CHAPTER 6: Placental Abnormalities

The placenta, as a rule, presents more or less rounded outlines, but now and again when inserted in the neighbourhood of the internal os it may take on a horseshoe-like appearance, its two branches running partially around the orifice.

—J. Whitridge Williams (1903)

INTRODUCTION

The placenta is a fantastic organ in its own right. As discussed in Chapter 5 (Blastocyst), it provides the indispensable interface between mother and fetus. Indeed, placental anatomy, physiology, and molecular structure remain some of the most intriguing and understudied topics in obstetrics.

Although a placental examination by the obstetrician is recommended, by consensus, routine pathological examination is not mandatory. Indeed, specific conditions that merit submission for detailed inspection are still debated. By way of example, the College of American Pathologists has recommended placental examination for an extensive list of indications (Langston, 1997). Data, however, are insufficient to support all of these. At minimum, the placenta and cord should be inspected in the delivery room. The decision to request pathological examination should be based on clinical and placental findings (Redline, 2008; Roberts, 2008). Listed in Table 6-1 are some of the indications at Parkland Hospital for placental anatomical and histopathological examination.

TABLE 6-1

Some Indications for Placental Pathological Examination[a]

Maternal Indications
Abruption
Antepartum infection with fetal risks
Anti-CDE alloimmunization
Cesarean hysterectomy
Oligohydramnios or hydramnios
Peripartum fever or infection
Preterm delivery
Postterm delivery
Severe trauma
Suspected placental injury
Systemic disorders with known effects
Thick or viscid meconium
Unexplained late pregnancy bleeding
Unexplained or recurrent pregnancy complications
Fetal and Neonatal Indications
Admission to an acute care nursery
Birth weight ≤10th or ≥95th percentile
Fetal anemia
Fetal or neonatal compromise
Neonatal seizures
Hydrops fetalis
Infection or sepsis
Major anomalies or abnormal karyotype
Multifetal gestation
Stillbirth or neonatal death
Vanishing twin beyond the first trimester
Placental Indications
Gross lesions
Marginal or velamentous cord insertion
Markedly abnormal placental shape or size
Markedly adhered placenta
Term cord <32 cm or >100 cm
Umbilical cord lesions

[a]Indications are organized alphabetically.

NORMAL PLACENTA

At term, the typical placenta weighs 470 g, is round to oval with a 22-cm diameter, and has a central thickness of 2.5 cm (Benirschke, 2012). It is composed of a placental disc, extraplacental membranes, and three-vessel umbilical cord. The disc surface that lies against the uterine wall is the *basal plate*, which is divided by clefts into portions—termed cotyledons. The fetal surface is the *chorionic plate*, into which the umbilical cord inserts, typically in the center. Large fetal vessels that originate from the cord vessels then spread and branch across the chorionic plate before entering stem villi of the placenta parenchyma. In tracing these, fetal arteries almost invariably cross over veins. The chorionic plate and its vessels are covered by thin amnion, which can be easily peeled away from a postdelivery specimen.

As recommended by the American Institute of Ultrasound in Medicine (2013), placental location and relationship to the internal cervical os are recorded during prenatal sonographic examinations. As visualized ultrasonically, the normal placenta is homogenous and 2 to 4 cm thick, lies against the myometrium, and indents into the amnionic sac. The retroplacental space is a hypoechoic area that separates the myometrium from the basal plate and measures less than 1 to 2 cm. The umbilical cord is also imaged, its fetal and placental insertion sites examined, and its vessels counted.

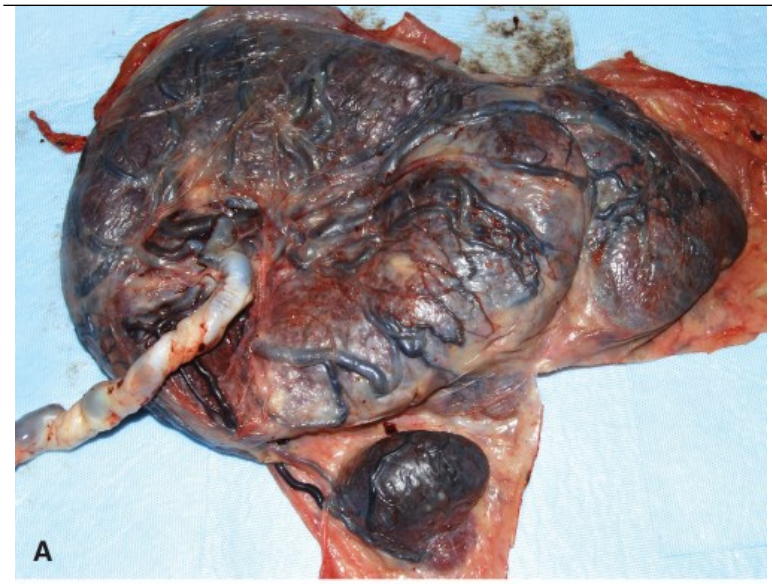
Many placental lesions can be identified grossly or sonographically, but other abnormalities require histopathological examination for clarification. A detailed description of these is beyond the scope of this chapter, and interested readers are referred to textbooks by Benirschke (2012), Fox (2007), and Faye-Petersen (2006) and their colleagues. Moreover, the placenta accrete syndrome and gestational trophoblastic disease are presented in detail in Chapters 20 and 41, respectively.

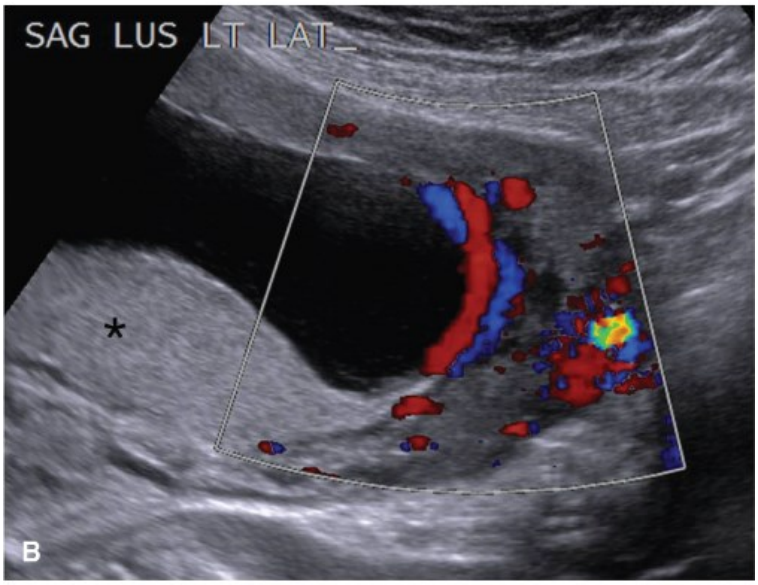
SHAPE AND SIZE VARIANTS

Of variants, placentas may infrequently form as separate, nearly equally sized discs. This *bilobate placenta* may also be called bipartite placenta or placenta duplex. In these, the cord inserts between the two placental lobes—either into a connecting chorionic bridge or into intervening membranes. A placenta containing three or more equivalently sized lobes is rare and termed *multilobate*. Unlike this equal distribution, one or more disparately smaller accessory lobes—*succenturiate lobes*—may develop in the membranes at a distance from the main placenta (Fig. 6-1). These lobes have vessels that course through the membranes. Of clinical importance, if these vessels overlie the cervix to create a vasa previa, dangerous fetal hemorrhage can follow vessel laceration (Remnants and Cysts). An accessory lobe can also be retained in the uterus after delivery to cause postpartum uterine atony and hemorrhage or later endometritis.

FIGURE 6-1

Succenturiate lobe. **A.** Vessels extend from the main placental disc to supply the small round succenturiate lobe located beneath it. (Used with permission from Dr. Jaya George.) **B.** Sonographic imaging with color Doppler shows the main placental disc implanted posteriorly (*asterisk*). The succenturiate lobe is located on the anterior uterine wall across the amnionic cavity. Vessels are identified as the long red and blue crossing tubular structures that travel within the membranes to connect these two portions of placenta.





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Rarely, the placental surface area varies from the norm. With placenta membranacea, villi cover all or nearly all the uterine cavity. This may occasionally give rise to serious hemorrhage because of associated placenta previa or accreta (Greenberg, 1991; Pereira, 2013). A ring-shaped placenta may be a variant of placenta membranacea. This placenta is annular, and a partial or complete ring of placental tissue is present. These abnormalities appear to be associated with a greater likelihood of antepartum and postpartum bleeding and fetal-growth restriction (Faye-Petersen, 2006; Steemers, 1995). With placenta fenestrata, the central portion of a placental disc is missing. In some instances, there is an actual hole in the placenta, but more often, the defect involves only villous tissue, and the chorionic plate remains intact. Clinically, it may erroneously prompt a search for a retained placental cotyledon.

During pregnancy, the normal placenta increases its thickness at a rate of approximately 1 mm per week. Although not measured as a component of routine sonographic evaluation, this thickness typically does not exceed 40 mm (Hoddick, 1985). *Placentomegaly* defines those thicker than 40 mm and commonly results from striking villous enlargement. This may be secondary to maternal diabetes or severe maternal anemia, or to fetal hydrops, anemia, or infection caused by syphilis, toxoplasmosis, parvovirus, or cytomegalovirus. In these conditions, the placenta is homogeneously thickened. Less commonly with placentomegaly, fetal parts are present, but villi are edematous and appear as small placental cysts, such as in cases of partial mole (Chap. 20, Diagnosis). Cystic vesicles are also seen with *placental mesenchymal dysplasia*. Vesicles in this rare condition correspond to enlarged

stem villi, but unlike molar pregnancy, trophoblast proliferation is not excessive (Woo, 2011).

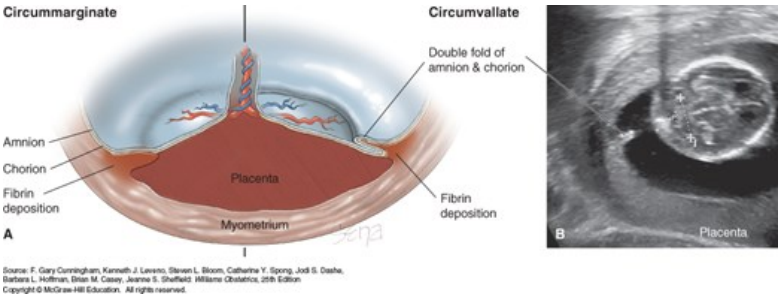
Rather than villous enlargement, placentomegaly often may result from collections of blood or fibrin, which impart heterogeneity to the placenta. Examples of this are discussed in Maternal Blood Flow Disruption and include massive perivillous fibrin deposition, intervillous or subchorionic thromboses, and large retroplacental hematomas.

EXTRACHORIAL PLACENTATION

The chorionic plate normally extends to the periphery of the placenta and has a diameter similar to that of the basal plate. With extrachorial placentation, however, the chorionic plate fails to extend to this periphery and leads to a chorionic plate that is smaller than the basal plate (Fig. 6-2). Circummarginate and circumvallate placentas are the two types. In a *circummarginate placenta*, fibrin and old hemorrhage lie between the placenta and the overlying sheer amniochorion. In contrast, with a *circumvallate placenta*, the chorion periphery is a thickened, opaque, gray-white circular ridge composed of a double fold of chorion and amnion. Sonographically, the double fold can be seen as a thick, linear band of echoes extending from one placental edge to the other. On cross section, however, it appears as two "shelves," with each lying above an opposing placental margin (see Fig. 6-2). This anatomy can help differentiate this shelf from amnionic bands and amnionic sheets, which are described in Amniochorion.

FIGURE 6-2

A. In this illustration, circummarginate (*left*) and circumvallate (*right*) varieties of extrachorial placentation are shown. A circummarginate placenta is covered by a single layer of amniochorion. **B.** This transabdominal gray-scale sonographic image shows a circumvallate placenta. The double fold of amnion and chorion creates a broad, opaque white ring and ridge on the fetal surface.



In relatively small observational studies of circumvallate placenta diagnosed postpartum, it was associated with increased risk for antepartum bleeding, abruption, fetal demise, and preterm birth (Lademacher, 1981; Suzuki, 2008; Taniguchi, 2014). In a prospective sonographic investigation of 17 cases, however, Shen and associates (2007a) found most circumvallate placentas to be transient. Persistent cases were benign. In general, most otherwise uncomplicated pregnancies with either type of extrachorial placentation have normal outcomes, and no increased surveillance is usually required.

CIRCULATORY DISTURBANCES

Functionally, placental perfusion disorders can be grouped into: (1) those in which maternal blood flow to or within the intervillous space is disrupted, and (2) those with disturbed fetal blood flow through the villi. These lesions are frequently identified in the normal, mature placenta. Although they can limit maximal placental blood flow, functional reserve within the placental prevents harm in most cases. Indeed, some estimate that up to 30 percent of placental villi can be lost without untoward fetal effects (Fox, 2007). If extensive, however, these lesions can profoundly limit fetal growth.

Lesions that disrupt perfusion are frequently seen grossly or sonographically, whereas smaller lesions are seen only histologically. With sonography, many of these, such as subchorionic fibrin deposition, perivillous fibrin deposition, and intervillous thrombosis, appear as focal sonolucencies within the placenta. Importantly, in the absence of maternal or fetal complications, isolated placental sonolucencies are considered incidental findings.

Maternal Blood Flow Disruption

Subchorionic Fibrin Deposition

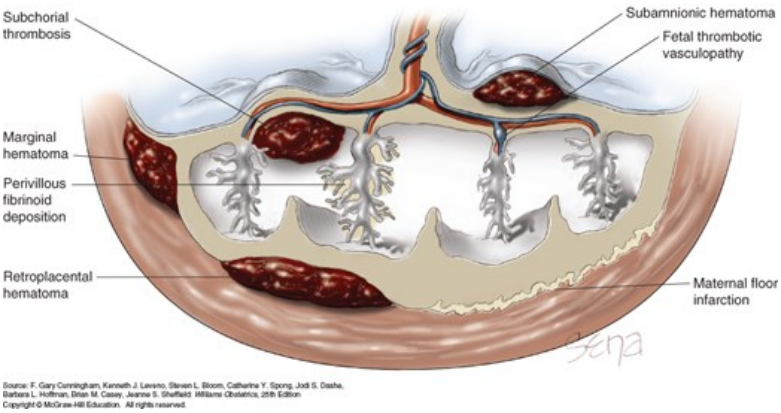
These collections are caused by slowing of maternal blood flow within the intervillous space. In the portion of this space near the chorionic plate, blood stasis is prominent and leads to subsequent fibrin deposition. In viewing the placental fetal surface, subchorionic lesions are commonly seen as white or yellow, firm, round, elevated plaques just beneath the chorionic plate.

Perivillous Fibrin Deposition

Stasis of maternal blood flow around an individual villus also results in fibrin deposition and can lead to diminished villous oxygenation and necrosis of syncytiotrophoblast (Fig. 6-3). These small yellow-white placental nodules are grossly visible within the parenchyma of a sectioned placenta. Within limits, these reflect normal placental aging.

FIGURE 6-3

Potential sites of maternally and fetally related placental circulatory disturbances. (Adapted from Faye-Petersen, 2006.)



Maternal Floor Infarction

This extreme variant of perivillous fibrin deposition is a dense fibrinoid layer within the placental basal plate and is erroneously termed an infarction. *Maternal floor infarction* has a thick, yellow or white, firm corrugated surface that impedes normal maternal blood flow into the intervillous space. In specific cases that extend beyond the basal plate to entrap villi and obliterate the intervillous space, the term *massive perivillous fibrin deposition* is used. The etiopathogenesis is unclear, but maternal auto- or alloimmunity appears contributory (Faye-Peterson, 2017; Romero, 2013). Antiphospholipid antibody syndrome and angiogenic factors involved with preeclampsia have also been implicated (Sebire, 2002, 2003; Whitten, 2013).

These lesions are not reliably imaged with prenatal sonography, but they may create a thicker basal plate. Affected pregnancies are associated with miscarriage, fetal-growth restriction, preterm delivery, and stillbirth (Andres, 1990; Mandsager, 1994). Importantly, these adverse outcomes can recur in subsequent pregnancies.

Intervillous Thrombus

This is a collection of coagulated maternal blood normally found in the intervillous space mixed with fetal blood from a break in a villus. Grossly, these round or oval collections vary in size up to several centimeters. They appear red if recent or white-yellow if older, and they develop at any placental depth. Intervillous thrombi are common and typically not associated with adverse fetal sequelae. Because there is potential for a communication between maternal and fetal circulations, large lesions can cause elevated maternal serum alpha-fetoprotein levels (Salafia, 1988).

Infarction

Chorionic villi themselves receive oxygen solely from maternal circulation supplied to the intervillous space. Any uteroplacental disease that diminishes or obstructs this supply can result in infarction of an individual villus. These are common lesions in mature placentas and are benign in limited numbers. If numerous, however, placental insufficiency can develop. When they are thick, centrally located, and randomly distributed, they

may be associated with preeclampsia or lupus anticoagulant.

Hematoma

As depicted in Figure 6-3, the maternal-placental-fetal unit can develop several hematoma types. These include: (1) retroplacental hematoma—between the placenta and its adjacent decidua; (2) marginal hematoma—between the chorion and decidua at the placental periphery—known clinically as subchorionic hemorrhage; (3) subamnionic hematoma—these are of fetal vessel origin and found beneath the amnion but above the chorionic plate, and (4) subchorial thrombus along the roof of the intervillous space and beneath the chorionic plate. With this last type, *massive subchorionic hematomas* are also known as a *Breus mole.*

Sonographically, hematomas evolve with time and appear hyperechoic to isoechoic in the first week after hemorrhage, hypoechoic at 1 to 2 weeks, and finally, anechoic after 2 weeks. Most subchorionic hematomas visible sonographically are fairly small and of no clinical consequence. However, extensive retroplacental, marginal, and subchorial collections have been associated with higher rates of miscarriage, stillbirth, placental abruption, and preterm delivery (Ball, 1996; Fung, 2010; Madu, 2006; Tuuli, 2011). In essence, placental abruption is a large, clinically significant retroplacental hematoma.

Fetal Blood Flow Disruption

Fetal Thrombotic Vasculopathy

Placental lesions that arise from fetal circulatory disturbances are also depicted in Figure 6-3. Deoxygenated fetal blood flows from the two umbilical arteries into arteries within the chorionic plate that divide and send branches out across the placental surface. These eventually supply individual stem villi, and their thrombosis will obstruct fetal blood flow. Distal to the obstruction, affected portions of the villus become nonfunctional. Thrombi in limited numbers are normally found in mature placentas. If many villi are affected, which can be seen with preeclampsia, the fetus may suffer growth restriction, stillbirth, or nonreassuring fetal heart rate patterns (Chisholm, 2015; Lepais, 2014; Saleemuddin, 2010).

Villous Vascular Lesions

There is a spectrum of villous capillary lesions. *Chorangiosis* describes an increased number of capillaries within terminal villi. Its definition requires ≥10 capillaries to be present in ≥10 villi in ≥10 fields viewed through a 10× microscope lens (Altshuler, 1984). Clinically, long-standing hypoperfusion or hypoxia is thought to be causative (Stanek, 2016). It is often associated with maternal diabetes mellitus (Ogino, 2000). *Chorangiomatosis* describes increased capillary number in stem villi, but terminal villi are spared. This finding has been linked with fetal-growth restriction and anomalies (Bagby, 2011). Despite these associations, the clinical significance of both vascular conditions remains unclear. *Chorioangiomas* are described subsequently.

Subamnionic Hematoma

As indicated earlier, these hematomas lie between the chorionic plate and amnion. They most often are acute events during third-stage labor when cord traction ruptures a vessel near the cord insertion.

Large chronic antepartum lesions may cause fetomaternal hemorrhage or fetal-growth restriction (Deans, 1998). They also may be confused with other placental masses such as chorioangioma. In most cases, Doppler interrogation will show absent internal blood flow within a hematoma and permit differentiation (Sepulveda, 2000).

PLACENTAL CALCIFICATION

Calcium salts can be deposited throughout the placenta but are most common on the basal plate. Calcification accrues with advancing gestation, and greater degrees are associated with smoking and increasing maternal serum calcium levels (Bedir Findik, 2015; Klesges, 1998; McKenna, 2005). These hyperechoic deposits can easily be seen sonographically, and a grading scale from 0 to 3 reflects increasing calcification with increasing numerical grade (Grannum, 1979). Following this scheme, a grade 0 placenta is homogeneous, lacks calcification, and displays a smooth, flat chorionic plate. A grade 1 placenta has scattered echogenicities and subtle chorionic plate undulations. Grade 2 shows echogenic stippling at the basal plate. Large, echogenic comma shapes originate from an indented chorionic plate, but their curve falls short of the basal plate. Last, a grade 3 placenta has echogenic indentations extending from the chorionic plate to the basal plate, which create discrete components that resemble cotyledons. Basal plate

densities also increase.

As a predictor, this grading scale is not useful for neonatal outcome near term (Hill, 1983; McKenna, 2005; Montan, 1986). However, data from two small studies link grade 3 placenta prior to 32 weeks with stillbirth and some other adverse pregnancy outcomes (Chen, 2011, 2015).

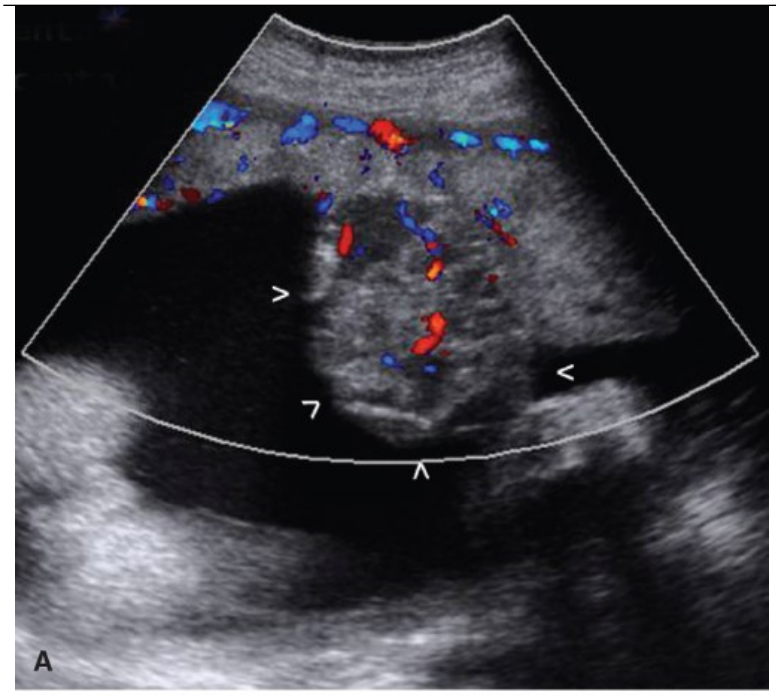
PLACENTAL TUMORS

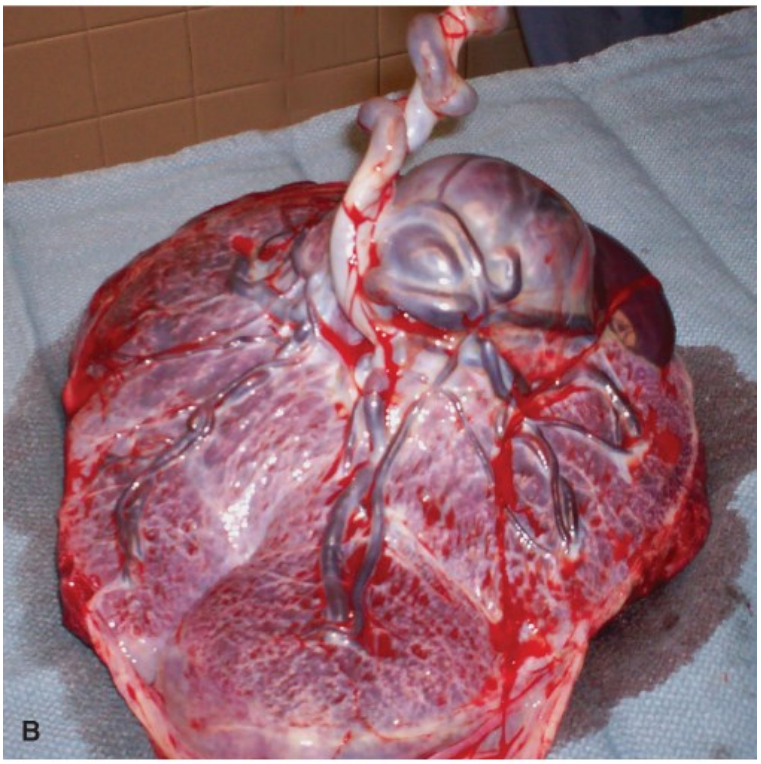
Chorioangioma

These benign tumors have components similar to blood vessels and stroma of the chorionic villus. Also called chorangiomas, these placental tumors occur with an incidence of approximately 1 percent (Guschmann, 2003). In some cases, fetal-to-maternal hemorrhage across tumor capillaries leads to elevated levels of maternal serum alpha-fetoprotein (MSAFP), prompting sonographic evaluation. Their characteristic sonographic appearance shows a well-circumscribed, rounded, predominantly hypoechoic lesion lying near the chorionic plate and protruding into the amnionic cavity (Fig. 6-4). Documenting increased blood flow by color Doppler helps to distinguish these lesions from other placental masses such as hematoma, partial hydatidiform mole, teratoma, metastases, and leiomyoma (Prapas, 2000). Although rare, *chorangiocarcinoma* tumors clinically mirror chorangiomas (Huang, 2015).

FIGURE 6-4

Placental chorioangioma. **A.** Color Doppler imaging displays blood flow through a large chorioangioma with its border outlined by white arrows. **B.** Grossly, the chorioangioma is a round, well-circumcised mass protruding from the fetal surface.





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Small chorioangiomas are usually asymptomatic. Large tumors, typically those measuring >4 cm, can create significant arteriovenous shunting within the placenta to cause high-output heart failure, hydrops, and fetal death (Al Wattar, 2014). Compression of fetal erythrocytes within tumor vessels can lead to hemolysis and microangiopathic anemia (Bauer, 1978). Hydramnios, preterm delivery, and fetal-growth restriction are other sequelae. Rare cases include tumor vessel rupture, hemorrhage, and fetal death (Batukan, 2001). At the other extreme, rare tumor infarction can lead to symptom reversal (Zalel, 2002).

Gray-scale and color Doppler interrogation of the placenta and amnionic fluid volume are used to identify these tumors. Diagnostic tools that can affirm associated fetomaternal hemorrhage include MSAFP level and Kleihauer-Betke stain. With fetal concern, echocardiography assesses cardiac function, whereas middle cerebral artery interrogation is used to identify fetal anemia.

Several fetal therapies interfere with the vascular supply to the tumor and reverse fetal heart failure. At specialized perinatal centers, endoscopic laser ablation of feeder vessels to the tumor is most frequently used and is associated with favorable fetal outcomes (Hosseinzadeh, 2015). Of other therapy, fetal transfusion can treat serious anemia, amnioreduction can temporize hydramnios, and digoxin therapy can assist fetal heart failure.

Metastatic Tumors

Maternal malignant tumors rarely metastasize to the placenta. Of those that do, melanomas, leukemias and lymphomas, and breast cancer are the most common (Al-Adnani, 2007). Tumor cells usually are confined within the intervillous space. As a result, metastasis to the fetus is uncommon but is most often seen with melanoma (Alexander, 2003).

Similarly, cases in which fetal malignancy metastasizes to the placenta are rare (Reif, 2014). These are predominantly fetal neuroectodermal tumors, and only one case in the literature describes transplantation of tumor to the maternal uterus (Nath, 1995).

AMNIOCHORION

Chorioamnionitis

Normal genital-tract flora can colonize and infect the membranes, umbilical cord, and eventually the fetus. Bacteria most commonly ascend after prolonged membrane rupture and during labor to cause infection. Organisms initially infect the chorion and adjacent decidua in the area overlying the internal os. Subsequently, progression leads to full-thickness involvement of the membranes—chorioamnionitis. Organisms often then spread along the chorioamnionic surface to colonize and replicate in amnionic fluid. Inflammation of the chorionic plate and of the umbilical cord—*funisitis*—may follow (Kim, 2015; Redline, 2012).

Most commonly, there is microscopic or occult chorioamnionitis, which is caused by a wide variety of microorganisms. This is frequently cited as a possible explanation for many otherwise unexplained cases of ruptured membranes, preterm labor, or both as discussed in Chapter 42 (Cervical Dysfunction). In some cases, gross infection is characterized by membrane clouding and is sometimes accompanied by a foul odor that depends on bacterial species.

Other Membrane Abnormalities

Amnion nodosum is a condition characterized by numerous small, light-tan nodules on the amnion overlying the chorionic plate. These may be scraped off the fetal surface and contain deposits of fetal squames and fibrin that reflect prolonged and severe oligohydramnios (Adeniran, 2007).

Two notable bandlike structures can be formed by the fetal membranes. Of these, *amnionic band sequence* is an anatomical disruption sequence in which amnion bands tether, constrict, or amputate fetal parts. Amnionic bands commonly cause limb-reduction defects, facial clefts, or encephalocele (Barzilay, 2015; Guzmán-Huerta, 2013). Umbilical cord compromise is another sequela (Barros, 2014; Heifetz, 1984b). Severe defects of the spine or ventral wall that accompany amnionic bands suggest a *limb-body wall complex*, described in Chapter 10 (Gastrointestinal Tract).

Clinically, sonography often first identifies the sequelae of this sequence rather than the bands themselves. As with any fetal anomaly, targeted sonography is indicated. Identification of a limb-reduction defect, an encephalocele in an atypical location, or an extremity with edema or positional deformity should prompt careful evaluation for amnionic bands.

Management depends on the degree of anatomic deformity. Fetoscopic laser interruption of the band may be suitable in highly selected antepartum cases (Javadian, 2013; Mathis, 2015).

In contrast, an *amnionic sheet* is formed by normal amniochorion draped over a preexisting uterine synechia. Generally, these sheets pose little fetal risk, although slightly higher rates of preterm membrane rupture and placental abruption have been described (Korbin, 1998; Nelson, 2010; Tuuli, 2012).

UMBILICAL CORD

Length

Most umbilical cords at delivery are 40 to 70 cm long, and very few measure <30 cm or >100 cm. Cord length is influenced positively by both amnionic fluid volume and fetal mobility (Miller, 1982). In retrospective studies, short cords have been linked with congenital malformations and intrapartum distress (Baergen, 2001; Krakowiak, 2004; Yamamoto, 2016). Excessively long cords are linked with cord entanglement or prolapse and with fetal anomalies (Olaya-C, 2015; Rayburn, 1981).

Because antenatal determination of cord length is technically limited, cord diameter has been evaluated as a predictive marker for fetal outcomes. Some have linked lean cords with poor fetal growth and large-diameter cords with macrosomia (Proctor, 2013). However, the clinical utility of this parameter is still unclear (Barbieri, 2008; Cromi, 2007; Raio, 1999b, 2003).

Coiling

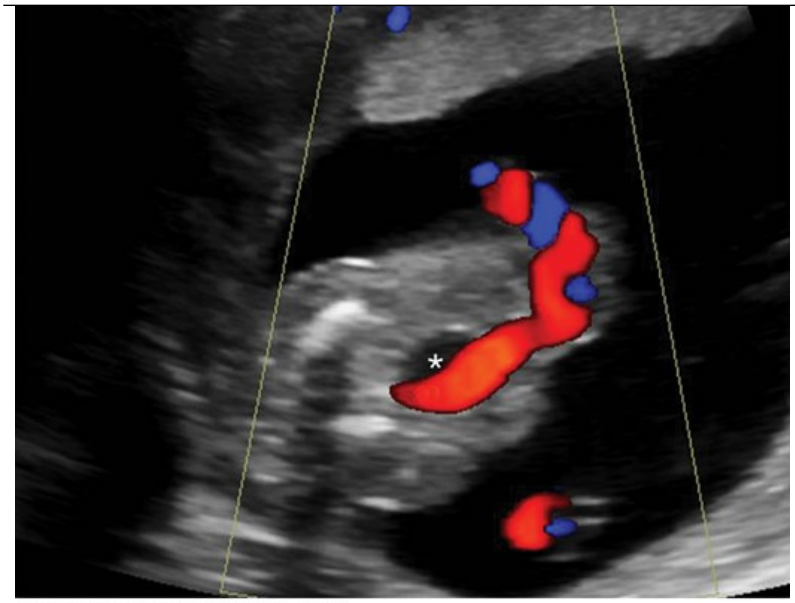
Cord coiling characteristics have been reported but are not currently part of standard sonographic evaluation. Usually the umbilical vessels spiral through the cord in a sinistral, that is, left-twisting direction (Fletcher, 1993; Lacro, 1987). The number of complete coils per centimeter of cord length is termed the *umbilical coiling index—UCI* (Strong, 1994). A normal, antepartum, sonographically derived UCI is 0.4, and this contrasts with a normal, postpartum, physically measured value of 0.2 (Sebire, 2007). UCIs <10th percentile are considered *hypocoiled,* and those >90th percentile are *hypercoiled*. Clinically, the significance of coiling extremes is controversial. Some studies evaluating large, unselected cohorts find no associations between UCI values and poor neonatal outcome (Jessop, 2014; Pathak, 2010). In others, extremes are linked with various adverse outcomes but most consistently with intrapartum fetal heart rate abnormalities, preterm labor, or fetal-growth restriction (Chitra, 2012; de Laat, 2006; Predanic, 2005; Rana, 1995).

Vessel Number

Counting cord vessel number is a standard component of anatomical evaluation during fetal sonographic examination and immediately after delivery (Fig. 6-5). Embryos initially have two umbilical veins. In the first trimester, the right vein typically atrophies to leave one large vein to accompany the two, thick-walled umbilical arteries. Four-vessel cords are rare and often associated with congenital anomalies (Puvabanditsin, 2011). If it is an isolated finding, however, prognosis can be good (Avnet, 2011).

FIGURE 6-5

Two umbilical arteries are typically documented sonographically in the second trimester. They encircle the fetal bladder (*asterisk*) as extensions of the superior vesical arteries. In this color Doppler sonographic image, a single umbilical artery, shown in red, runs along the bladder wall before joining the umbilical vein (blue) in the cord. Below this, the two vessels of the cord, seen as a larger red and smaller blue circle, are also seen floating in a cross section of a cord segment.



The most common aberration is that of a single umbilical artery (SUA), with a cited incidence of 0.63 percent in liveborn neonates, 1.92 percent with perinatal deaths, and 3 percent in twins (Heifetz, 1984a). Fetuses with major malformations frequently have a single artery. Thus, its identification often prompts consideration for targeted sonography and possibly fetal echocardiography. The most frequent anomalies are cardiovascular and genitourinary (Hua, 2010; Murphy-Kaulbeck, 2010). In an anomalous fetus, a single artery greatly increases the aneuploidy risk, and amniocentesis is recommended (Dagklis, 2010; Lubusky, 2007).

If target sonography finds otherwise normal anatomy, an isolated single artery in an otherwise low-risk pregnancy does not significantly increase the fetal aneuploidy risk. However, as in isolated finding, it has been associated with fetal-growth restriction and perinatal death in some but not all studies (Chetty-John, 2010; Gutvirtz, 2016; Hua, 2010; Murphy-Kaulbeck, 2010; Voskamp, 2013). Thus, while clinical monitoring of growth is reasonable, the value of sonographic surveillance is unclear.

A rare anomaly is that of a fused umbilical artery with a shared lumen. It arises from failure of the two arteries to split during embryological development. The common lumen may extend through the entire cord, but, if partial, it is typically found near the placental insertion site (Yamada, 2005). In one report, these were associated with a higher incidence of marginal or velamentous cord insertion, but not congenital fetal anomalies (Fujikura, 2003).

Found in most placentas, the *Hyrtl anastomosis* is a connection between the two umbilical arteries and lies near the cord insertion into the placenta. This anastomosis acts as a pressure-equalizing system between the arteries (Gordon, 2007). As a result, redistribution of pressure gradients and blood flow improves placental perfusion, especially during uterine contractions or during compression of one umbilical artery. Fetuses with a single umbilical artery lack this safety valve (Raio, 1999a, 2001).

Remnants and Cysts

Several structures are housed in the umbilical cord during fetal development, and their remnants may be seen when the mature cord is viewed transversely. Indeed, Jauniaux and colleagues (1989) sectioned 1000 cords, and in one fourth of the specimens, they found remnants of vitelline duct, allantoic duct, and embryonic vessels. These were not associated with congenital malformations or perinatal complications.

Cysts occasionally are found along the course of the cord. They are designated according to their origin. *True cysts* are epithelium-lined remnants of the allantoic or vitelline ducts and tend to be located closer to the fetal insertion site. In contrast, the more common *pseudocysts* form from local degeneration of Wharton jelly and occur anywhere along the cord. Both have a similar sonographic appearance. Single umbilical cord cysts identified in the first trimester tend to resolve completely, however, multiple cysts may portend miscarriage or aneuploidy (Ghezzi, 2003; Hannaford, 2013). Cysts persisting beyond this time are associated with a risk for structural defects and chromosomal anomalies (Bonilla, 2010; Zangen, 2010).

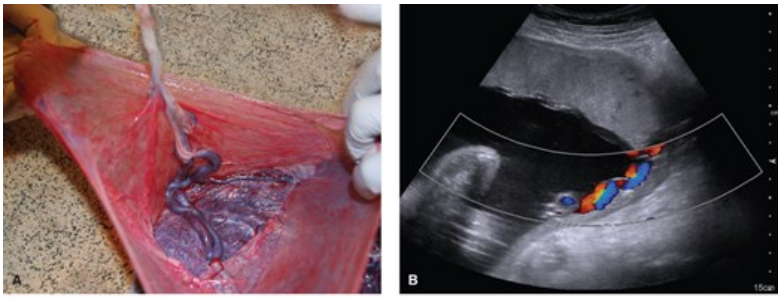
Insertion

The cord normally inserts centrally into the placental disc, but eccentric, marginal, or velamentous insertions are variants. Of these, eccentric insertions in general pose no identifiable fetal risk. Marginal insertion is a common variant—sometimes referred to as a *battledore placenta*—in which the cord anchors at the placental margin. In one population-based study, the rate was 6 percent in singleton gestations and 11 percent in twins (Ebbing, 2013). This common insertion variant rarely causes problems, but it and velamentous insertion occasionally result in the cord being pulled off during delivery of the placenta (Ebbing, 2015; Luo, 2013). In monochorionic twins, this insertion may be associated with weight discordance (Kent, 2011).

With velamentous insertion, the umbilical vessels characteristically travel within the membranes before reaching the placental margin (Fig. 6-6) The incidence of velamentous insertion approximates 1 percent but is 6 percent with twins (Ebbing, 2013). It is more commonly seen with placenta previa (Papinniemi, 2007; Räisänen, 2012). Antenatal diagnosis is possible sonographically, and with velamentous insertion, cord vessels are seen traveling along the uterine wall before entering the placental disc. Clinically, vessels are vulnerable to compression, which may lead to fetal hypoperfusion and acidemia. Higher associated rates of low Apgar scores, stillbirth, preterm delivery, and small for gestational age have been noted (Ebbing, 2017; Esakoff, 2015; Heinonen, 1996; Vahanian, 2015). Accordingly, monitoring of fetal growth is reasonable either clinically or sonographically (Vintzileos, 2015).

FIGURE 6-6

Velamentous cord insertion. **A.** The umbilical cord inserts into the membranes. From here, the cord vessels branch and are supported only by membrane until they reach the placental disc. **B.** When viewed sonographically and using color Doppler, the cord vessels appear to lie against the myometrium as they travel to insert marginally into the placental disc, which lies at the top of this image.



Last, with the very uncommon furcate insertion, umbilical vessels lose their protective Wharton jelly shortly before they insert. As a result, they are covered only by an amnion sheath and prone to compression, twisting, and thrombosis.

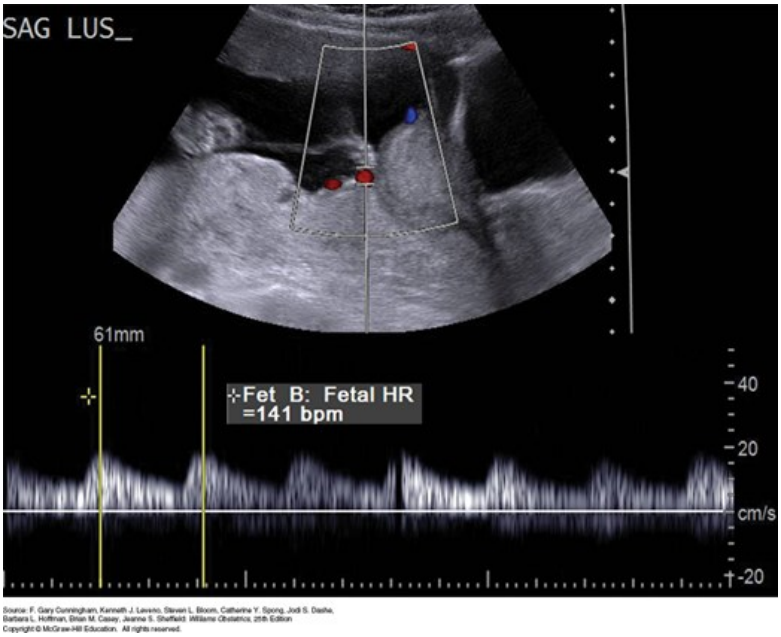
Vasa Previa

With this condition, vessels travel within the membranes and overlie the cervical os. There, they can be torn with cervical dilatation or membrane rupture, and laceration can lead to rapid fetal exsanguination. Over the cervix, vessels can also be compressed by a presenting fetal part. Fortunately, vasa previa is uncommon and has an incidence of 2 to 6 per 10,000 pregnancies (Ruiter, 2016; Sullivan, 2017). Vasa previa is classified as type 1, in which vessels are part of a velamentous cord insertion, and type 2, in which involved vessels span between portions of a bilobate or a succenturiate placenta (Catanzarite, 2001). Two other risks are conception with in vitro fertilization and second-trimester placenta previa, with or without later migration (Baulies, 2007; Schachter, 2003).

Compared with intrapartum diagnosis, antepartum diagnosis greatly improves the perinatal survival rate, which ranges from 97 to 100 percent (Oyelese, 2004; Rebarber, 2014; Swank, 2016). Thus, vasa previa is ideally identified early, although this is not always possible. Clinically, an examiner is occasionally able to palpate or directly see a tubular fetal vessel in the membranes overlying the presenting part. Effective screening for vasa previa begins during scheduled midtrimester sonographic examination. In suspicious cases, transvaginal sonography is added and shows cord vessels inserting into the membranes—rather than directly into the placenta—and vessels running above the cervical internal os (Fig. 6-7). Routine color Doppler interrogation of the placental cord insertion site, particularly in cases of placenta previa or low-lying placenta, may aid its detection. With this, the vessel waveform reflects the fetal heart rate. In one systematic review, the median prenatal detection rate was 93 percent (Ruiter, 2015).

FIGURE 6-7

Vasa previa. Using color Doppler, an umbilical vessel (*red circle*) is seen overlying the internal os. At the bottom, the Doppler waveform seen with this vasa previa has the typical appearance of an umbilical artery, with a pulse rate of 141 beats per minute.



Once vasa previa is identified, subsequent imaging is reasonable because 6 to 17 percent of cases ultimately resolve (Rebarber, 2015; Swank, 2016). Bed rest apparently has no added advantage. Antenatal corticosteroids can be provided as indicated or given prophylactically at 28 to 32 weeks' gestation to cover possible urgent preterm delivery. Antenatal hospitalization may be considered at 30 to 34 weeks to permit surveillance and expedited delivery for labor, bleeding, or rupture of membranes. Data supporting this is limited, and admission may best serve women with risk factors that portend early delivery (Society for Maternal-Fetal Medicine, 2015). A few cases of antepartum fetoscopic surgery with vessel laser ablation are described (Hosseinzadeh, 2015; Johnston, 2014). However, current practice is early scheduled cesarean delivery. Robinson and Grobman (2011) performed a decision analysis and recommend elective cesarean delivery at 34 to 35 weeks' gestation to balance the risks of perinatal exsanguination versus preterm birth morbidity. The Society for Maternal-Fetal Medicine (2015) considers planned cesarean delivery at 34 to 37 weeks' gestation reasonable.

At delivery, the fetus is expeditiously delivered after the hysterotomy incision in case a vessel is lacerated during uterine entry. Delayed cord clamping is not encouraged.

In all pregnancies, otherwise unexplained vaginal bleeding either antepartum or intrapartum should prompt consideration of vasa previa and a lacerated fetal vessel. In many cases, bleeding is rapidly fatal, and infant salvage is not possible. With less hemorrhage, however, it may be possible to distinguish fetal versus maternal bleeding. Various tests may be used, and each relies on the increased resistance of fetal hemoglobin to denaturing by alkaline or acid reagents (Odunsi, 1996; Oyelese, 1999).

Knots, Strictures, and Loops

Various mechanical abnormalities in the cord can impede blood flow and sometimes cause fetal harm. Of these, *true knots* are found in approximately 1 percent of births. These form from fetal movement, and associated risks include hydramnios and diabetes (Hershkovitz, 2001; Räisänen, 2013). Knots are especially common and dangerous in monoamnionic twins, which are discussed in Chapter 45 (Unique Fetal Complications). When true knots are associated with singleton fetuses, the stillbirth risk is increased four- to tenfold (Airas, 2002; Sørnes, 2000).

Knots can be found incidentally during antepartum sonography, and a "hanging noose" sign is suggestive (Ramon y Cajal, 2006). Three-dimensional and color Doppler aid diagnostic accuracy (Hasbun, 2007). With these knots, optimal fetal surveillance is unclear but may include umbilical artery Doppler velocimetry, nonstress testing, or subjective fetal movement monitoring (Rodriguez, 2012; Scioscia, 2011). Allowing vaginal delivery is suitable,

but abnormal intrapartum fetal heart rate tracings are more often encountered. That said, cesarean delivery rates are not increased, and cord blood acid-base values are usually normal (Airas, 2002; Maher, 1996).

In contrast, *false knots* form from focal redundancy and folding of an umbilical cord vessel. These lack clinical significance.

Cord strictures are focal narrowings of the diameter that usually develop near the fetal cord insertion site (Peng, 2006). Characteristic pathological features include an absence of Wharton jelly and stenosis or obliteration of cord vessels at the narrow segment (Sun, 1995). In most instances, the fetus is stillborn (French, 2005). Even less common is a cord stricture caused by an amnionic band.

Cord loops are frequently encountered and are caused by coiling around various fetal parts during movement. A cord around the neck—a *nuchal cord*—is common, and vaginal delivery is suitable. One loop is reported in 20 to 34 percent of deliveries; two loops in 2.5 to 5 percent; and three loops in 0.2 to 0.5 percent (Kan, 1957; Sørnes, 1995; Spellacy, 1966). During labor, up to 20 percent of fetuses with a nuchal cord have moderate to severe variable heart rate decelerations, and these are associated with a lower umbilical artery pH (Hankins, 1987). Cords wrapped around the body can have similar effects (Kobayashi, 2015). Despite their frequency, nuchal cords are not associated with greater rates of adverse perinatal outcome (Henry, 2013; Sheiner, 2006).

Last, a *funic presentation* describes when the umbilical cord is the presenting part in labor. These are uncommon and most often are associated with fetal malpresentation (Kinugasa, 2007). A funic presentation in some cases is identified with placental sonography and color flow Doppler (Ezra, 2003). Overt or occult cord prolapse can complicate labor. Thus, once identified at term, cesarean delivery is typically recommended.

Vascular

Cord hematomas are rare and generally follow rupture of an umbilical vessel, usually the vein, and bleeding into the Wharton jelly. Hematomas have been associated with abnormal cord length, umbilical vessel aneurysm, trauma, entanglement, umbilical vessel venipuncture, and funisitis (Gualandri, 2008). Most are identified postpartum, but hematomas are recognized sonographically as hypoechoic masses that lack blood flow (Chou, 2003). Sequelae include stillbirth or intrapartum abnormal fetal heart rate pattern (Abraham, 2015; Barbati, 2009; Sepulveda, 2005; Towers, 2009).

Umbilical cord vessel thromboses are rare in utero events and seldom diagnosed antepartum. Approximately 70 percent are venous, 20 percent are venous and arterial, and 10 percent are arterial thromboses (Heifetz, 1988). These all have high associated rates of stillbirth, fetal-growth restriction, and intrapartum fetal distress (Minakami, 2001; Sato, 2006; Shilling, 2014). If these are identified antepartum as hypoechoic masses without blood flow, data from case reports support consideration of prompt delivery if of viable age (Kanenishi, 2013).

An *umbilical vein varix* can complicate either the intraamnionic or fetal intraabdominal portion of the umbilical vein. Sonographically and complemented by color Doppler, rare intraamnionic varices show cystic dilatation of the umbilical vein that is contiguous with a normal-caliber portion. Of complications, an intraamnionic varix may compress an adjacent umbilical artery or can rupture or thrombose. In cases without these, White and colleagues (1994) recommend fetal surveillance and delivery once fetal maturity is confirmed. However, data are limited and derived from case reports.

The rare *umbilical artery aneurysm* is caused by congenital thinning of the vessel wall with diminished support from Wharton jelly. Indeed, most form at or near the cord placental insertion site, where this support is absent. These are associated with single umbilical artery, trisomy 18, amnionic fluid volume extremes, fetal-growth restriction, and stillbirth (Hill, 2010; Vyas, 2016). At least theoretically, these aneurysms could cause fetal compromise and death by compression of the umbilical vein. These aneurysms may appear sonographically as a cyst with a hyperechoic rim. Within the aneurysm, color flow and spectral Doppler interrogation demonstrate either low-velocity or turbulent nonpulsatile flow (Olog, 2011; Sepulveda, 2003; Shen, 2007b). Although not codified, management may include fetal karyotyping, antenatal fetal surveillance, and early delivery to prevent stillbirth (Doehrman, 2014).

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