

PEDIATRIC UMBILICAL PROBLEMS

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EMBRYOLOGY

Umbilical problems encountered in the pediatric population are often associated with retained umbilical cord structures or with failure of the umbilical ring to close at birth. Therefore, an understanding of the embryologic development of the umbilical cord and the umbilicus is essential when considering the differential diagnosis for pathology found in this region.

The development of the embryo begins with establishment of the posterior body wall.¹⁰ At this stage, the amniotic cavity is dorsal to the posterior body wall and the yolk sac is ventral (Fig. 1A). As the embryo grows, the anterior abdominal wall develops by simultaneous cranial, caudal, and lateral infolding. This infolding attenuates the yolk sac. Part of the yolk sac becomes intracoelomic to later develop into the midgut and hindgut (Fig. 1B). The attenuated connection between the extracoelomic yolk sac and the midgut is called the vitelline or omphalomesenteric duct, and is supplied by the vitelline artery and vein.

As growth continues, the vitelline duct in the yolk stalk fuses with the chorion or body stalk that contains the umbilical vein and arteries and the allantois, which is a projection of the hindgut portion of the intracoelomic yolk sac that later becomes the bladder. The paired vitelline veins develop into the hepatic and portal veins, and the vitelline artery contributes to the creation of the superior mesenteric artery.¹⁹

The umbilical cord represents the fusion of the yolk stalk containing the vitelline duct, and the body stalk containing the paired umbilical arteries, the umbilical vein, and the allantois (Fig. 1C). It also contains primitive mesenchymal tissue (Wharton's jelly) and is covered by an outer layer of amnion.

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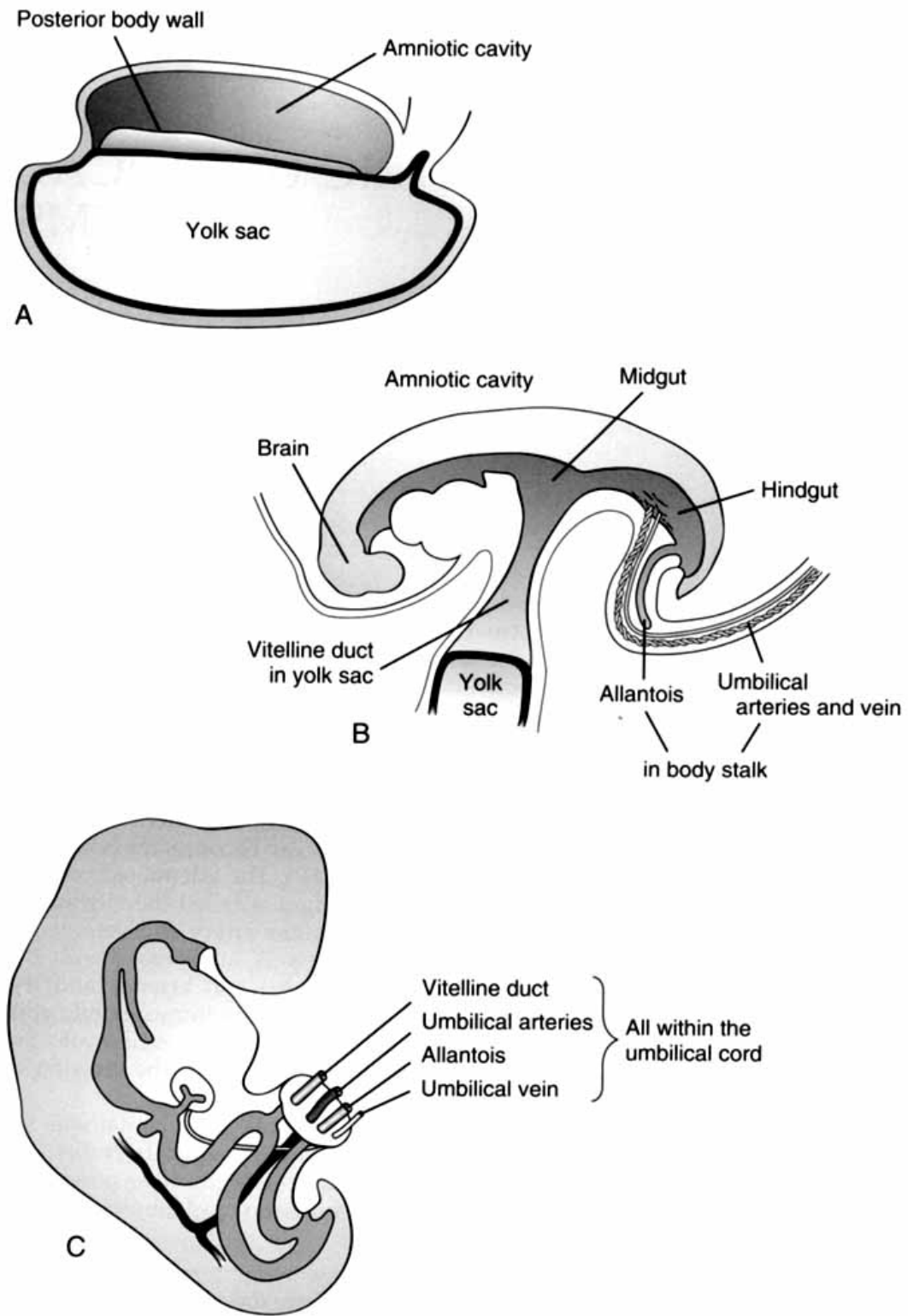


Figure 1. Development of the umbilical cord. *A*, Posterior body wall is established. *B*, Vitelline duct forms with craniocaudal and lateral infolding. *C*, The umbilical cord forms with the fusion of the yolk stalk and body stalk.

Normally, the vitelline duct obliterates between the 5th and 9th week of intrauterine life. The umbilical arteries and vein close shortly after birth and become the lateral umbilical ligaments and the ligamentum teres. The urachus or median umbilical ligament is the obliterated portion of the allantois that runs from the dome of the bladder to the umbilicus. The urachus forms between the 8th and 16th week of gestation. The area of umbilical cord insertion contracts to a fibromuscular ring that closes at birth. The umbilical cord separates from the abdominal wall 5 to 8 days after birth.

Pathology at the umbilicus can manifest as inflammation, drainage, a palpable mass, or herniation. These signs may occur in combination and may or may not be associated with retained umbilical cord elements.

OMPHALITIS

Omphalitis is defined as erythema and edema of the umbilical region with or without drainage from the umbilicus.⁴ Bacterial colonization of the umbilical stump occurs soon after delivery. The devitalized stump is a medium that supports bacterial growth. Most commonly, omphalitis in the newborn is owing to either poor hygiene or a hospital-acquired infection.¹⁸

Mild cases of omphalitis usually respond to local application of alcohol and drying of the inflamed area.⁴ However, omphalitis can rapidly progress from a localized infection to necrotizing fasciitis of the abdominal wall. Therefore, most cases of erythema and edema at the umbilicus, especially when associated with drainage, warrant hospital admission, pediatric surgical consultation, and treatment with intravenous antibiotics. Antibiotic coverage should be effective against *Staphylococcus*, *Streptococcus*, and gram-negative rods. The margins of erythema should be marked and recession of these margins followed closely. With intravenous antibiotics and local wound care, the erythema and drainage should improve within 12 to 24 hours.²¹

In a recent report, approximately 16% of those neonates admitted for omphalitis went on to develop necrotizing fasciitis of the abdominal wall.²¹ All were full-term infants with no apparent risk factors for sepsis. The discrimination between omphalitis and necrotizing fasciitis is an important one as necrotizing fasciitis has a mortality rate of greater than 50%. Neonates with necrotizing fasciitis show signs of systemic sepsis with tachycardia, leukocytosis or leukopenia, and fever. The bacteria grown from culture of the umbilical drainage in necrotizing fasciitis tends to be polymicrobial rather than the single organism that is usually isolated in simple omphalitis.²⁰ The appearance of the periumbilical skin can be helpful in making the diagnosis of necrotizing fasciitis. Periumbilical skin that is violaceous or has a peau d'orange appearance indicates involvement of deep tissue layers. Necrotizing fasciitis is a surgical emergency with the only chance for cure being debridement of all devitalized tissue.

UMBILICAL GRANULOMA

When the fibromuscular ring of the umbilicus closes and the umbilical cord sloughs, the ring is covered anteriorly by skin and posteriorly by peritoneum. After cord separation, there may be incomplete epithelialization over the ring and an area of beefy red tissue or granulation tissue is visible. Granulation tissue formation is a normal stage in wound healing and represents endothelial cell division and migration to form a rich bed of new capillaries. Granulation tissue

can overgrow, and at the umbilicus can result in an umbilical granuloma. Umbilical granulomas can be treated by one or two applications of silver nitrate that results in a chemical cautery. If the granuloma is too large, or fails treatment with several applications of silver nitrate, it may need to be excised. When excised, all tissue is sent to pathology to rule out retained umbilical remnants (i.e., vitelline duct or urachal remnants). Once the granulation tissue is ablated, the base of the wound will heal with complete epithelialization.^{18, 23}

Umbilical granuloma is the most common umbilical abnormality in the neonate causing inflammation and drainage. If it does not respond to silver nitrate treatment, the abnormality may in fact be a remnant of the vitelline duct or urachus and referral to a pediatric surgeon is required.¹⁵

VITELLINE DUCT ANOMALIES

Two percent of the population can be expected to have a vitelline duct remnant, also called omphalomesenteric duct remnants.²⁵ A *Meckel's diverticulum* is the most common type (Fig. 2). It is a true diverticulum of the small bowel occurring approximately 60 cm from the ileocecal valve. Since a Meckel's diverticulum alone will not cause symptomatology at the umbilicus, it can present with (1) massive rectal hemorrhage when retained gastric mucosa is present, (2) as a small bowel obstruction when it serves as a lead point for intussusception, or (3) when the diverticulum is attached to the umbilicus by a fibrous band (Fig. 2B) around which the small bowel may volvulize. The majority of people with Meckel's diverticulum (70% to 95%) remain asymptomatic. Children repre-

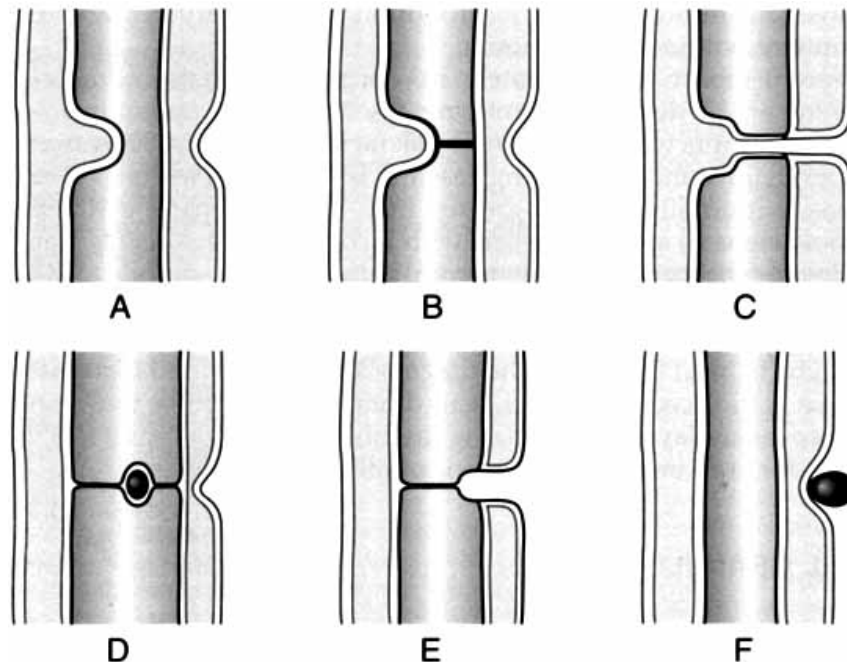


Figure 2. Vitelline duct anomalies. *A*, Meckel's diverticulum. *B*, Fibrous band attaching a Meckel's diverticulum to the abdominal wall. *C*, Umbilical fistula. *D*, Vitelline cyst. *E*, Umbilical sinus. *F*, Umbilical polyp.

sent the majority of symptomatic patients. A detailed discussion of Meckel's diverticulum is presented in the article by Irish et al elsewhere in this issue.

An *umbilical fistula*, which in a recent report represented only 2% of children treated over a 16-year period for vitelline duct anomalies,²⁵ is a patent vitelline duct (Fig. 2C). Usually, persistent drainage at the umbilicus is present. The drainage may be bilious, or if the tract is secondarily infected, may be purulent. The diagnosis of an umbilical fistula may not be apparent on physical examination alone. A fistulogram to define the abnormality is helpful. A small catheter is inserted into the umbilical opening and radiopaque dye is injected. The diagnosis of an umbilical fistula is confirmed when the dye is visualized in the lumen of the small bowel.

When the vitelline duct is only partially obliterated, a *vitelline cyst* (Fig. 2D) or a *vitelline sinus* (Fig. 2E) may be present. Vitelline cysts are rare, but can present as an umbilical mass. The vitelline sinus presents with persistent umbilical drainage and is usually diagnosed in the older child. Again, injection of contrast material may be helpful and a blind-ending sinus will be visualized.

An *umbilical polyp* (Fig. 2F) also presents as an umbilical mass and may initially be confused with an umbilical granuloma. After the cord separates, it is seen as a glistening, cherry-red nodule. Unlike the umbilical granuloma, the polyp is a remnant of the vitelline duct and consists of small bowel mucosa. In addition, it will not regress with the application of silver nitrate. Since the origin of the *umbilical polyp* has been exclusively attributed to a remnant of the vitelline duct, there is now one case report where an umbilical polyp appears to have originated from the urachus.¹⁶

All symptomatic vitelline duct anomalies need to be excised. Unlike a Meckel's diverticulum that may present as a surgical emergency, those anomalies presenting with umbilical inflammation and drainage may be electively repaired after local wound care and intravenous antibiotics clear the area of infection. The umbilicus must be fully explored and all vestiges of the vitelline duct excised including intra-abdominal components.¹⁴

URACHAL ANOMALIES

Analogous to the vitelline duct anomalies, complete or partial patency of the urachus can present with drainage and infection at the umbilicus.⁹ When the urachus is patent, a *vesicoumbilical fistula* is present (Fig. 3). Here, the child may have recurrent cystitis and urine draining at the umbilicus. A fistulogram will show contrast dye tracking caudally to the bladder in the extraperitoneal space and will distinguish this anomaly from a patent vitelline duct. A urinary outflow obstruction such as posterior urethral valves should be ruled out as the origin of the the patent urachus.²²

A *urachal sinus* may be a cause of umbilical drainage and infection and results from a distally patent urachus (Fig. 3B). A proximally patent urachus represents a bladder diverticulum analogous to a Meckel's diverticulum.

A *urachal cyst*, the most common of the urachal anomalies,⁹ results from a patent mid-portion of the urachal lumen with the distal and proximal lumens obliterated. A urachal cyst may present as a midline infraumbilical mass. It may become infected and rupture either through the umbilicus or rarely into the peritoneal cavity causing peritonitis.

Like the vitelline duct anomalies, vestiges of a symptomatic patent urachus should be surgically excised. This requires general anesthesia, and can be done through an infraumbilical incision. Complete excision of the urachus to the

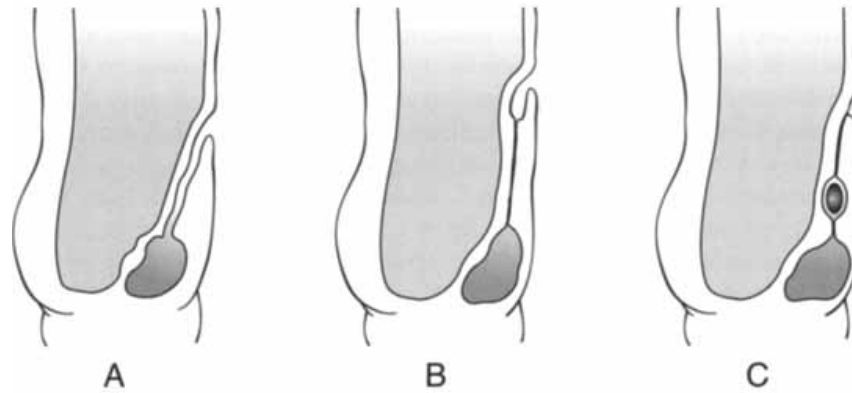


Figure 3. Urachal anomalies. A, Fistula. B, Sinus. C, Cyst.

dome of the bladder should be carried out and can now be performed laparoscopically.⁸

DELAYED UMBILICAL STUMP SEPARATION

Recent publications have raised concerns regarding children with a history of separation of the cord later than 3 weeks.^{1, 7, 12, 26} Umbilical cord separation is dependent upon infiltration with neutrophils and monocytes, and subsequent necrosis, after which the most common abnormal lesion is a granuloma. These frequently regress with local treatment, or may be excised. On occasion, intestinal mucosal remnants are seen in removed specimens, and these children have a 30% or greater incidence of having a significant omphalomesenteric or vitelline duct remnant.¹⁴ An increased susceptibility to bacterial infections as a reproducible effect of complement receptor 3 (CR3) deficiency has been described.¹⁷ The exact nature of the immune defect associated with delayed umbilical separation has been the subject of extensive investigation. A familial occurrence has been noted, and an X-linked dominant mode of transmission postulated.⁶ Early reports identified a neutrophil defect,³ and further studies implicated malfunction of monocytes and natural killer cells.^{1, 3} Chemotaxis, phagocytosis, and the oxidative burst were all affected.¹¹ The availability of monoclonal antibodies to specific cell surface proteins and glycoproteins on the surface of phagocytes led to the discovery of a deficiency of the membrane surface antigen CR3.¹⁷ In patients with delayed umbilical cord separation, either the primary care physician or the pediatric surgeon should refer the infant to a pediatric immunologist for CR3 evaluation. Identification of a CR3 defect, whether partial or complete, will aid in subsequent surveillance and treatment of bacterial infections in both the child and siblings. Although this defect is rare, even partial lack of CR3 can lead to fatal sepsis.¹²

UMBILICAL HERNIA

An umbilical hernia develops when the umbilical ring fails to close after cord separation. The defect is covered posteriorly by peritoneum and anteriorly by skin. It is noticed as a protrusion at the umbilicus that is more pronounced

when the infant strains and reduces when the infant is supine and at rest. The diameter of the defect is rarely greater than 2 cm. The entire circumference of the umbilical ring should be carefully palpated. If the fascial ring is not completely intact, spontaneous closure is unlikely, and referral to a pediatric surgeon is appropriate.

There is a racial predilection for umbilical hernia with African-American infants 6 to 10 times more likely to incur the defect than white infants.² Low birth weight is also a predisposing factor. More than 80% of infants weighing less than 1200 g have evidence of an umbilical hernia compared with 21% of infants weighing over 2500 g at birth.¹⁸

Umbilical hernias in the pediatric population rarely incarcerate. The natural history of umbilical hernia is that the majority close by the age of 3 years. Spontaneous closure of the umbilical hernia is unlikely after the age of 4 to 5 years. Children greater than 3 years of age with a defect greater than 1.5 cm are also unlikely to have spontaneous closure. The risk of incarceration and strangulation increases with increasing age. Therefore, it is recommended that all defects that have not spontaneously closed by age 4 or 5, be surgically repaired.

Surgical repair is through an infraumbilical incision. The hernia sac is excised and the fascial defect closed with suture. The under surface of the umbilical skin is tacked to the fascia to recreate an umbilical invagination and the skin incision is closed.

PROBOSCOID UMBILICAL HERNIAS

The proboscoid umbilical hernia is an important subset of umbilical defects, representing 17% of all umbilical hernias repaired at our institution. They have a similarly sized fascial defect as all umbilical hernias, but they differ in the amount of redundant overlying skin. As a result, they have a characteristic protuberant or *proboscoid* appearance (Fig. 4). As in all umbilical hernias, time alone may result in spontaneous closure of the fascial defect. However, proboscoid hernias may require early prophylactic repair for social, cosmetic, or surgical concerns. The authors advocate early repair if:

1. the hernia is aesthetically displeasing to the patient or parents,
2. if there are concerns regarding excessive manipulation of the hernia by the child,
3. if a female child attempts to urinate from the hernia, or
4. if the proboscis increases in size.

OTHER UNUSUAL UMBILICAL PROBLEMS

Drainage from the umbilicus in neonates and infants is usually owing to a remnant of the vitelline duct or a patent urachus. There have been case reports of rarer anomalies such as an infected vestigial umbilical artery,⁵ persistence of the vitelline artery,²⁷ and a fistula between the vermiform appendix and a patent vitelline duct.¹³

Pilonidal disease of the umbilicus has been reported. The most recent study describes a series of young men with an average age of 21 years who presented with umbilical discharge, edema, and cellulitis. In all cases, the umbilicus was excised and the acute inflammation was owing to the presence of hair within what appeared to be an acquired umbilical sinus.²⁴

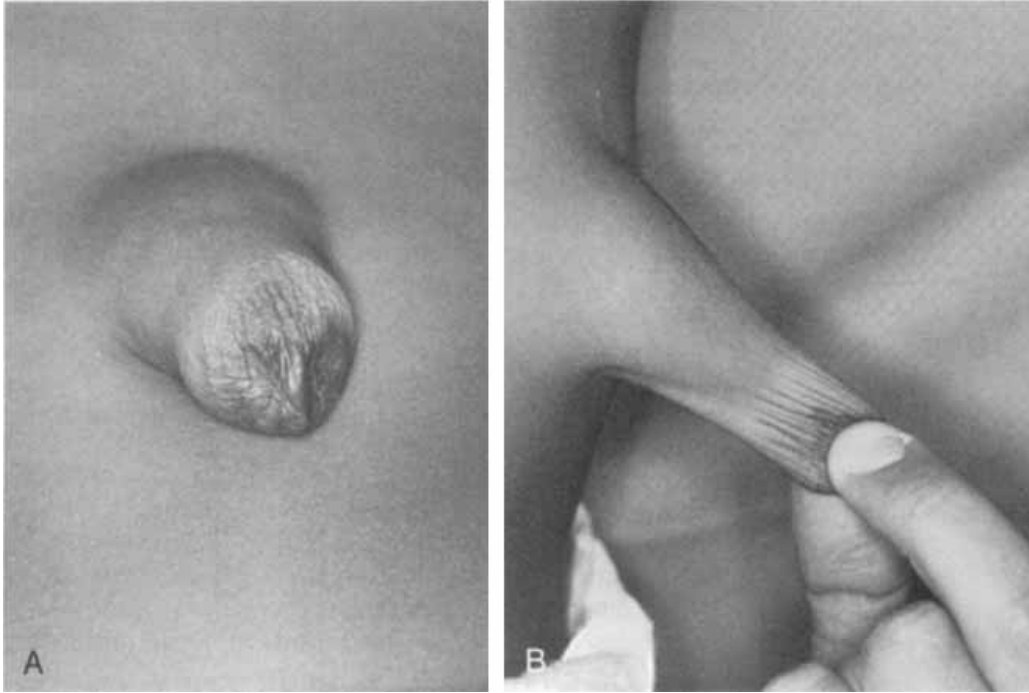


Figure 4. Proboscoïd umbilical hernia. *A*, Anterior view. *B*, Lateral view.

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