embryology of the umbilical cord helps in understanding the pathogenesis of congenital umbilical anomalies.

In the fourth week of embryogenesis, the flat trilaminar embryonic disc folds and becomes a cylindrical C-shaped embryo, which narrows the opening of the yolk sac to the embryo (figure 2). This narrowed opening contains the umbilical vessels,

the urachus, and the omphalomesenteric duct. The omphalomesenteric duct connects the yolk sac to the developing gut. At the same time, the allantois, a diverticulum of the caudal hindgut, forms and becomes the urachus. The urachus connects the developing genitourinary tract (bladder) to the umbilicus.

In normal development, both the omphalomesenteric duct and urachus involute. After involution, no remnant of the omphalomesenteric duct persists, but, in contrast, a persistent remnant of the urachus can be seen in some individuals extending from the bladder to the umbilicus in the preperitoneal midline.

UMBILICAL CORD AT BIRTH

Newborn examination — After delivery, the clamped umbilical cord of the newborn infant is inspected for its general appearance, including the presence of discharge, an unusually thick cord, or a single umbilical artery (SUA).

Thick cord — A thick cord may contain bowel, embryonic remnants (omphalomesenteric duct or urachal elements), or a vascular anomaly [10-12]. Thus, evaluation of infants with a thick cord should include a careful examination of the transected cord. If there is suspicion of urachal or omphalomesenteric duct remnants, an ultrasound should be ordered [13]. Thin cords less than 1 cm in circumference are associated with infants who are postdates or small for gestational age (SGA).

Single umbilical artery — An SUA is present in 0.2 to 0.6 percent of live births, occurring more frequently in SGA and premature infants as well as twins [14-17]. Multiple studies have shown that 20 to 30 percent of neonates with SUA had major structural anomalies, often involving multiple organs [14-17]. The most commonly affected organs are the heart, gastrointestinal tract, and central nervous system. Meta-analysis based on reviews of the literature suggest that SUA is associated with lower birth weight, oligo- and polyhydramnios, admission to the neonatal intensive care unit, and increased mortality [18,19]. (See "Single umbilical artery", section on 'Assessment for associated abnormalities'.)

However, in the majority of cases (70 to 80 percent), SUA is an isolated finding. Based on available data, we do **not** perform further imaging for healthy term infants with an isolated SUA, as there is a low likelihood of renal or urologic abnormalities, illustrated by the following:

- A meta-analysis of seven studies that included 204 infants who were screened for renal malformations either by ultrasonography or intravenous pyelography reported a 16 percent detection rate of a renal anomaly [16]. However, the abnormality was persistent and considered clinically significant in only one-half (8 percent). Vesicoureteral reflux (VUR) of grade 2 or higher was the major renal finding and was found in 3 percent of the total study population.
- A subsequent prospective study of infants born over six years at a single English institution identified 137 of 33,057 liveborn infants (4.1/1000) with SUA [20]. Eight infants had major congenital anomalies (eg, complex heart disease, cloaca, anorectal, abnormal karyotypes, cystic adenomatoid malformation, and multiple anomalies), and three had minor anomalies (eg, preauricular skin tag, preaxial polydactyly, and syndactyly). Infants with SUA had a lower gestational age and were more likely to be SGA than control infants with two umbilical arteries. In infants with isolated SUA, 117 of the 122 infants who underwent renal ultrasonography had normal scans. Two infants had significant renal abnormalities: a hypodysplastic kidney, which was already detected prenatally, and a single kidney. Renal anomalies were more common among infants with isolated SUA compared with the control cohort (4.1 versus 0.9 percent). There was no statistical difference in the risk of significant clinical renal abnormalities defined by the authors (1.6 versus 0.4 percent).
- Similar results were seen in a study of 65 infants with SUA from the Netherlands that reported a clinically insignificant renal abnormality in 1 of 57 cases of isolated SUA [21].

Cord care — The risk of subsequent umbilical infection is dependent upon the quality of care at delivery and postnatally [22,23]. In all settings, the cord should be cut with a sterile blade or scissors, preferably using sterile gloves, to prevent bacterial contamination, which can lead to omphalitis or neonatal tetanus [22].

- Low medical resources In the setting of low medical resources where sterile equipment is not available, there is an increased risk of omphalitis. In this case, the use of <u>chlorhexidine</u> (an antiseptic topical agent) is a beneficial and inexpensive option that reduces neonatal morbidity and mortality. In a meta-analysis of 12 community-based trials, the use of chlorhexidine reduced all-cause mortality (relative risk [RR] 0.81, 95% CI 0.71-0.92) and risk of omphalitis/infections (RR 0.48, 95% CI 0.4-0.57) compared with dry cord care [24].
- Hospital delivery In the hospital setting, where aseptic care is routine in the clamping and cutting of the umbilical cord, dry cord care is recommended. Additional antiseptic topical care is not necessary, as it does not significantly reduce the

already low risk of omphalitis in settings that use routine aseptic care to clamp and cut the umbilical cord, and is associated with rare complications ($\boxed{1}$ table 1) [24-26].

Inappropriate cord care also can increase umbilical infections. As an example, neonatal tetanus has been reported as a complication of inappropriate application of cow dung or bentonite clay [27]. (See "Tetanus", section on 'Neonatal tetanus'.)

Cord separation — Cord separation normally occurs one week after birth, although there can be delays up to three weeks [28]. This was illustrated in a subsequent study of the previously mentioned Nepalese community-based trial that demonstrated a mean umbilical separation time of 4.2, 4.3, and 5.3 days in infants who received dry care, soap/water, or <u>chlorhexidine</u> umbilical cord care, respectively [28]. In addition, multiple cleansings with chlorhexidine increased the time for cord separation to a mean duration of 7.5 days [29]. Longer time periods to separation are associated with other antiseptic topical agents used in cord care including salicylate sugar powder (mean 5.6 days) [30], 70 percent alcohol (mean 12 to 16.9 days) [30,31], and triple dye (range three to eight weeks) [32].

Umbilical cord separation is initiated by thrombosis and contraction of the umbilical vessels followed by phagocyte-mediated tissue breakdown and epithelialization of the cord stump. The umbilical stump is colonized by bacteria derived from the maternal genital tract or environment after delivery, and in some cases these organisms can cause umbilical infections. (See 'Omphalitis' below.)

Delayed separation — Delayed cord separation does not have a specific definition, primarily because of the variation in normal cord separation. In general, any cord that persists after three weeks probably represents delayed cord separation. Delayed cord separation can be associated with underlying immunodeficiency, infection, or urachal abnormality [33]. Neutrophil function should be evaluated in infants with delayed cord separation and signs of umbilical infection because infants with leukocyte adhesion defects often present with these findings. (See "Leukocyte-adhesion deficiency", section on 'LAD I'.)

Although there are no data on the care of cords that fail to separate in the normal time frame of three weeks, these cords usually will separate without intervention eventually. Rubbing alcohol should not be applied to the cord as it may kill bacteria that assist in cord drying and separation. Drying of the cord may be helped by keeping the diaper folded below the cord, thereby exposing the cord to air. In some instances, after the cord has desiccated, it may be removed by a health care provider using a scissor or scalpel by dividing the desiccated tissue just distal to the normal skin.

OMPHALOMESENTERIC DUCT ANOMALIES

Partial or complete failure of involution of the omphalomesenteric duct can lead to a spectrum of anomalies in the newborn infant due to varying degrees and location of duct patency (figure 3).

- Complete patency results in the omphalomesenteric duct directly connecting the umbilicus to the terminal ileum. This can lead to drainage from the umbilicus. These infants will often appear to have a "stoma" in the umbilicus after cord separation.
- A persistent omphalomesenteric duct at the umbilicus with no intestinal connection results in an umbilical polyp. (See 'Umbilical polyp' below.)
- Persistent tissue at the ileum, with no connection to the umbilicus results in Meckel's diverticulum. (See "Meckel's diverticulum".)
- Patent mid-duct with closure at both the umbilical and ileal ends of the omphalomesenteric duct results in an omphalomesenteric duct cyst. Because the two ends are fixed, this can lead to small bowel obstruction if loops of bowel twist around the cyst.
- Persistent fibrous cord between the umbilicus and the ileum, which can lead to small bowel obstruction.

In a case series of 217 children with omphalomesenteric duct anomalies, 85 (39 percent) were symptomatic [34]. The most common symptoms were rectal bleeding due to Meckel's diverticulum and intestinal obstruction due to fibrous bands; abdominal pain and bilious umbilical drainage were much less common [34]. Among the 132 asymptomatic patients, a Meckel's diverticulum was incidentally found at laparotomy.

Because more than one omphalomesenteric duct anomaly can be present, radiologic evaluation should be performed in patients with an omphalomesenteric duct anomaly. This should include ultrasonography and possibly a Meckel scan with 99m technetium pertechnetate, which has an affinity for gastric mucosa. (See "Lower gastrointestinal bleeding in children: Causes and diagnostic approach", section on 'Meckel's diverticulum'.)

Surgical excision is performed for all symptomatic omphalomesenteric duct remnants, or remnants such as fibrous bands or cysts that place the patient at risk for bowel obstruction [35]. In our center, surgical resection is suggested for children with either symptomatic or incidentally found, asymptomatic Meckel's diverticulum because of the increased risk of complications over a lifetime [36]. In adults with an asymptomatic Meckel's diverticulum, the decision to resect is based on the relative risk of leaving the diverticulum in place versus resection [36]. (See "Meckel's diverticulum", section on 'Treatment approach'.)

UMBILICAL HERNIA

Umbilical hernia is due to a persistent opening of the umbilical ring, which normally spontaneously closes.

Natural history of umbilical ring closure — The fascial opening (umbilical ring) exists to allow passage of the umbilical vessels from the mother into the fetus. After birth, this fascial opening closes spontaneously with continued growth of the rectus abdominis muscles toward one another. Ultimately, complete closure occurs with fusion of the peritoneal and fascial layers within a small fibrous area of the umbilicus. Closure of the umbilical ring is complete in most children by five years of age.

Although closure is complete in most children by five years of age, this is not true for all children, as closure continues well into the teenage years for some children [37,38]. This was illustrated in a cross-sectional study of 4052 individuals that noted the following rates of umbilical closure of 665 Black children between 4 and 11 years of age based upon age [37].

- 4 to 5 years of age (n = 51) 14 percent
- 6 to 7 years of age (n = 142) 4 percent
- 8 to 9 years of age (n = 221) 3 percent
- 10 to 11 years of age (n = 251) 2 percent

Spontaneous closure is less likely to occur in patients who have a fascial opening that is greater than 1.5 cm, a significant amount of protruding skin, are older, or have an underlying predisposing condition [39,40]. Umbilical hernias due to failure of spontaneous closure are frequently seen in patients with Ehlers-Danlos [41], Beckwith-Wiedemann syndrome [40], Down syndrome, mucopolysaccharidoses [42], hypothyroidism [43], or trisomy 18. Increased intraabdominal pressure from ascites or peritoneal dialysis also can prevent closure of the umbilical ring [44].

Clinical findings — Although the vast majority of pediatric patients with umbilical hernias are asymptomatic, in rare cases the hernia can interfere with feeding, especially in young infants with hernias that contain bowel. In children, umbilical hernias rarely become incarcerated (inability to be reduced by manipulation) [45] or strangulated (vascular compromise of the contents of an incarcerated hernia) [46] or, even more rarely, rupture.

Umbilical hernias are detected during the newborn abdominal examination, particularly when there is increased intraabdominal pressure from crying. Umbilical hernias are easily reduced even if they are quite large, and the borders of the fascial defects can be palpated through the skin. The fascial defect, not the degree of protrusion, is most indicative of whether spontaneous closure will occur. It is important to differentiate umbilical hernias from supraumbilical hernias, which will not close spontaneously [47].

Management — Because the natural course of the umbilical ring is eventual closure, most umbilical hernias will spontaneously resolve [48]. In general, asymptomatic children with an umbilical ring that continues to decrease can be observed, regardless of their age. Surgery before age four years is not generally recommended, since the risk of recurrence is higher in children who undergo umbilical hernia repair before age four years, and surgery results in higher rates of posthospitalization and emergency department visits (条 algorithm 1) [49,50].

As a result, surgical intervention is required only in a minority of patients. The following are indications for surgical repair:

- Incarcerated or strangulated hernia is an absolute indication for surgical repair at any time that it is observed.
- For children <4 years of age, indications for surgery are for children in whom it is unlikely that the hernia will spontaneously close:
 - Large, proboscoid (trunk-like) hernias (picture 1) without any decrease in the size of the umbilical ring defect over the first two years of life.
 - Hernias associated with genetic and syndromic conditions, including children with Ehlers-Danlos syndrome [41], Beckwith-Wiedemann syndrome [40], Down syndrome, mucopolysaccharidoses [42], hypothyroidism [43], trisomy 18, or those with ascites or undergoing peritoneal dialysis [44].

- Behavioral concerns (eg, poor feeding, pulling on the hernia, bullying, and shame).
- For children ≥4 years of age, surgical repair may be considered for children with the following [51]:
 - Large defects (>1.5 cm/diameter)
 - No decrease in the size of the fascial defect during a year or so of observation
 - Behavioral concerns

Although there is folklore about "adhesive taping" an umbilical hernia to promote closure, this practice can lead to skin complications such as maceration and infection and should not be performed.

UMBILICAL MASSES

Infants — Umbilical masses in the neonate are most commonly umbilical granulomas, polyps, or ectopic tissue. These conditions are typically differentiated from one another by physical examination or failure to respond to initial treatment used to treat granulomas. However, if there is any question of whether an umbilical mass in a neonate is a polyp or granuloma, histopathologic evaluation of the lesion should be performed. If a polyp is diagnosed, further evaluation for associated embryologic anomalies (eg, Meckel's diverticulum) should be performed. (See 'Omphalomesenteric duct anomalies' above and 'Urachal anomalies' below.)

Umbilical granuloma — In neonates, umbilical granuloma is the most common cause of an umbilical mass. It is a soft, moist, pink, usually pedunculated, friable lesion of granulation tissue that varies in size from 3 to 10 mm in length (picture 2).

Umbilical granuloma forms in the first few weeks of life from excess tissue that persists at the base of the umbilicus after cord separation [52]. Granuloma formation is more likely to occur when there is inflammation of the umbilical cord, usually due to infection, which also delays cord separation.

Umbilical granuloma is most often detected after the cord has separated because of persistent drainage of serous or serosanguineous fluid, or moisture around the umbilicus.

In our center, we use topical 75% silver nitrate, usually applied by a wooden applicator with premounted silver nitrate. The lesion is treated once or twice a week for several weeks, but generally only a few applications are required for successful treatment. Caution should be exercised in applying silver nitrate because it can cause chemical burns or staining of the surrounding skin.

Limited data suggest that topical <u>clobetasol</u> propionate cream is successful in treating umbilical granulomas [53]. However, there are concerns regarding this intervention because of frequent adverse effects (skin atrophy and hypopigmentation), treatment failure, and the potential for systemic absorption. As a result, we would **not** routinely use topical clobetasol propionate until further studies have shown that it is effective and safe [54]. There is also a report of the successful use of common table salt as a single application that is covered with adhesive tape for 24 hours [55]. However, further confirmation of the safety and efficacy of this approach is needed.

In cases that fail to respond to topical <u>silver nitrate</u>, ligation of the granulation tissue can be performed in the office without discomfort [56]. Before ligation, the umbilicus should be carefully examined to rule out other causes of umbilical masses, such as umbilical polyp. Failure of the granuloma to resolve with ligation and/or silver nitrate should also increase the suspicion that the lesion is actually an umbilical polyp.

Umbilical polyp — Umbilical polyps are firm masses comprised of intestinal epithelium or uroepithelium, which are omphalomesenteric or urachal embryologic remnants [57,58]. Umbilical polyps, although they resemble a granuloma, are much less common, are often larger than granulomas, and do not respond to <u>silver nitrate</u> therapy. These lesions require surgical excision.

Ectopic tissue — Ectopic tissue in the umbilical cord is a very rare lesion that presents as a solid mass. Ectopic tissue can include pancreas, which most likely arises from growth of pluripotent cells derived from the omphalomesenteric duct [39,59], or liver, which results from mechanical entrapment as the umbilical ring closes [39,60]. Surgical excision is required for removal of ectopic tissue in the umbilicus.

Older children and adults — Umbilical masses in older children and adults are uncommon. Both benign and malignant umbilical tumors have been reported [39].

- Benign lesions include hamartomas, pyogenic granulomas, nevi, inclusion cysts, hemangiomas, dermatofibromas, neurofibromas, granular cell tumors, desmoid tumors, and lipomas.
- Primary malignancies are rare and include melanoma, urachal adenocarcinoma, squamous cell carcinoma, and basal cell carcinoma.
- Metastatic lesions have been reported from many primary sources (eg, stomach, pancreas, endometrium, ovary, cervix, colon, small bowel, gallbladder, prostate, lung, and breast). Metastatic lesions to the umbilicus are called "Sister Mary Joseph's node," named after the nun who worked with Dr. William Mayo, the surgeon who developed the surgical approach to umbilical hernia repair [61].

Other umbilical lesions include omphaliths, which results from lint accumulation in the umbilicus that become "ossified". These lesions are often hard and black, and thus can be misdiagnosed as a melanoma. In addition, keloid formation within or near the umbilicus can occur and mimic a primary tumor.

URACHAL ANOMALIES

The urachus normally involutes, resulting in a fibrous cord between the umbilicus and the bladder in the preperitoneal space. Disruption of this process can lead to a spectrum of rare anomalies ($\sqrt{\frac{1}{2}}$ figure 4):

- Patent urachus Complete patency results in a patent urachus with free communication between the bladder and the umbilicus. These children may present at birth with a giant umbilical cord, as the cord may fill with urine. Older children generally present with a persistently wet or draining umbilicus, and occasionally with a urinary tract infection.
- Umbilical polyp Persistent tissue at the umbilicus with no connection to the bladder results in an umbilical polyp. (See 'Umbilical polyp' above.)
- Bladder diverticulum Persistent tissue at the bladder with no connection to the umbilicus results in a bladder diverticulum. Bladder diverticulum can cause ureteral obstruction at the site of bladder insertion.

• Urachal cyst – Patency in the mid-duct with closure at both the umbilicus and the bladder results in a urachal cyst. The cyst can present as a mass, especially in older children and adults. The cyst may become infected with gram-positive skin flora or gram-negative Enterobacteriaceae and present with associated signs or symptoms of abdominal pain, erythema, or swelling, usually located below the umbilicus.

The true incidence of urachal anomalies is unknown. In a retrospective review of the radiologic database from a Canadian tertiary pediatric center, urachal anomalies were found incidentally in 1 percent of all patients who underwent abdominal imaging studies performed between 2000 and 2012 [62]. Diagnoses of the 721 patients (mean age at time of diagnosis was 6.2 years) with urachal anomalies included urachal remnants (89 percent), urachal cysts (9 percent), and patent urachus (1.5 percent).

In another case series of 103 patients from a single tertiary care center diagnosed at a median age of 1.8 months between 2006 and 2010, anomalies included urachal cyst (n = 38), patent urachus (n = 21), urachal diverticulum (n = 13), urachal sinus (n = 11), and nonspecific urachal remnant (n = 20) [63].

Clinical findings — Clinical manifestations in children include [62,63]:

- Umbilical drainage
- Abdominal pain
- Abnormal appearance of the umbilicus including a palpable mass
- Infection
- Asymptomatic presentation due to incidental finding on abdominal imaging study

Urachal anomalies may not be detected until adulthood, and patients may present with hematuria, pain, dysuria, or as incidentally during surgery for another disorder [64]. In adults undergoing resection of a urachal remnant, one-half of the pathologic specimens demonstrate evidence of adenocarcinoma. However, malignancy has not been reported in children with urachal anomalies.

Evaluation and management — A renal ultrasound and, if indicated, a voiding cystourethrogram (VCUG), should be performed in patients with urachal anomalies to confirm there are no associated genitourinary tract abnormalities.

In adults, if there is an opening at the umbilicus, a sinogram (radiocontrast injection into urachal opening) can be used to diagnose a patent urachus or urachal sinus. However, sinograms are difficult to perform in children and rarely add beneficial information to the other imaging modalities in pediatric patients.

Surgical excision has been the treatment of choice. However, if a small urachal remnant is found adjacent to the bladder, many surgeons now opt to follow these patients since the risk to benefit ratio for resection is not clear [62,63,65,66]. If a urachal anomaly is not resected, the caregiver and patient should be made aware of a potential future risk of malignancy and the need for lifelong screening.

OTHER ABNORMALITIES

Appendicoumbilical fistula — Appendicoumbilical fistula is rare and appears to occur in two settings: during development with entrapment of the appendix in the closing umbilical ring; and after a perforated appendicitis. The presenting manifestation is drainage of stool from the umbilicus.

Enteric fistula — Enteric fistula to the umbilicus has been reported in patients with Crohn disease or following surgery for tuberculous peritonitis [8,67]. (See "Clinical manifestations and complications of inflammatory bowel disease in children and adolescents".)

UMBILICAL INFECTION

Umbilical infections, which can progress to systemic infections, occur primarily in the newborn because of the following predisposing factors:

- Immediately following birth, the umbilicus becomes colonized with a diverse flora of microorganisms. Staphylococcal species and other Gram-positive cocci are present within hours, and enteric organisms follow shortly thereafter [68-70].
- Devitalized tissues of the umbilical cord stump provide an excellent growth medium for bacteria.

• The thrombosed blood vessels within the umbilical cord stump provide an entry for microorganisms into the bloodstream of the neonates, potentially leading to sepsis.

Omphalitis — Omphalitis is an infection of the umbilicus and/or surrounding tissues. It is predominantly a disease of the neonate and is characterized by purulent discharge from the umbilical cord stump with surrounding induration, erythema, and tenderness (picture 3A-B and picture 4).

Epidemiology and pathogenesis — Omphalitis is rare in developed countries, with a reported incidence of 0.7 percent [71]. However, the practice of intentional umbilical nonseverance (ie, the umbilical cord is not separated from the placenta after birth), also referred to as lotus birth, is associated with an increased risk of omphalitis [72]. In resource-limited settings, the estimated incidence is as high as 8 percent of infants born in hospitals and 22 percent of those born at home [73,74]. In the resource-limited setting, antiseptic topical care of the umbilicus stump reduces the risk of omphalitis and neonatal mortality. (See 'Cord care' above.)

Omphalitis is a polymicrobial infection. Historically, the predominant pathogens included *Staphylococcus aureus*, *Streptococcus* pyogenes, and Gram-negative bacteria such as Escherichia coli, Klebsiella pneumoniae, and Proteus mirabilis [75,76]. However, with the routine use of antistaphylococcal cord care regimens, Gram-negative infections of the umbilicus have increased [77,78]. In addition, anaerobic bacteria such as Bacteroides fragilis, Clostridium perfringens, and Clostridium tetani can contribute to umbilical infections, especially in infants born to mothers with chorioamnionitis [79]. In these infants, foul smelling umbilical drainage is a typical finding.

In the newborn, risk factors for the development of omphalitis include low birth weight, prolonged labor, prolonged rupture of membranes or maternal infection, nonsterile delivery, umbilical catheterization, and home birth [73,77,80]. Improper cord care also increases the risk of omphalitis, such as cultural application of cow dung. Abnormalities of the immune system, such as defects in leukocyte adhesion, neutrophil or natural killer lymphocyte function, and interferon production, can contribute to the development of omphalitis [80,81].

Clinical features — Mild discharge from the umbilical stump in the absence of inflammatory signs may be a normal occurrence, even when accompanied by some odor. Umbilical stump bleeding may occur with omphalitis because the infection delays thrombosis of the umbilical vessels.

Systemic signs, including lethargy, fever, irritability, and poor feeding are suggestive of more severe infection or complication. The most common complication of omphalitis is sepsis [77]. Other complications include septic umbilical arteritis, portal vein thrombosis, liver abscess, peritonitis, intestinal gangrene, small bowel evisceration, necrotizing fasciitis, and death [12,68].

Management — Whenever possible, cultures of the discharge should be obtained prior to the start of antibiotic therapy. Blood and cerebrospinal fluid cultures should also be obtained in infants with systemic signs (eg, fever) as they are more likely to be septic or develop meningitis.

Antibiotic treatment of omphalitis is required and is directed against Gram-positive and Gram-negative organisms [77]. In the neonate, parenterally administered antistaphylococcal penicillin and aminoglycoside agents are administered to decrease the risk of significant complications, such as sepsis and necrotizing fasciitis. In communities with a high prevalence of methicillin-resistant *S. aureus*, <u>vancomycin</u> should be used in place of an antistaphylococcal penicillin. <u>Clindamycin</u> or <u>metronidazole</u> also has been suggested in the treatment of infants with omphalitis for anaerobic coverage, especially those with foul smelling discharge or born to mothers with amnionitis [52,79]. We typically administer a 10-day course of intravenous antibiotics in neonatal patients, which can be modified dependent upon the patient's clinical response and whether complications develop.

Some clinicians treat infants with minimal symptoms with topical applications such as alcohol, <u>bacitracin</u>, or <u>mupirocin</u>. However, there is no evidence of efficacy of this practice or on the efficacy of the administration of oral antibiotics in these infants.

In older patients, similar antibiotic coverage can be administered orally, and is modified based on culture results and clinical improvement.

Mortality — Among infants with omphalitis, mortality rate is estimated between 7 and 15 percent [82]. Male sex, prematurity, septic delivery (including unplanned home delivery), and abnormal temperature are reported risk factors for poor prognosis in infants with omphalitis. However, data are limited and firm conclusions cannot be drawn regarding the role of these factors in mortality [82]. Although rare, the development of necrotizing fasciitis is associated with a higher mortality rate. However, necrotizing fasciitis is often difficult to diagnose without surgical exploration. For that reason, it is better to err on the side of surgical exploration in critically ill infants. (See 'Necrotizing fasciitis' below.)

Necrotizing fasciitis — Neonatal necrotizing fasciitis is a rare complication of omphalitis [52]. It is a polymicrobial infection of the skin, subcutaneous fat, and superficial and deep fascia. It is characterized by rapid spread of infection and inflammation, and signs of systemic toxicity. Infants presenting with fasciitis have a high incidence of bacteremia, shock, and death [52,77]. Reported mortality rates are as high as 60 to 85 percent [39,83]. Prompt aggressive surgery, broad-spectrum antibiotics, and supportive care are critical in the management of necrotizing fasciitis [39]. In addition to debridement of the involved abdominal wall, it is important to resect the umbilical vein, both umbilical arteries, and any urachal remnant that is present, as these may be involved in the necrotizing infection (even if they look normal). (See "Necrotizing soft tissue infections".)

Funisitis — Funisitis is inflammation of the umbilical cord that occurs with chorioamnionitis (in response to intraamniotic infection). Funisitis involves only the external surface of the cord and Wharton's jelly and does not involve the umbilical vessels. The inflammatory cells seen migrating through the fetal vessels in the cord are evidence of a fetal response to an external pathologic factor that is maternal in origin.

Funisitis does not involve the umbilical stump, which differentiates it from omphalitis. Necrotizing funisitis occurs with longstanding infection and is characterized by inflammatory debris and calcification of umbilical cord tissues. Infants with funisitis can be born healthy, but should be treated with broad-spectrum antibiotics (similar to the regimen used for omphalitis) for a minimum of seven days of therapy. (See "The placental pathology report".)

Infected umbilical piercing — Infected umbilical piercings are characterized by purulent discharge from the pierced skin with surrounding induration, erythema, and tenderness.

Local wound care and topical antibiotics usually are sufficient for resolution of local infection in older children, and it is usually not necessary to remove the device. When infection persists or occurs in a child younger than one year of age, oral antibiotics aimed at treating S. aureus and S. pyogenes are recommended. Neonates with an infected umbilicus require an evaluation for sepsis and intravenous antibiotics. (See 'Omphalitis' above and "Body piercing in adolescents and young adults", section on 'Localized infection'.)

INFORMATION FOR PATIENTS

UpToDate offers two types of patient education materials, "The Basics" and "Beyond the Basics." The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or e-mail these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on "patient info" and the keyword(s) of interest.)

• Basics topic (see "Patient education: Meckel's diverticulum (The Basics)")

SUMMARY AND RECOMMENDATIONS

- **Newborn examination** At delivery, the clamped umbilical cord is inspected to detect any alterations of the normal characteristics (thickness, length, and coiling) of the cord, which can be associated with an increased risk of significant pathology in the newborn infant. (See <u>'Umbilical cord'</u> above.)
 - Umbilical drainage In the newborn, the most common cause of umbilical drainage is umbilical granuloma. Other causes of umbilical drainage include omphalitis and umbilical anomalies, such as omphalomesenteric duct and urachal abnormalities. (See 'Newborn examination' above and 'Umbilical granuloma' above and 'Other abnormalities' above and 'Omphalitis' above.)
 - Single umbilical artery A single umbilical artery (SUA) is seen in 0.2 to 0.6 percent of live births, of whom 20 to 30 percent will have other major structural anomalies. Although infants with an isolated finding of SUA have an increased risk of occult renal anomalies, these are generally not clinically significant. As a result, in patients with isolated SUA, we suggest **not** to perform a screening renal ultrasonography (**Grade 2C**). (See <u>'Single umbilical artery'</u> above.)

- Newborn care In the neonate, umbilical cord care is directed toward reducing the risk of umbilical infection and is dependent upon the quality of care at delivery and postnatally. In resource-limited countries where there is an increased risk for omphalitis, we recommend antiseptic topical cord care (eg, <u>chlorhexidine</u>) rather than dry cord care (<u>Grade 1B</u>). (See <u>'Cord care'</u> above.)
 - Umbilical cord separation usually occurs within the first week of life. Delayed cord separation is associated with underlying immunodeficiency, infection, or urachal anomaly. (See 'Cord separation' above.)
- Umbilical hernias Umbilical hernias are commonly found in children, generally easily reducible, and usually resolve without intervention over the first five years of life. Surgical repair is always required in the rare patient with an incarcerated hernia. Indications for surgical repair are based on the probability of spontaneous closure and whether the child is symptomatic (algorithm 1). (See <u>'Umbilical hernia'</u> above.)
- Umbilical granuloma Umbilical granuloma is the most common umbilical mass in neonates. It is usually detected after cord separation because of persistent drainage. Topical <u>silver nitrate</u> is most commonly used to treat this lesion. (See 'Umbilical granuloma' above.)
- Omphalomesenteric duct anomalies Failed embryologic closure of the omphalomesenteric duct results in a range of anomalies including completely patent duct, umbilical polyp, Meckel's diverticulum, omphalomesenteric duct cyst, or intraabdominal fibrous band. (See 'Embryology' above and 'Omphalomesenteric duct anomalies' above.)
- Urachal anomalies Failed embryologic closure of the urachus results in a range of anomalies including patent urachus, umbilical polyp, bladder diverticulum, or urachal cyst. (See <u>'Embryology'</u> above and <u>'Urachal anomalies'</u> above and 'Umbilical polyp' above.)
- Omphalitis Omphalitis, a polymicrobial infection of the umbilicus and surrounding tissue, occurs predominantly in the neonate. Complications include sepsis and necrotizing fasciitis, which has a high mortality rate. (See 'Omphalitis' above and 'Necrotizing fasciitis' above.)
 - Antibiotic treatment of omphalitis is required and is directed against Gram-positive and Gram-negative organisms. We suggest initial parenteral administration of antistaphylococcal and aminoglycoside agents rather than topical or oral

antibiotic therapy, in infants with omphalitis to reduce the risk of severe complications (Grade 2C). In communities with a high prevalence of methicillin-resistant Staphylococcus aureus, we suggest vancomycin be administered rather than an antistaphylococcal penicillin (**Grade 2C**). (See 'Omphalitis' above.)

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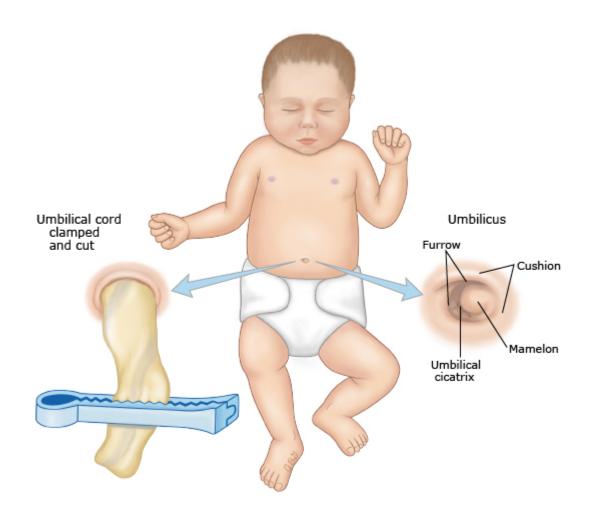
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Topic 5009 Version 45.0

GRAPHICS

The normal umbilicus

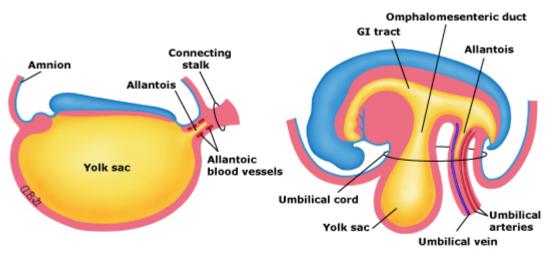


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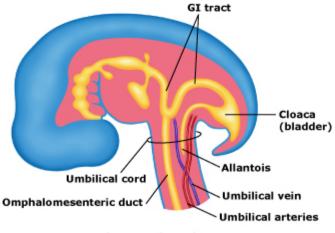
Graphic 74008 Version 3.0

Umbilical cord development third to fifth week gestation



A. Three-week embryo

B. Four-week embryo



C. Five-week embryo

GI: gastrointestinal.

Graphic 57342 Version 3.0

Antiseptic topical cord care agents and rare associated complications

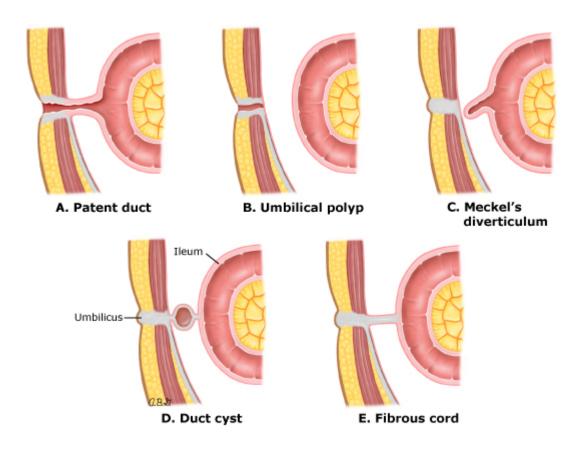
Agent	Rare complication
1 percent basic fuchsine	None
Bacitracin	None
Colloid silver-benzyl-peroxide	None
Micronized green clay powder	None
Chlorhexidine	Slight delay in cord separation
Hexachlorophene	Spongiform myelinopathy
Isopropyl alcohol	Percutaneous toxicity, central nervous system depression, skin necrosis
Neomycin	Neural deafness
Povidone-iodine	Transient hypothyroxinemia, hypothyroidism
Salicylic acid	Metabolic acidosis
Silver sulfadiazine	Kernicterus (sulfa)
Triple dye	Carcinogenicity

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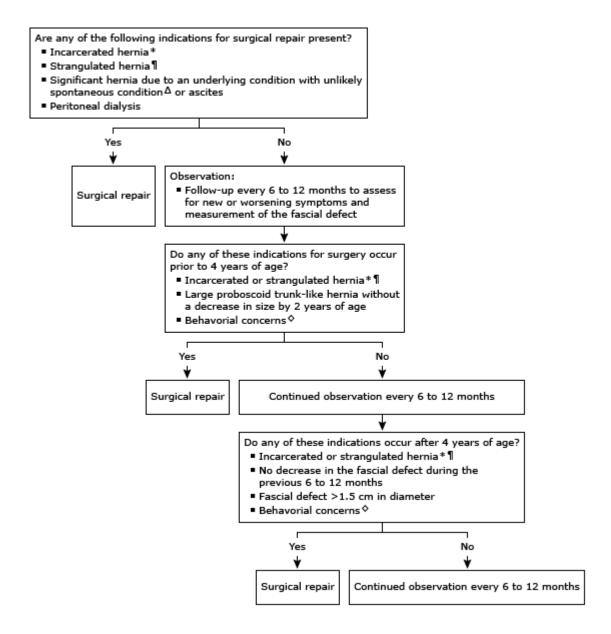
Graphic 76549 Version 4.0

Anomalies of the omphalomesenteric duct



Graphic 62526 Version 2.0

Management of umbilical hernia in children



The management of umbilical hernia in children is based on the natural course of spontaneous resolution in most asymptomatic individuals and a higher recurrence rate if surgical correction is performed before 4 years of age. Refer to the UpToDate topic on umbilical hernia for additional details.

- * Incarcerated hernia is defined as inability to reduce hernia by manipulation.
- ¶ Strangulated hernia is defined as vascular compromise of the contents of an incarcerated hernia.

 Δ Underlying conditions in which spontaneous closure is unlikely include: Ehlers-Danlos syndrome, Beckwith-Wiedemann syndrome, Down syndrome, mucopolysaccharidoses, hypothyroidism, and trisomy18.

♦ Behavioral concerns include poor feeding, chronic pulling on the hernia, bullying, and shame.

Graphic 132454 Version 1.0

Proboscoid hernia warranting early repair



Courtesy of Mary L Brandt, MD.

Graphic 63270 Version 2.0

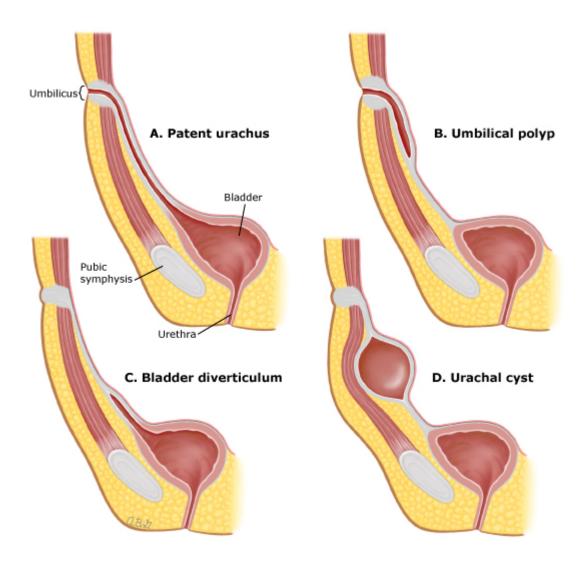
Umbilical granuloma



Courtesy of Mary L Brandt, MD.

Graphic 58481 Version 2.0

Anomalies of the urachus



Graphic 79324 Version 3.0

Omphalitis



This picture shows omphalitis, or infection of the umbilical cord stump, in an infant with leukocyte-adhesion deficiency.

Graphic 63960 Version 5.0

Omphalitis



Omphalitis with induration and erythema surrounding the umbilicus in a newborn.

Courtesy of Martin I Lorin, MD.

Graphic 71634 Version 3.0

Omphalitis shoelace



Omphalitis with induration and erythema surrounding the umbilicus in a newborn Native American infant. The umbilicus was tied with a shoelace.

Courtesy of Martin I Lorin, MD.

Graphic 50750 Version 3.0

Contributor Disclosures

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