Anomalies, abnormalities, and care of the umbilicus

Albert Pomeranz, MD

Department of Pediatrics, Medical College of Wisconsin, Children's Hospital of Wisconsin, Downtown Health Center, 1020 North 12th Street, Milwaukee, WI 53233, USA

The umbilical cord, along with the anterior abdominal wall, begins development by the end of the third week of gestation. By the twelfth week, embryonic remnants leave one umbilical vein and two umbilical arteries covered by amnion, which are eventually protected and incased by Wharton’s jelly. Although it is shed after birth, remnants remain clinically relevant in the newborn period and early childhood. This article discusses abnormalities that involve the umbilical cord and issues related to its care after birth.

Umbilical cord anomalies and abnormalities

Cord characteristics

The mean length of the cord is 55 cm [1]. Short cords are associated with decreased fetal movement as a result of either maternal or fetal causes. Maternal causes include oligohydramnios and uterine abnormalities that result in less fetal space; fetal causes include structural limb defects, such as amniotic bands and neurologic limb defects, as seen in Werdnig-Hoffman syndrome [2]. Long cords (> 70 cm) are associated with fetal entanglements, true knots, and cord prolapse. The umbilical cord contains several loops or coils that are established early in fetal development. These coils are believed to supply protection from compressive forces placed on the cord. Abnormal coiling of the cord is associated with various adverse neonatal outcomes, including premature delivery and fetal demise [3–5].

Single umbilical artery

A single umbilical artery (SUA) is a relatively common congenital malformation and often is associated with other significant malformations. In a meta-
analysis in which the diagnosis was made by examining placental and umbilical cord specimens from live born infants or by examining the umbilicus after birth, the mean incidence was 0.55%, with 27% of these having an associated malformation [6]. This statistic emphasizes the importance of examining the newborn cord closely and performing a careful search for additional abnormalities if a two-vessel cord is detected. The associated malformations varied, with a substantial number being multiple and having associated chromosomal abnormalities. Although the increased risk of morbidity and mortality in newborns with SUA is largely related to the concurrent fetal and placental malformations, even nonmalformed infants with SUA may have an increased perinatal mortality rate [4].

The controversial subject is what to do about isolated SUA without any physical evidence of associated anomalies. In the meta-analysis of the newborns with isolated SUA in which urologic evaluations were performed, 16% had renal anomalies [6]; however, 54% of these were considered minor or self-limiting, which left 7.3% of cases as clinically significant. The authors of the study believed that screening was not cost effective and that many of the anomalies would be diagnosed in follow-up, with the value of early detection unclear. The number of patients (204) in this study with asymptomatic SUA was relatively small, and no control groups without SUA were available.

The results and opinion of the authors of another study illustrated the subjectivity involved in the recommendation regarding screening of newborns with isolated SUA. In the study by Bourke et al [7], which was one of the studies included in the previously discussed meta-analysis, the authors performed screening ultrasounds over a 6-year period on all newborns born in their hospital with isolated SUA. They identified 112 cases and found significant renal abnormalities in 8 (7.1%) of these newborns. Four of these patients had grade 2-3 ureteropelvic reflux, yet the authors believed that screening was justified. Until a prospective study that compares the rate of significant renal abnormalities in isolated SUA with a healthy control group is performed, a firm recommendation cannot be made. Currently, however, this author is in favor of screening these infants with renal ultrasound.

Urachal abnormalities

The urachus is an embryonic remnant of the communication between the bladder and the umbilicus and generally exists as a fibrous cord. Four clinical anomalies have been described: urachal sinus, urachal cyst, patent urachus, and urachal diverticulum [8]. In a study of 48 patients by Cilento et al [8], the presenting complaint was periumbilical discharge in 42%, followed by a mass or cyst and abdominal or periumbilical pain. In another study of 21 patients, 57% presented with periumbilical discharge [9]. The most common anomalies were urachal sinus and urachal cyst.

The urachal sinus may present with drainage, pain, or periumbilical redness [9]. If the discharge empties into the bladder, symptoms of a urinary tract in-
fection may occur [8]. The differential diagnosis includes a patent urachus, patent omphalomesenteric duct, umbilical granuloma, and omphalitis. If an opening can be cannulated, sinography is the study of choice because it allows the diagnosis of a patent urachus or patent omphalomesenteric duct that communicates with the bowel [8].

Urachal cysts present in several ways, typically in early childhood. Although urachal cysts may present as a palpable periumbilical mass, they most often present as an infected cyst with abdominal pain, erythema, and periumbilical swelling, generally below the umbilicus. The cyst may rupture and drain from the umbilicus or intraperitoneally, which results in an acute abdomen. *Staphylococcus aureus* is the most common isolated organism. Occasionally they may be detected incidentally during other radiologic procedures secondary to calcification. The diagnosis usually can be made by ultrasound [8,9].

A patent urachus involves free communication between the bladder and the umbilicus, which allows urine to flow from the umbilicus. It presents with a persistent wet or draining cord and occasionally as a urinary tract infection [10]. The drainage is usually thin and clear. The differential diagnosis includes abnormalities that result in a wet or draining cord, similar to those discussed for a urachal sinus. The umbilical granuloma may be distinguished by the nature of the discharge and presence of the granuloma on examination. A sinogram aids in diagnosis in most cases and should reveal the presence of a patent omphalomesenteric duct. A cystogram is often still recommended to rule out a urinary obstructive lesion, such as posterior urethral valves. The patent urachus is believed to act as a “pop off” valve in these obstructed cases [11].

**Omphalomesenteric duct (vitelline) abnormalities**

The omphalomesenteric duct forms a connection between the intestinal tract and abdominal wall during fetal development. Several possible remnants are produced, the most common being a Meckel’s diverticulum. Symptomatic Meckel’s diverticulum most commonly presents in the young child as painless rectal bleeding from ectopic gastric mucosa, although it may be the lead point for intussusception in neonates and infants. If the duct remains patent to the umbilicus, fecal drainage from the umbilicus can occur. The differential diagnosis includes the urachal remnants and an umbilical granuloma, but the nature of the discharge is often helpful [10,11]. Other remnants include bands, which result from the attachment between the abdominal wall and intestine, and cysts. These bands may result in volvulus and intestinal obstruction; cysts may become infected and less commonly result in obstruction.

In a large study of 217 pediatric cases of omphalomesenteric duct remnants, neonates and infants most commonly presented with intestinal obstruction [12]. Three neonates and one older infant (6 weeks old) with a patent omphalomesenteric duct all presented with bilious drainage from the umbilicus. An umbilical sinogram with contrast aided in diagnosis in these cases. All of the urachal and omphalomesenteric abnormalities require surgical intervention.
Umbilical granuloma

Umbilical granulomas commonly come to the attention of parents because of persistent drainage or moisture involving the umbilicus after the cord has dried and separated. It is not a congenital abnormality but represents continuing inflammation of granulation tissue that has not yet epithelialized. The lesion is a round, wet, often pink, pedunculated lesion of variable size, usually 3 to 10 mm. The contribution of colonized bacteria and low-grade infection is unclear. Urachal duct and omphalomesenteric duct remnants that produce umbilical wetness or discharge must be considered in the differential diagnosis. Umbilical polyps, which represent a distal omphalomesenteric remnant, appear similar to a granuloma. Failure of a presumed umbilical granuloma to respond to treatment with silver nitrate should raise suspicion that one is dealing with one of these other abnormalities.

The most commonly used treatment of umbilical granuloma is silver nitrate cauterization that comes mounted on a wooden stick applicator in a concentration of 75%. This chemical acts as an antiseptic, astringent, or caustic agent depending on the concentration [13]. The application may be repeated every few days as necessary but generally requires only a few treatments for success. Its use can result in burns to periumbilical skin, so caution should be exercised during application [14]. Surgical removal of the granuloma rarely is required.

Umbilical cord care, omphalitis, and necrotizing fasciitis

Umbilical cord care

The umbilicus is often the first site of colonization of S aureus and other organisms [15]. The presence of S aureus and the degree of colonization increase the risk of infection at the periumbilical site and other sites [16,17]. The care of the umbilicus in the newborn period has been directed at controlling this colonization in hopes of decreasing infection, particularly omphalitis and fasciitis. Various antibiotic and antiseptic agents have been used for this purpose and have been demonstrated to reduce colonization and infection rates [18–21]. Group B streptococcus colonization rates also have been inhibited by various antimicrobial agents [19,22].

Several different antimicrobial agents have been used to reduce colonization rates, including triple dye, bacitracin, silver sulfadiazine, povidone-iodine, chlorhexidine, and hexachlorophene. Although a recent Cochrane Review article was unable to determine the best cord care in developed countries, all of the agents mentioned previously have been shown to reduce colonization rates, but their effectiveness varies by the study [23]. Alcohol is not effective in controlling colonization, nor does it seem to shorten the time until cord separation [24].

Triple dye is one of the more commonly used antibacterial agents, and it seems to be effective at reducing group B streptococcus colonization and S aureus
More than other agents, however, this agent results in prolonged cord separation times [27] and leaves the cord discolored. Side effects or toxicities from the other agents rarely have been reported. Hexachlorophene bathing of infants was common up to 1971, at which time the Food and Drug Administration and American Academy of Pediatrics issued a statement discouraging its use because of fear concerning possible neurotoxicity after cutaneous absorption [28]. Its use on a small surface area, such as the umbilical stump, is not viewed as a problem, however. When used extensively and repeatedly on the newborn, povidone-iodine has been reported to result in transient thyroid suppression [29], and there is some concern over the possibility of falsely elevated thyroid-stimulating hormone levels on the newborn screen when used only on the cord. A study conducted by Lin et al [30] demonstrated significantly more false-positive thyroid-stimulating hormone levels in screened newborns on whose umbilical cords povidone-iodine was used compared with alcohol or triple dye.

Several hospitals have discontinued antimicrobial treatment of the cord, which is partially a result of changes in maternity care. *S. aureus* cross-contamination of the umbilical stump from infant to infant via hospital personnel has been demonstrated [31]. It is assumed that the increase in single rooms and infants “rooming-in” with mothers should result in fewer personnel being involved and, therefore, less colonization [20]. Significant omphalitis and its major complication, necrotizing fasciitis, also are relatively rare [32,33].

A recent study by Janssen et al [20] illustrated the uncertainty surrounding cord care. The authors reported on a randomized trial that involved 766 newborns on whom triple dye/alcohol versus dry cord care was used; the dry cord care consisted of soap and water to the periumbilical area. Although only one case of omphalitis was reported in the dry cord care group compared with the treated group, which was not statistically significant, there was significantly greater colonization of *S. aureus*, group B streptococcus, and *Escherichia coli*. This statistic is concerning because of the association of *S. aureus* colonization, particularly when heavy with subsequent infection [34]. Community nurses also observed cord exudates and foul odor significantly more often in the dry cord care group during the home visit.

Prolonged cord separation times are a concern to parents and physicians. Antimicrobial treatment of the cord is considered to be a major cause [27,35]. A suggested explanation for this effect is that the lower colonization rates result in a decrease in leukocyte infiltration of the cord and its subsequent digestion by leukocytes. The mean cord separation times are reported to be 7 days with dry cord care [36] and approximately 15 days with triple dye [37,38]. In a study by Wilson et al [38], 10% of cords to which triple dye was applied in the nursery were still attached at 3 weeks.

Prolonged cord separation is important primarily for two reasons. The delay in separation may result in anxiety on the part of parents and health care personnel [39]. The delayed separation, particularly when associated with omphalitis, also may be the first sign of leukocyte adhesion deficiency. This is a rare disorder of neutrophils characterized by defective adhesion-dependent functions, such
as chemotaxis, which result in severe recurrent infections during infancy [40,41]. In most cases, cord separation was delayed beyond 3 weeks, with some infections presenting after cord separation. Newborns with cord separation times beyond 3 weeks should be monitored for signs of omphalitis and other infections commonly seen with this disease, such as aseptic meningitis, perianal infections, and otitis media [41]. Because these infants often have persistent leukocytosis with significant neutrophilia even when not infected [41], it is reasonable to screen asymptomatic infants with delayed cord separation times, such as beyond 4 weeks, with a complete blood count with differential. If leukocytosis is present, further immunologic evaluation is indicated. A less serious form of the disease, leukocyte adhesion deficiency type II, results from partial absence of the adhesion molecule.

In addition to the previously discussed causes of delayed separation of the cord, Razvi et al [42] reported two infants with delayed cord separation attributed to urachal cysts. Both infants were diagnosed by ultrasound and had negative test results for immune defects. One infant had developed umbilical discharge at 8 weeks of age.

In conclusion, when dry cord care of the umbilicus is used, parents must be educated about signs of infection and the need to contact their health care provider immediately if any of these signs are present. In less developed countries, where adequate cord care cannot be guaranteed, application of antimicrobial and aseptic agents to the cord is recommended.

Omphalitis

Omphalitis is an infection of the umbilical cord stump or the surrounding tissue that results from organisms that have colonized the area. One of the main reasons for antiseptic treatment of the umbilical cord in the newborn is prevention of this disease. Because the umbilicus presents access to the portal vein via the umbilical vein, infection in this area may result in portal vein thrombosis, phlebitis, and liver abscesses. A long-term complication is extrahepatic portal hypertension. The process also may spread through fascial plains and result in peritonitis [43]. Although rare, the development of necrotizing fasciitis may occur and is associated with a high mortality rate [33,44]. Omphalitis is a polymicrobial infection; the most common organisms involved include S. aureus, group A streptococcus, and gram-negative bacteria [44]. Although anaerobes rarely have been documented in this infection, they have been recovered in up to 39% of patients when looked for aggressively [44,45].

The clinical presentation of omphalitis is characterized by drainage from the umbilical stump and signs of periumbilical inflammation, such as erythema induration and swelling of the skin. Associated systemic signs, including lethargy, fever, irritability, and poor feeding, indicate more severe infection or complications. The difficulty with the diagnosis lies with the mild presentation of discharge from the umbilical stump without inflammatory signs, particularly after cord separation. This occurrence may be normal even when associated
with some odor [44]. Some physicians treat infants with minimal symptoms with a topical agent, such as alcohol, and application of topical antibiotics, such as bacitracin or mupirocin [44], although there is no evidence of the efficacy of this practice. Infants with omphalitis must be followed closely for any signs of inflammation, particularly erythema of the periumbilical skin beyond a short distance (>0.5 cm) from contact with the cord [20]. In infants without inflammation, it is important to consider other causes of moisture or discharge from the cord, including umbilical granuloma and urachal and omphalomesenteric duct anomalies.

Patients with omphalitis who have signs of inflammation should have the discharge or pus cultured after the skin has been decontaminated and blood cultures obtained, particularly if they are febrile. Investigation of other sites where infection may have spread is indicated based on the clinical status of the patient. Systemic antibiotics that cover the organisms most commonly involved, including S aureus, streptococcus, and gram-negative bacteria, are necessary. Coverage for anaerobes also should be considered, especially for infants with foul-smelling drainage or infants born of mothers with amnionitis [44]. In cases that involve mild signs of inflammation but no systemic signs or symptoms, oral antibiotic therapy may be reasonable but must be accompanied by close follow-up [43].

Necrotizing fasciitis

Necrotizing fasciitis is a rare, often fatal complication of omphalitis that involves infection of the skin, subcutaneous fat, and superficial and deep fascia. It progresses rapidly. In a review of the literature from 1999 [32], which yielded 66 cases of necrotizing fasciitis, the overall mortality rate was 59%. Most cases were secondary to omphalitis and were polymicrobial. The most common aerobic bacteria were S aureus, E coli, and enterococcus; clostridium species and bacteroides were the predominant anaerobic bacteria. The rapid progression of inflammation along with marked tissue edema and signs of systemic toxicity are characteristic of the infection [44]. Early diagnosis is essential because in addition to antibiotic therapy, immediate surgical débridement results in the greatest chance for survival.

Summary

An SUA is the most common anomaly of the umbilical cord and is often associated with other congenital anomalies. When present as an isolated phenomenon without other physical abnormalities, the decision to screen for associated renal anomalies is debatable but recommended by this author. Urachal and omphalomesenteric anomalies may present in the newborn period with umbilical cord drainage or wetness. They must be distinguished from an umbilical granuloma, which is by far the more common abnormality.
The use of antimicrobial and antiseptics agents on the umbilical stump prevents bacterial colonization and infection, particularly omphalitis and necrotizing fasciitis. With changes in maternal care resulting in less bacterial cross-contamination of infants and the relative rarity of these infections, many hospitals have adopted dry cord care. In this situation, parents must be educated carefully about signs of infection and the need to contact a health care provider immediately if present. Although long cord separation times may result from the use of antimicrobial agents on the cord stump, they also may be associated with leukocyte adhesion abnormalities, especially when associated with infections in the neonatal period. For infants whose cords persist beyond 3 to 4 weeks without associated infection, a screening complete blood count to detect leukocytosis is a reasonable screen for this disorder. If leukocytosis is present or neonatal infection occurs, further immunologic testing is required.

References