



# YOUR SURGICAL CONSULTANT

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We at Pediatric Surgical Associates greatly appreciate our collegial relationship with referring physicians such as you. Best wishes for the coming year to you and your families, and to the members of your staff.

**Please note: For your patients' convenience, beginning February 1 we will be opening an office in Grapevine located at 2020 W Hiway 114 Suite 110, near the Baylor Grapevine Medical Center. We will be available for preoperative consultations and postoperative follow-up visits each Tuesday afternoon from 1 to 5 pm and each Thursday morning from 9 to 12 am.**

## UMBILICAL DISORDERS OF CHILDHOOD

Abnormalities of the umbilicus are interesting and varied. The classic treatise is a 1916 text by Thomas Cullen.

**Embryology.** At 3 weeks of gestational age, the midgut is connected to the yolk sac at the umbilicus by the omphalomesenteric/vitelline duct with its artery and vein. By 4 weeks, the duct has been incorporated into the cord. At 6½ weeks, the duct contracts, the intestine elongates, and the liver enlarges forcing the intestine into the cord. The midgut rotates 90° counterclockwise around the duct. Between 8 and 10 weeks, the intestines exit the cord and reenter the peritoneal cavity. The allantois persists as the urachus.

The left umbilical vein and both umbilical arteries persist. The vitelline artery normally obliterates distally but persists proximally as the superior mesenteric artery. The vitelline vein also normally obliterates distally but persists proximally as the portal vein. The omphalomesenteric duct ordinarily obliterates completely; failure to do so results in potentially serious conditions; some are reviewed below.

**Normal and Abnormal Anatomy of the Cord and Umbilicus.** Normal cross-sectional anatomy of the cord reveals two umbilical arteries and one umbilical vein. A single umbilical artery is found in 0.5 to 0.9% of births; it is more common in females and in whites. It is also occasionally associated with infants of diabetic mothers, with certain

trisomy anomalies, and with the usage of certain drugs during pregnancy. There is a reported incidence of associated congenital abnormalities of up to 53%, including cardiovascular, pulmonary, gastrointestinal, genitourinary, cerebrospinal, musculoskeletal, facial, and ocular anomalies. A thorough physical examination, especially for dysmorphism, parental counseling, and an earlier and more detailed follow-up examination are indicated. Special screening studies for cardiac, renal, or other anomalies are obtained only if clinically indicated. In one series, (29%) of all infants with a single umbilical artery died perinatally, (16%) lived but were malformed, and (55%) were alive and normal.

Although the appearance of a "normal" umbilicus is far from consistent, certain syndromes have characteristic umbilical dysmorphism as part of their clinical presentations. Most patients with Aarskog syndrome (short stature with facial, digital, and genital anomalies) have a prominent and protruding central portion of the umbilicus surrounded by a deep ovoid depression. Many patients with Reiger syndrome (goniodysgenesis and hypodontia) have a broad prominent umbilicus with redundant umbilical skin. The Robinow syndrome (fetal face, forearm brachymelia, genital hypoplasia, moderate dwarfing) includes an abnormally high positioned, broad, and poorly epithelialized umbilicus.

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**Disorders of the Umbilical Stump.** With a competent immune system and routine care, the umbilical stump should atrophy and separate 12 to 14 days following birth. A stalk persisting longer than three to four weeks should be surgically explored and excised since persistent omphalomesenteric remnants or ectopic viscera may be present within a persistent stump. Simple ligation could be disastrous because bowel might be inadvertently ligated.

An umbilical granuloma, or pyogenic granuloma of the umbilicus, occurs commonly within a week or two after the cord stump separates. A granuloma represents a low-grade infection of the site with the resultant formation of granulation tissue which typically produces a slight discharge. The granuloma is usually small and cherry-red, and may have a stalk; it must be differentiated from an umbilical polyp (see below) by application of silver nitrate which is curative for a granuloma, while a polyp will remain unchanged. There are no anomalies associated with umbilical granulomas.

**Incomplete Obliteration of the Omphalomesenteric Duct.** Partial disappearance of the omphalomesenteric duct produces an umbilical polyp or sinus, an omphalomesenteric cyst, fibrotic bands between the umbilicus and intestine, or a Meckel's diverticulum. Complete persistence results in an omphalomesenteric fistula.

Ectopic intestinal or gastric mucosa comprises an umbilical polyp. Pancreatic tissue may be present. Skin excoriation around such a polyp signifies the presence of gastric or pancreatic tissue. Proper treatment consists of excision with histologic verification. Since more than one-half of such polyps are associated with underlying omphalomesenteric anomalies, a mini-laparotomy should be performed at the time of excision. Prognosis is excellent with surgical repair; if not, a volvulus, internal hernia, or complications of a Meckel's diverticulum (see below) may occur.

An umbilical sinus is prone to infection and should be excised. Surgical treatment resembles that of the polyp. The lack of fecal discharge from the umbilicus distinguishes an umbilical sinus from an omphalomesenteric fistula.

Omphalomesenteric cysts may occur within the abdominal wall or within a fibrotic omphalomesenteric band. These are usually not diagnosed unless they undergo torsion, become infected, or enlarge due to retained secretions. Persistent fibrous bands or patent vessels may connect the underside of the umbilicus to normal bowel or to a Meckel's diverticulum. Although often asymptomatic, these bands may provide an axis around which a volvulus may rotate.

A Meckel's diverticulum is a remnant of the intestinal portion of the omphalomesenteric duct and may contain gastric or pancreatic tissue. A Meckel's diverticulum is present in about 2% of asymptomatic individuals. Several complications of a diverticulum may require emergency surgical intervention. Diverticulitis may progress to perforation. Acid secretion by ectopic gastric mucosa may cause an ulcer with resultant hemorrhage—classically "brick-red" with guaiac "positive" stools—or perforation. Obstruction may be caused by intussusception of a non-tethered Meckel's diverticulum or by torsion around a persistent vascular or fibrotic band. A technetium<sup>99m</sup> pertechnetate or "Meckel's scan," is not entirely reliable but may confirm clinical suspicions.

The vitelline duct may infrequently persist as an omphalomesenteric fistula. This anomaly occurs more commonly in males than in females and usually becomes apparent within the first week of life when stool is noted from the umbilicus. A radiographic study performed through the umbilicus is diagnostic. Treatment consists of expeditious surgical resection and anastomosis. Repair should take place as soon after diagnosis as possible, usually within the first few weeks of life. Associated anomalies may be present.

**Incomplete Obliteration of the Allantois.** Complete persistence of the allantois causes a patent urachus; partial persistence may cause a urachal sinus, cyst, or diverticulum.

As with an umbilical sinus, a urachal sinus may result in intermittent purulent umbilical discharge. A sinogram is diagnostic. A sinus should be excised when diagnosed.

A urachal cyst is analogous to the omphalomesenteric cyst but is not free to undergo volvulus. Symptoms may begin at nearly any age, and there are no associated anomalies. These cysts usually present as asymptomatic or infected lower abdominal masses. Treatment consists of excision.

A bladder diverticulum may be completely asymptomatic, but should be resected if it causes recurrent cystitis.

A patent urachus is far more common in males than in females and is often recognized at birth. Urine from the umbilicus is usually the presenting sign and a fistulogram is diagnostic. Other urogenital or gastrointestinal anomalies are common and must be excluded prior to surgical repair.

**Umbilical Hernia.** Embryologically, the umbilical ring constricts and closes after the intestine has returned to the abdominal cavity; umbilical hernias are the result of an incomplete closure. A hernia usually appears during infancy but increased intraabdominal pressure due to a tumor will occasionally cause an umbilical hernia to develop later. Umbilical hernias are more common in blacks than whites. About 85% of all umbilical hernias will close without surgical repair before 4 years of age, although a proboscoid or "elephant trunk" hernia is unlikely to close. Umbilical hernias in children rarely incarcerate or rupture.

All manner of folk remedies have been employed to encourage nonoperative closure of the hernia. One of the more common practices consists of taping a coin over the defect. Not only has this been shown not to promote closure, but this may actually cause inflammation or pressure necrosis and is contraindicated. Simple observation is preferred with surgical closure if the hernia persists past 4 years of age.

**Other Umbilical Disorders.** Uncommon umbilical findings include Cullen's sign (periumbilical bruising) which usually is indicative of acute pancreatitis or retroperitoneal bleeding, and Hoffstätter's sign (bluish discoloration of an umbilical hernia) indicating intra-abdominal hemorrhage. Crohn's disease may present in ectopic intestinal mucosa within the umbilicus. Umbilical tumors are rare in children, but if primary, are usually hemangiomas or congenital arteriovenous malformations. Primary sarcoma of the umbilicus, probably arising from urachal remnants, has been found in children, but metastases from tumors in remote areas are a more common cause of malignant disease in this area. A mass due to metastatic tumor in the umbilicus is called a Sister Joseph's nodule after Dr. William Mayo's surgical assistant who first drew his attention to the finding.

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**Disclaimer:** All material is intended for informational purposes only and is not intended, and should not be used, to replace medical advice offered by a qualified physician. We are always available and willing to discuss questionable conditions with you and we invite your request for our assistance.