
The Umbilical Cord

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Normal Anatomy of the Umbilical Cord

The umbilical cord normally contains three vessels, two arteries and one vein, surrounded by a connective tissue known as "Wharton's jelly" (Figs. 11-1, 11-2, 11-3). At term, the mean length of the umbilical cord is 55 cm. A short cord is less than 35 cm in length (lower 6th percentile), and a long cord measures more than 80 cm (upper 6th percentile).³ The mean umbilical cord circumference at 40 weeks is 3.6 cm (range 2.6 to 6.0 cm).² The 90th percentile for the area of the umbilical cord at term is 1.3 cm square.⁴

Sonographic Biometry of the Umbilical Cord

There is a paucity of data concerning the biometry of the umbilical cord during gestation. The only nomogram available concerning the umbilical cord refers to the size of the umbilical vein in a free-floating cord (Fig. 11-4).¹

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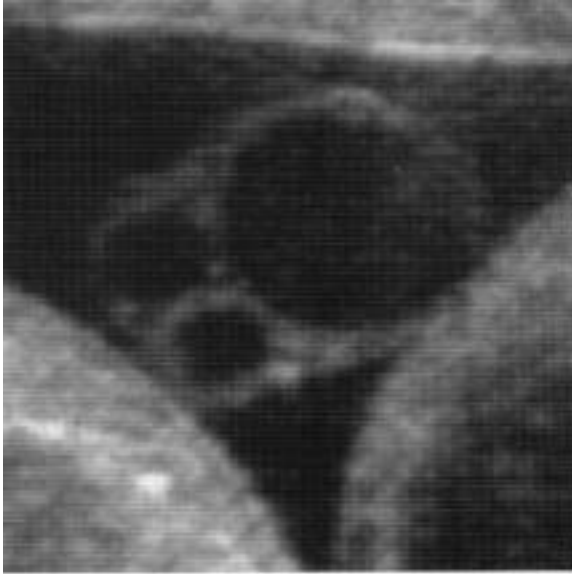


Figure 11-1. Transverse section of the umbilical cord showing two arteries and one vein.



Figure 11-3. Insertion of the umbilical cord in the placental mass. Demonstration of the insertion site is important for percutaneous umbilical cord sampling. This section excludes velamentous insertion.

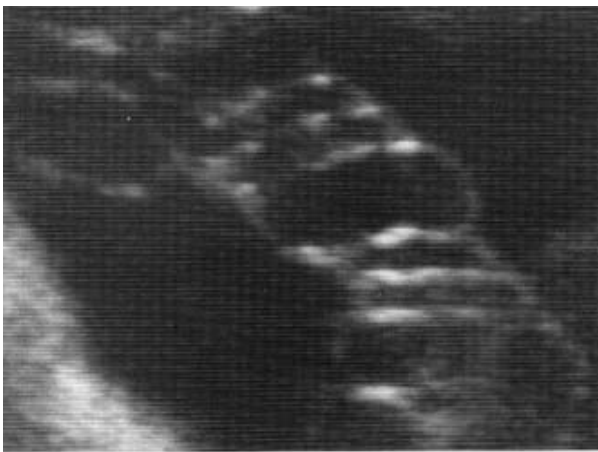


Figure 11-2. Longitudinal section of the umbilical cord showing the typical braided appearance.

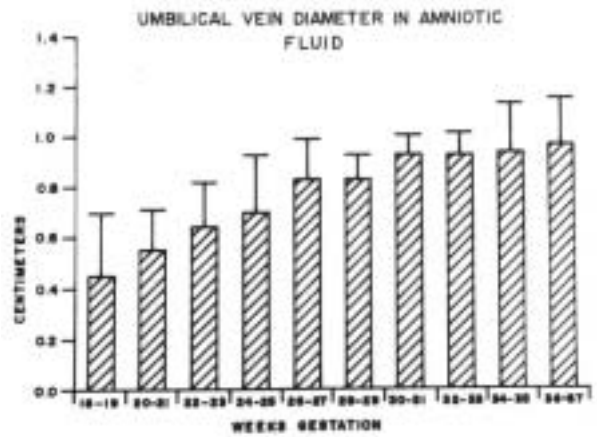


Figure 11-4. Nomogram for the umbilical vein diameter. (Reproduced with permission from DeVore et al.: *Am J Obstet Gynecol* 141:464, 1981.)

Single Umbilical Artery

Synonyms

Absence of an umbilical artery, umbilical artery agenesis, umbilical artery atrophy, and missing umbilical artery.

Definition

Single umbilical artery (SUA) is the absence of one Umbilical artery.

Epidemiology

Prospective studies indicate that SUA is present in 1 percent of all deliveries. The method of examination of the umbilical cord and the patient's race are important factors in determining the prevalence of this anomaly. For example, gross examination of the umbilical cord underestimates the prevalence.^{14,18,21} The location of the section is also important, since the two arteries may fuse close to the placental insertion of the cord,^{2,8,32} and examination at this point would overestimate the prevalence of this anomaly.^{2,22} SUA is less common in Japanese and blacks and more common in eastern Europeans.^{11,12,14,15,26,31} SUA is more common in autopsy series and in stillbirths.^{10,29} The prevalence is higher in the third trimester than in very early embryos (less than 8 weeks old),^{10,33} This suggests that a developmental atrophy of a normally formed umbilical artery may occur in some fetuses.^{28,34} The male to female ratio is

0.85:1.^{5,13,15,16,18,21,27,39} There is a greater tendency of males to be malformed in prospective series. The prevalence of SUA is three to four times higher in multiple gestations.^{4,13,15,16,21,27} There is no evidence of an epidemiologic association between maternal age, parity,^{15,21,38} month of the last menstrual period,^{7,14-16} and the prevalence of SUA.

Etiology

There is no evidence of a familial tendency of this disorder. A genetic etiology is unlikely. The increased incidence in twin gestations is not observed in monozygotic twins.¹⁴

The three theories about the pathogenesis of SUA are (1) primary agenesis of one of the umbilical arteries, (2) secondary atrophy of a previously normal artery, and (3) persistence of the original single allantoic artery of the body stalk. There is no statistical difference between atrophy and aplasia in SUA and associated malformations.¹

Prenatal Diagnosis

The normal umbilical cord contains two arteries and one vein readily visible in transverse or longitudinal sections.^{17,35} In longitudinal sections, the helicoidal shape provides a typical braided appearance to the umbilical cord. A single umbilical artery can be seen readily in transverse sections by identifying a cord

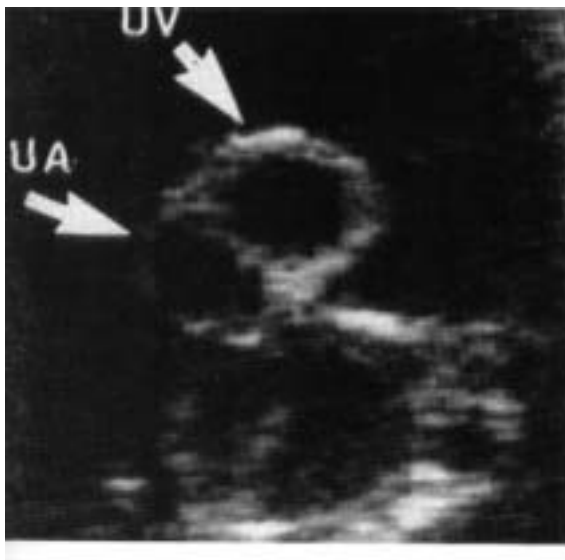


Figure 11-5. Transverse section of the umbilical cord demonstrating only two vessels. UA, umbilical artery; UV, umbilical vein.

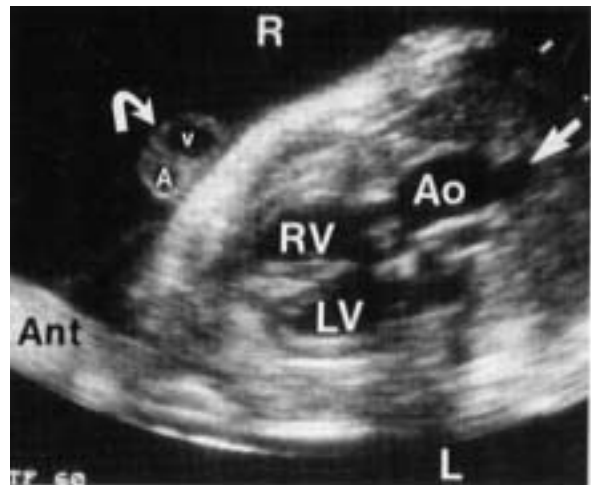


Figure 11-6. Transverse section of the fetal chest in a fetus with double outlet right ventricle. The umbilical cord is seen anterior to the chest (*curved arrow*). RV, right ventricle; LV, left ventricle; Ao, aorta; Ant, anterior; L, left; R, right; V, umbilical vein; A, single umbilical artery.

with only two vessels (Figs. 11-5, 11-6). The vein is typically larger than the artery. In longitudinal sections, a loss of the braided pattern of the umbilical cord can be visualized (Figs. 11-2, 11-3).

Identification of an SUA is an indication for a careful search for associated anomalies including echocardiography (see section on associated anomalies). These infants are also at risk for intrauterine growth retardation, and serial examinations are recommended.

Associated Anomalies

Infants with SUA have a higher prevalence of congenital anomalies, intrauterine growth retardation,^{1,4,5,13,15,26} prematurity,^{1,4,5,13,20} and a higher perinatal mortality^{1,5,13,15,25,31} than infants born with two umbilical arteries. Twenty-one percent of infants with SUA have associated anomalies in prospective series,^{1,4,5,7,11,13,15,18,22,24,26,31,36,37} and the incidence is three times higher in autopsy series.^{20,29} Heifetz has estimated that the risk of anomalies is seven times greater in infants with SUA than in control infants with two umbilical arteries.¹⁴ Table 11-1 illustrates the associated anomalies in 158 autopsy cases examined at the Armed Forces Institute of Pathology and reported by Heifetz.¹⁴ It is clear that many of these anomalies are subtle (e.g., absence of the uvula) and, therefore, nondetectable with ultrasound. In other cases, the severity of the anomaly is such (e.g., bilateral renal agenesis) that identification of SUA is practically irrelevant. The mean number of malformations per infant varies between 2 and 5.

Abnormalities that are detectable with ultrasound and are most commonly associated with SUA include cardiovascular abnormalities (particularly ventricular septal defects and conotruncal anomalies), cleft lip, ventral wall defects, esophageal atresia, spina bifida, central nervous system defects (hydrocephaly, holoprosencephaly), diaphragmatic hernia, cystic hygromas, genitourinary abnormalities (hydronephrosis, dysplastic kidneys), and digital abnormalities (polydactyly, syndactyly). All fetuses with SUA should have echocardiography performed, since cardiovascular abnormalities are among the most frequent defects.

SUA is associated with a higher incidence of marginal and velamentous insertion of the umbilical cord while these anomalies have been found in 5.9 percent and 1.2 percent of all placentas, respectively,³⁰ in SUA, their incidence is 18 percent and 9.3 percent, respectively.^{4,13,15,22,24-26} In two different series, the association of SUA with velamentous insertion of the umbilical cord slightly increased the risk for other anomalies.^{9,14} The prevalence of chromosomal abnormalities in term infants with SUA is unknown. Isolated reports have documented that

SUA can occur in association with autosomal trisomies.^{1-3,19,20,23,26,27} A recent pathologic study examining fetuses with SUA delivered before the 28th week of gestation reported an incidence of chromosomal anomalies of 67 percent (6 out of 9).⁶ This is higher than the 31 percent (24 out of 74) observed in infants born with malformations other than SUA.⁶ In this small series (n = 9), all infants with SUA and chromosomal anomalies had severe malformations. From these data, performance of amniocentesis seems justified when SUA is associated with severe anomalies.

Prognosis

The mean perinatal mortality for infants with SUA has been reported to be 20 percent.¹⁴ Two thirds of the perinatal deaths are stillbirths,^{12,15,21,37,39} and among these, three quarters have occurred antepartum and one quarter intrapartum.^{9,13} The main cause of death is the presence of associated anomalies. However, the perinatal mortality remains elevated in infants with SUA but without associated malformations.^{5,12,13} This is mainly due to prematurity and intrauterine growth retardation.

Infants with SUA are at risk for internal malformation even if external anomalies cannot be detected.^{11,25} However, if these infants remain asymptomatic during the neonatal period, their risk for lethal or serious anomalies is not higher than that of non-SUA infants.^{5,11,26,37} The long-term prognosis for growth-retarded infants with SUA is good, since these infants attain growth rates comparable to nonaffected infants.¹¹

Obstetrical Management

The detection of a single umbilical artery should prompt a search for associated anomalies. Echocardiography is indicated. Karyotype determination should be performed if associated anomalies are detected. The risk of chromosomal abnormalities in SUA without gross anomalies detected by ultrasound has not been established. Serial sonography for identification of IUGR is recommended. Intrapartum fetal heart monitoring is indicated, since some series suggest that these infants are at risk for intrapartum fetal distress and death. Pediatricians should be alerted to the diagnosis of SUA, and noninvasive techniques like neonatal ultrasound should be used freely to detect subclinical anomalies. Invasive procedures for diagnostic purposes in an otherwise asymptomatic infant do not seem justified. Data from the Collaborative Perinatal project show that infants born with SUA had a higher incidence of inguinal hernias (5.5 percent versus 1.1 percent) than a control group in a follow-up period of 4 years. The IQ of nonmalformed infants with SUA is not different from that of infants with two umbilical arteries.^{5,11,26,36}

TABLE 11-1. MALFORMATIONS IDENTIFIED IN 158 AFIP AUTOPSY CASES OF SUA

1. Urogenital Tract		4. Gastrointestinal Tract	
Renal agenesis or hypoplasia	18	Agenesis of uvula	1
Renal dysplasia or dysgenesis	30	Tracheoesophageal fistula	12
Horseshoe kidney	16	Esophageal atresia or stenosis	6
Pelvic kidney	2	Gastric atresia	1
Hydroureter and hydronephrosis	17	Duodenal atresia	1
Other ureteral anomalies	6	Midgut aplasia or stenosis	2
Urethral anomalies	12	Meckel's diverticulum	4
Persistent cloaca	7	Malrotation	10
Urachal anomalies	2	Intestinal duplication	1
Malformed or absent ext. genitalia	15	Imperforate anus	25
Testicular agenesis	3	Colorectal atresia or stenosis	3
Hypospadias	4	Liver anom./gallbladder agenesis	5
Agenesis of vas and prostate	1	Miscellaneous	1
Hydrocele	1		<u>72</u>
Agenesis of ovary and tube	4	5. Central Nervous System	
Cervical agenesis	3	Anencephaly	17
Uterine fundal anomalies	9	Meningocele, spina bifida	12
Vaginal anomalies	3	Hydrocephalus	4
	<u>153</u>	Holoprosencephaly, cebocephaly	7
2. Cardiovascular System		Microcephaly	2
Patent ductus arteriosus	21	Cerebellar anomalies	6
Patent foramen ovale	10	Microphthalmia	4
Atrial septal defect	14	Cranial nerve abnormalities	3
Ventricular septal defect	33	Miscellaneous	7
Hypoplastic left ventricle	5		<u>62</u>
Tetralogy of Fallot	2	6. Respiratory System	
Truncus anomalies	11	Choanal atresia	2
Transposition	3	Atresia nasal septum	1
Anomalous pulmonary venous return	2	Laryngeal sten./trach. agenesis	3
Coarctation	7	Pulmonary hypoplasia	26
Dextrocardia	3	Abnormal lobation	14
Dextroposition of aorta	3	Pulmonary aplasia	1
Valve anomalies	11	Diaphragmatic hernia	12
Subaortic stenosis	1		<u>59</u>
Coronary artery anomalies	4	7. Integument	
Other thoracic vessel anomalies	12	External ear abnormalities	47
	<u>142</u>	Skin tags	4
3. Musculoskeletal System		Epicanthal folds	5
High arched palate	2	Hemangiomas	2
Cleft lip and palate	27	Lymphang./cystic hygroma/web	14
Rib and sternal anomalies	12	Hydrops	2
Vertebral anomalies	15	Hypoplastic fingernails	1
Sacral agenesis	2		<u>75</u>
Amelia and phocomelia	4	8. Miscellaneous	
Other long bone anomalies	8	Endocrine gland anomalies	7
Wrist and ankle deformities	4	Accessory spleen	4
Talipes	19	Situs inversus	1
Rocker bottom feet	5	Sacrococcygeal teratoma	4
Hip dislocation	5	Pharyngeal teratoma	1
Polydactyly	12		<u>17</u>
Syndactyly or clinodactyly	6		
Other finger or toe anomalies	15	Total	753
Mandibular hypoplasia	4		
Abnormal sphenoid	1		
Achondroplasia	1		
Prune-belly	4		
Abdominal wall hernia	1		
Femoral hernia	1		
Omphalocele	21		
Gastroschisis	4		
	<u>173</u>		

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CYSTIC LESIONS OF THE UMBILICAL CORD

Omphalomesenteric Cyst

Synonym

Omphalomesenteric duct cyst.

Definition

A cystic lesion of the umbilical cord due to persistence and dilatation of a segment of the omphalomesenteric duct lined by epithelium of gastrointestinal origin.

Embryology

The omphalomesenteric duct joins the embryonic gut and the yolk sac. It is formed during the 3d week of gestation and is closed by the 16th week of gestation. Small vestigial remnants of this duct are found frequently in normal umbilical cords.²

Incidence

A total of nine cases have been described in the literature² The male to female ratio is 3:5, which is consistent with the predominance of omphalomesenteric anomalies seen in males (e.g., Meckel's

diverticula, intraabdominal mesenteric cysts, fistulas, polyps or cysts of the umbilicus, polyps or cysts of the umbilical cord).

Pathology

The cysts are generally located in close proximity to the fetus. Cysts vary in size, with the largest being 6 cm in diameter. The lining is epithelium of the gastrointestinal type (stomach, small intestine, and colon). On occasion, the surface of the cysts may have an angiomatoid appearance.²

Diagnosis

Identification of an umbilical cord cyst, which at pathology proved to be an omphalomesenteric cyst, has been reported. The cyst was identified at 20 weeks and did not change in dimensions during pregnancy (Fig. 11-7). The infant was normal. Differential diagnosis includes other hypoechoogenic lesions of the umbilical cord, like allantoid cysts, and hematomas of the umbilical cord.

Prognosis

The prognosis is excellent. One exception is a case reported by Blanc and Allan, in which acid produced by a cyst lined by gastric mucosa eroded the umbilical vein and caused fetal exsanguination.¹

Infants with other types of remnants of the omphalomesenteric duct have been found to have a 62 percent incidence of internal lesions, of which the most common is Meckel's diverticulum.⁴ Therefore, identification of this lesion should be called to the attention of parents and pediatricians. If abdominal symptoms develop later in life, Meckel's diverticulum should be considered as part of the differential diagnosis.

Obstetrical Management

The diagnosis of a cystic lesion of the umbilical cord should be an indication for serial examinations to verify normal growth of the fetus and changes in the size of the lesion. A theoretical risk is the development of vascular compression by an expanding cyst. Doppler examination may be helpful in these instances. Delivery could be recommended as soon as lung maturity is documented in patients with sizable lesions.



Figure 11-7. An omphalomesenteric cyst can be seen as a hypoechoic mass (arrow) protruding from the umbilical cord (U). (Reproduced with permission from Rosenberg, et al: *J Ultrasound Med* 5:719, 1986.)

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Allantoid Cyst

Definition

A cystic dilatation of an allantoid remnant.

Embryology

The allantoid is one of the four fetal adnexa (amnion, chorion, yolk sac, and allantoid). It becomes a fibrous band after the 3d month of gestations

Pathology

These cysts are generally located close to the fetus. They are lined by a flattened epithelium or in some areas by transitional epithelium.¹ In one case, analysis of the cystic fluid revealed a specific gravity of 1.016, BUN of 22 mg/100 ml, and no protein content.

Diagnosis

This entity appears as a cystic structure of the umbilical cord close to the fetus² (Fig. 11-8). The differential diagnosis with other cystic lesions, such as omphalomesenteric cysts and old hematomas of the umbilical cord, is not possible. Visualization of a patent urachus in the abdomen should raise the index of suspicion.

Associated Anomalies

It may be associated with a patent urachus and lower genitourinary obstruction.

Prognosis

We are aware of only one case reported in the literature.² The infant was born without complications.

Obstetrical Management

The diagnosis of a cystic lesion of the umbilical cord should be an indication for serial examinations to verify normal growth of the fetus and changes in the

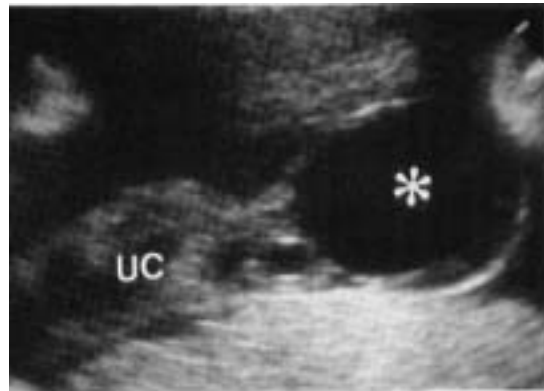


Figure 11-8. Cystic lesion of the umbilical cord corresponds to an allantoid cyst. UC, umbilical cord.

size of the lesion. A theoretical risk is the development of vascular compression by an expanding cyst. Doppler examination may be helpful in these instances. Delivery could be recommended as soon as lung maturity is documented in patients with sizable lesions.

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VASCULAR LESIONS OF THE UMBILICAL CORD

Thrombosis of the Umbilical Vessels

Definition

Occlusion of one⁶ or more³ vessels of the umbilical cord. This entity refers primarily to thrombosis of the umbilical vein, since this vessel is the only source of oxygenated blood coming from the placenta.

Incidence

Approximately 20 cases have been reported in the literature.^{6,10,16} Only 3 cases were found in an autopsy series of 4000 neonatal deaths and stillbirths weighing more than 400 g.¹² The incidence of umbilical cord thrombosis is higher in infants born to diabetic mothers (1:82) than in infants born to nondiabetic mothers (2:3918).¹² Also, the incidence of systemic thrombosis is higher in infants with diabetic mothers.

Etiology and Pathology

Thrombosis of the umbilical vessels may be a primary event¹⁶ or may be secondary to localized increased resistance in the umbilical circulation (torsion, knotting, looping, compressions, hematoma).^{2,6,7,11,13,15} Usually, the anatomic abnormality is close to the thrombus. However, in one fetus the thrombosis occurred at the opposite end of the torsion.³ Aneurysmic dilatation has also been reported in association with this disorder.^{5,8}

Other etiologic factors could be phlebitis and arteritis.¹⁰ In one documented case, the mother had rheumatoid arthritis.¹ The incidence of systemic thrombotic accidents has been reported to be higher in infants born to mothers with overt diabetes than to nondiabetic mothers (15.8 percent versus 0.8 percent).¹²

Thrombosis of the umbilical vein has been observed in association with nonimmune hydrops.^{1,4,14} Tense ascites could impair blood flow in the abdominal portion of the umbilical vein and create conditions favoring the development of thrombosis.

Diagnosis

The sonographic visualization of a thrombosed umbilical vein has been demonstrated in a report in which three fetal demises were carefully examined for thrombosis. Increased echogenicity of umbilical vessels was the diagnostic finding. In one case, this occurred in the intraabdominal portion of the umbil-

ical vein. This condition has been prenatally diagnosed in live fetuses.

Associated Anomalies

In most reports, no associated anomalies have been reported. In one patient, dysmorphic features, such as low set ears, hypertelorism, and long limbs, were reported.¹⁶ A report at variance with the literature is that of Konstantinova, who cited eight patients with varices and thrombosis of the umbilical cord having severe associated anomalies (hydrocephaly, anencephaly, trisomy 21, renal agenesis, spina bifida, cor triloculare, and phocomelia).⁹

Prognosis

There are only four infants who have survived thrombosis of the umbilical vein.^{8,10,13,16} The prognosis in all other patients is understandably poor.

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Hemangiomas of the Umbilical Cord

Synonyms

Angiomyxomas of the umbilical cord, cavernous hemangioma, hemangiofibromyxoma, myxoangioma, and telangioectasic myxosarcoma.

Definition

Hemangioma of the umbilical cord is a tumor arising from the endothelial cells of the vessels of the umbilical cord.

Incidence

After excluding those cases in which the tumor does not arise along the umbilical cord (placental hemangiomas) or lacks an endothelial component (representing, therefore, hematomas), only 18 cases are reported in the literature.^{3-11,13}

Etiology and Pathology

The lesions can measure up to 15 cm. They consist of an angiomatous nodule surrounded by edema and myxomatous degeneration of the Wharton's jelly. The tumor is more frequently located toward the placental end of the umbilical cord. The sites of origin are the main vessels of the umbilical cord, in order of frequency: arteries, veins, and vitelline capillaries. The lesions may involve more than one vessel. The typical microscopic appearance is that of multiple channels lined by benign endothelium accompanied by edema and myxoid degeneration of the stroma of the cord. The differential diagnosis between hemangiomas and hematomas of the umbilical cord is provided by the presence in the former of capillaries lined by endothelium and showing a positive immunoperoxidase staining for factor VIII-related antigen.

Associated Anomalies

This entity has been associated with nonimmune hydrops.^{2,11,12} In one case, there was an association between a hemangioma of the umbilical cord and severe diffuse skin hemangiomas.¹¹ In placental hemangiomas, the incidence of associated vascular

neoplasms is 10 percent. Although the same risk has not been reported with umbilical cord hemangiomas, it is possible that these infants are at a greater risk.⁸

Diagnosis

We have made a prenatal diagnosis of this tumor in a 34-week-old fetus. The lesion appeared as a hyperechogenic mass, extending from the fetal umbilicus for a distance of about 5 cm. The cord appeared edematous for most of its length, and a localized collection of edematous fluid (pseudocyst) could be seen close to the nodule (Fig. 11-9). Three umbilical vessels were present with normal waveform at Doppler examination.

A hemangioma of the umbilical cord appears as a hyperechogenic mass. Differential diagnosis for this type of lesion includes teratomas and an umbilical cord hematoma (see respective sections in this chapter). Associated localized edema of the umbilical cord has been noted in the two cases identified prenatally with ultrasound. In one case, the solid component of the lesion was not recognized because of its small size.¹¹ The mechanism responsible for the pseudocyst formation is unknown. It has been suggested that it results from transudation of fluid. A precise diagnosis of hemangioma of the umbilical cord may not be possible.

An umbilical cord hemangioma has been associated with elevated alphafetoprotein.¹ However, in our case, maternal serum alphafetoprotein determination in midtrimester was normal.

Prognosis

Data are limited because of the rarity of the disease. The development of hydrops would logically suggest compromise. Increased morbidity (12 of 18) and mortality (7 of 18) have been observed.^{3,4} It is unclear whether this represents a truly poor prognosis or detection biases. The excessive mortality is due to other seemingly unrelated complications, such as

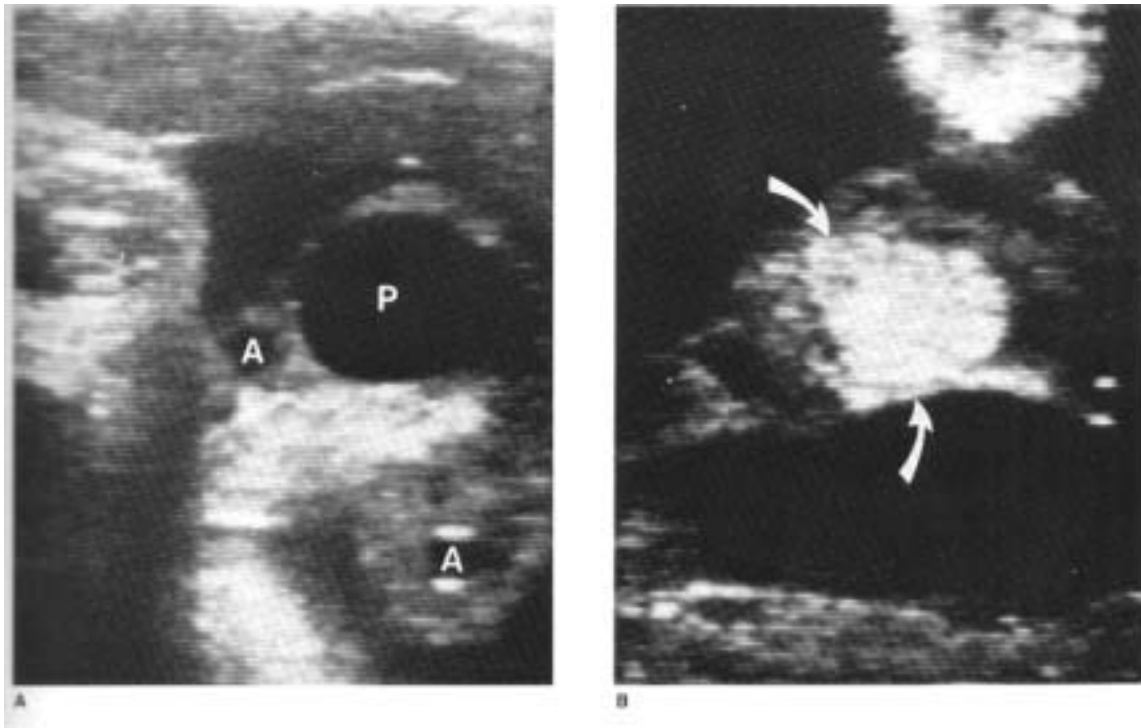


Figure 11-9. **A.** Umbilical cord close to the level of insertion into the fetal abdomen. A hyperechogenic nodule is clearly visible within the cord. A, umbilical arteries, P, pseudocyst due to localized accumulation of fluid within the Wharton's jelly. **B.** Loop of cord free floating. The dense nodule is visible, surrounded by edema of the Wharton's jelly.

prematurity, postmaturity, birth trauma, preeclampsia or short umbilical cord. The mechanism by which this tumor causes fetal compromise is probably mechanical impairment of the fetal circulation.

Obstetrical Management

The diagnosis of a lesion of the umbilical cord should probably be an indication for serial examinations to verify normal growth of the fetus, changes in the size of the lesion and signs of non-immune hydrops. A theoretical risk is the development of vascular compression by an expanding lesion. Doppler examination may be helpful in these instances. Delivery could be recommended as soon as lung maturity is documented in patients with sizable lesions.

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Hematoma of the Umbilical Cord

Definition

Extravasation of blood into the Wharton's jelly.

Incidence

Between 1 in 5505⁵ and 1 in 12,699.⁴

Etiology and Pathology

There is no adequate explanation for this phenomenon. A traumatic insult (torsion, loops, knots, traction, and prolapse) in an area of local weakness of the vessel wall has been advocated by some.^{3,5,14} With the use of invasive techniques of prenatal diagnosis, such as fetoscopy and percutaneous umbilical cord puncture, it is possible that this entity occurs iatrogenically.¹² The dimensions of the reported hematomas have ranged from 1 to 4 cm, and the length has been up to 42 cm. The most frequently involved vessel is the umbilical vein, with a vein: artery ratio of 9:1.⁵ The vitelline vessels could also be involved. In 11 of 61 cases, the hematoma was multiple.^{2,5} The most common site was near the fetal insertion of the cord^{3,6,14} but hematomas have been reported also in its central portion.^{7,9} A serious complication is the rupture of the hematoma into the amniotic cavity, since this may lead to exsanguination.² Another complication, reported by Fletcher,⁷ is neonatal myocardial infarction attributed to embolization of fragments released from the hematoma.

Diagnosis

Prenatal identification of this condition has been reported twice (Fig. 11-10).^{13,15} The first case pre-

sented as a 6- by 8-cm hypoechogenic septated mass.¹³ The hypoechogenic nature of the mass suggests that the clot was old, because a fresh clot is expected to be hyperechogenic. The second case showed a hyperechogenic lesion, which was recognized after an initially bloody amniocentesis.¹⁵ The differential diagnosis between this entity and other masses of the umbilical cord is difficult. Hematomas may be more irregular than other cystic lesions.

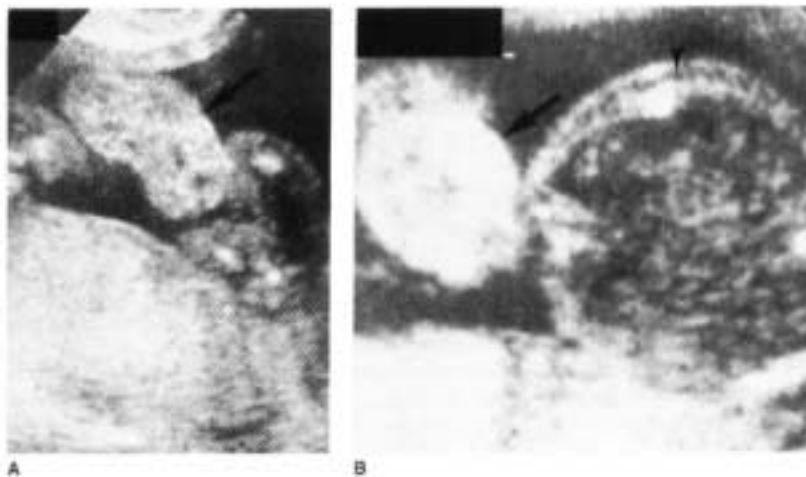
Prognosis

The fetal mortality reported by Dippel is 47 percent.⁵ The overall perinatal mortality in all reported cases is 52 percent (26 of 50).^{1,3,5,7,8-11,13,14} Death is caused mainly by exsanguination and compression of the vessels.

Obstetrical Management

If a hematoma is suspected in a fetus, an amniocentesis should be performed to establish lung maturity. In the presence of a mature fetus, there would be little advantage to prolonging the pregnancy. The optimal mode of delivery has not been established. However, 10 of 17 fetal deaths have occurred during labor. It is not known whether the accident occurs predominantly during labor or whether labor per se is dangerous for the infant with an umbilical cord hematoma. Doppler examination may provide a noninvasive means of examining umbilical vascular resistance.

Figure 11-10. **A.** Sausage-shaped umbilical cord hematoma (*arrow*) is hyperechoic and markedly thickened. **B.** Cross-section of umbilical cord hematoma (*arrow*) adjacent to but not contiguous with the fetal abdomen at the level of the intrahepatic umbilical vein. (Reproduced with permission from *Sutro et al.*: *AJR* 142:802, 1984.)



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OTHER PATHOLOGIC CONDITIONS OF THE UMBILICAL CORD

Strictures or Coarctation of the Umbilical Cord

Synonyms

Constriction of the umbilical cord,¹¹umbilical cord occlusions and fibrosis circumscripta of the umbilical cord.⁶

Definition

Coarctation is characterized by a localized narrowing of the cord with disappearance of the Wharton's jelly, thickening of the vascular walls, and narrowing of their lumens.¹⁰ Generally, torsion of the umbilical cord is present.⁹

Incidence

Approximately 30 cases have been described in the literature. However, this may not represent the real incidence of this entity. Tavares-Fortuna and Lourdes-Pratas conducted a prospective study and reported an incidence of 1 in 250 deliveries.¹⁰

Etiology and Pathology

The mechanisms responsible for coarctation of the umbilical cord are not understood. Edmonds' suggested that coarctation and subsequent torsion are a

postmortem event caused by necrobiosis of the Wharton's jelly. Weber¹¹ pointed out the following objections to this hypothesis: (1) liveborn fetuses have been reported to have constrictions and (2) coarctations are extremely rare in stillbirths. Perhaps this complication should be viewed as a local failure of Wharton's jelly development that creates a weak point in the umbilical cord. Fetal motion could lead to torsion around this point.

The site of the torsion is generally close to the fetus.³ A localized edematous area is frequently reported distal to the torsion point.⁸ Multiple strictures along the length of the umbilical cord can be found.^{4,10}

Diagnosis

To date, this condition has not been diagnosed with ultrasound, although recognition should be possible.

Associated Anomalies

The following anomalies have been reported with coarctation of the umbilical cord: type C tracheoesophageal fistula, cleft lip,⁵ anencephaly, anoph-

thalmia and exophthalmos,⁶ polyhydramnios, ventricular septal defect and trisomy 18,² and generalized subcutaneous edema.⁷

Prognosis

Most reported cases of coarctation result in stillbirths. However, this may represent reporting biases. Recurrences in subsequent pregnancies have not been noted in the literature.^{3,11}

Obstetrical Management

Should this diagnosis be made, serial monitoring of preterm infants would be indicated. Delivery as soon as there is a reasonable chance for survival would seem logical.

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Teratomas

Definition

A germ cell tumor containing elements from the three germinal layers.

Incidence

Four cases have been reported in the literature.²⁻⁵

Embryology

Germ cells arise in the dorsal wall of the yolk sac; from there they migrate to the genital ridge. During the course of this process, germ cells are found in the wall of the primitive gut. Since in early pregnancy, the primitive gut sends an evagination into the umbilical cord, germ cells may reach the cord at this stage.¹

It is possible that some teratomas are indeed acardiac twins, and viceversa.⁴

Pathology

This tumor has been reported to measure up to 9 cm in diameter. Histologically, it shows elements originating from the three germinal layers, and it may undergo calcification. Teratomas are found at any point along the length of the umbilical cord. The differential diagnosis with an acardiac twin is based on the following: (1) the acardiac twin has a separate,

although rudimentary, umbilical cord, (2) the twin cannot be entirely intrafunicular, and (3) the twin shows some evidence of body organization (e.g., cranium, spine) whereas teratomas are completely disorganized.

Diagnosis

An umbilical cord teratoma has not been diagnosed prenatally. The entity must be considered as part of the differential diagnosis, of a complex lesion of the umbilical cord.

Prognosis

In one of the three available cases for review, the fetus died.⁴

Obstetrical Management

Diagnosis of a lesion of the umbilical cord should probably be an indication for serial examinations to verify normal growth of the fetus, changes in the size of the lesion, and development of nonimmune hydrops. A theoretical risk is the development of vascular compression by an expanding lesion. Doppler examination may be helpful in these instances. Delivery could be recommended as soon as lung

maturity is documented in patients with sizable lesions.

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True Knots of the Umbilical Cord

Definition

Knots of the umbilical cord are classified as false or true. The former consist of simple dilatations of the umbilical vessels that look like knots but are considered devoid of any clinical significance. True knots are formed when the fetus passes through a loop of umbilical cord.

Incidence

In recent prospective series, it varied from 0.04 to 1 percent.²⁻⁷

Etiology and Pathology

Factors predisposing to true knots are commonly believed to include monoamniotic twins, long cords, and polyhydramnios. It seems logical that knots are formed in early pregnancy, when it is possible for the fetus to go through a loop of umbilical cord. Tightening of the knot can occur during labor. Knots can be single or multiple.

Diagnosis

This diagnosis is feasible with ultrasound. We have seen one fetus with isolated ascites who later proved to have a true knot of the umbilical cord.

Associated Anomalies

A recent study has suggested that infants with true knots of the umbilical cord have an increased incidence of congenital anomalies.¹

Prognosis

Perinatal mortality varies from 8 to 11 percent.^{4,7}

Obstetrical Management

A cesarean section would seem the most logical approach if a diagnosis is made.

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Cord Presentation

Synonym

Occult prolapse.

Definition

Prolapse refers to protrusion of the umbilical cord through the cervix into the vagina. Occult prolapse occurs when the cord lies alongside the presenting part. Cord presentation is defined as a cord lying between the presenting part and the lower pole of the intact membranes.

Incidence

The mean incidence of cord prolapse is 1 in 200 births. The frequency of cord presentation (diagnosed with ultrasound) is 0.61 percent.¹

Etiology

Predisposing factors include abnormal presentation, prematurity, polyhydramnios, unengaged present-

ing part, and long umbilical cord.² Prolapse occurs after either artificial or spontaneous rupture of the membranes.

Diagnosis

The diagnosis is easily made by demonstrating loops of umbilical cord in the lower segment below the presenting part (Fig. 11-11).

Prognosis

Lange et al.¹ have conducted a prospective study demonstrating that 9 of 1471 (0.61 percent) patients had cord presentation. This diagnosis was based on the demonstration of loops of umbilical cord in the lower uterine segment below the presenting part. Of the nine patients diagnosed to have cord presentation, seven were delivered by cesarean section and two vaginally. In four of the seven cases delivered by cesarean section, a cord presentation was found. In the other three it was suspected. Of the two vaginal deliveries, one was a stillbirth associated with a cord prolapse and the other underwent a spontaneous version with resolution of cord presentation.

Obstetrical Management

In patients at risk for cord presentation (malpresentation, polyhydramnios), a specific evaluation of the position of the cord should be performed. If a cord presentation is found in a term infant, admission to the hospital for delivery is recommended. Delivery can be accomplished by cesarean section. If the diagnosis is made in a viable but preterm gestation, expectant management is the most prudent approach. Serial examinations are recommended.

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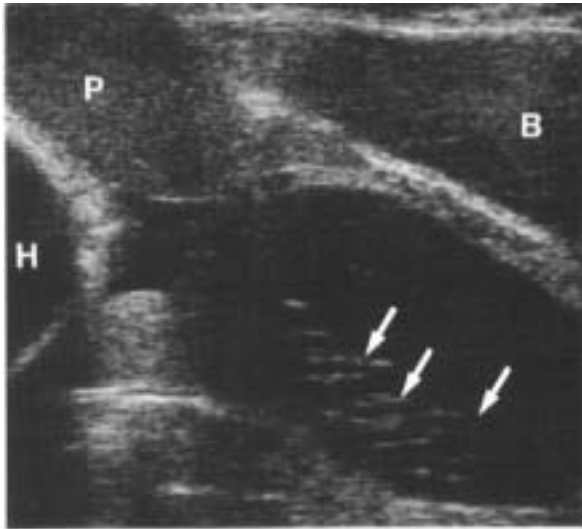


Figure 11-11. Longitudinal scan of the pelvis in a patient with preterm labor at 27 weeks. Note the multiple loops of umbilical cord (arrows), which are in front of the fetal head. B, bladder; H, head; P, placenta.

Velamentous Insertion

Definition

The term "velamentous insertion" refers to attachment of the cord to the membranes rather than to the placental mass. Marginal insertions refer to implantation of the cord into the edge of the placenta.

Incidence

The incidence is 0.09 to 1.8 percent.^{5,6,11} This condition is more common in multiple pregnancies.

Etiology

Benirschke and Driscoll¹ favor the concept of trophotropism. This view proposes that velamentous insertion occurs when most of the placental tissue grows laterally, leaving the initially centrally located umbilical cord in an area that will become atrophic. An alternative hypothesis suggests that there is a primary defect in the implantation of cord that occurs in a site of trophoblast in front of the decidua capsularis instead of the area of trophoblast that forms the placental mass.⁹

Pathology

Implantation of the umbilical vessels occurs in the chorion laeve, and therefore, the umbilical vessels lie on the membrane surface.

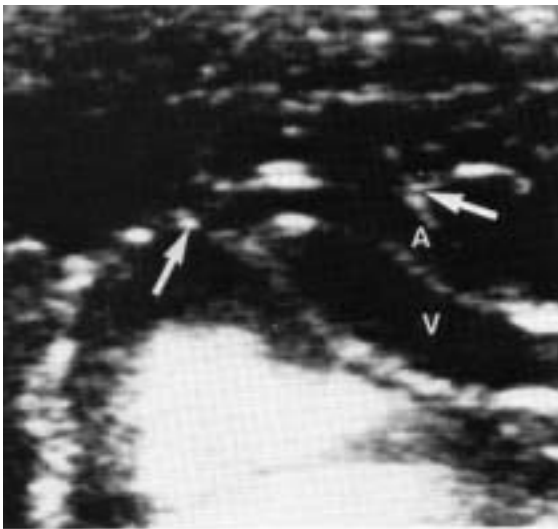


Figure 11-12. Velamentous insertion (arrows) of the umbilical cord on the amnion. A, umbilical artery, V, umbilical vein.

Associated Anomalies

The incidence of associated anomalies is increased and varies from 5.9 percent³ to 8.5 percent.⁷ The reported anomalies have included esophageal atresia, obstructive uropathies, congenital hip dislocation, asymmetrical head shape, spina bifida, ventricular septal defects, cleft palate, and trisomy 21 (one case). Bilobated placenta has also been found in association with the velamentous insertion of the umbilical cord. The birth weight of infants with velamentous insertion is lower than that of a control group even when malformed infants are excluded (3098 g, SD = 765, versus 3416 g, SD = 712).^{2,4} The incidence of intrauterine growth retardation (defined as 2 SD below the mean for gestational age) is 7.5 percent,^{2,8} and the incidence of premature births is 17.2 percent.² In twin gestations, the twin with velamentous insertion has a lower birth weight than the unaffected twin.²

Diagnosis

This diagnosis has been made several times in our institution. The critical point is establishing the relationship between the insertion of the umbilical cord and the placental mass (Fig. 11-12).

Prognosis

Infants with velamentous insertion are at increased risk for IUGR, preterm birth, and congenital anomalies.⁶ Data collected in Norway between 1969 and 1981 showed that infants with low birth weight and velamentous insertion had some difficulty in attaining normal weight and length.³ There were also some anomalies detected in childhood that were missed at birth, including esophagobronchial fistula, tetralogy of Fallot, obstructive uropathy, osteogenesis imperfecta, and muscle dystrophy.³

Obstetrical Management

If this diagnosis is made, a careful examination for associated anomalies is required. It is important to demonstrate a stomach, since esophageal atresia has been the most common anomaly detected in a large series (4 of 305). Echocardiography should be included. Serial sonographic examinations are indicated to exclude IUGR. Labor is a critical period of time because the umbilical vessels could be ruptured (vasa previa). For velamentous insertions located in the uterine fundus, no change in standard obstetrical management seems to be required. If the velamentous insertion is located in the lower uterine segment,

an elective section to avoid rupture of a vasa previa may be considered.

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