SONOGRAPHIC ASSESSMENT OF THE UMBILICAL CORD

CORD LENGTH

The umbilical cord is derived from the allantois and stalk of the yolk sac. The average length of the umbilical cord is 59 cm with a range of 22 to 130 cm\(^3\). The two factors that determine umbilical cord length are sufficient space in the amniotic cavity for movement and the tensile force applied to the umbilical cord during fetal movements\(^2\). If embryo/fetal movement is impeded, the tensile stretch placed on the cord will be less and the eventual cord length will be shorter. For example, the umbilical cord length in twins is generally shorter than for singletons\(^3\). In fetal rats, the exposure to alcohol during prenatal development is associated with a shortened umbilical cord\(^4\). In abnormal pregnancies the frequency and velocity of fetal movements decreases\(^5\). This would explain the reduction in cord length in fetuses who subsequently die in utero. However, if the reason for the intrauterine demise was acute, cord length would not be affected\(^6\). Cord length is longer in fetuses with polyhydramnios\(^7\).

For the first half of the first trimester the amniotic cavity diameter and crown-rump length approximate one another (Fig. 1). Hence, crown-rump length and cord length are roughly equivalent\(^6\) (Fig. 2). After 10 weeks' gestation amniotic fluid volume increases rapidly due to the initiation of fetal voiding\(^8\). As a result, the amniotic cavity increases in size and the cord length becomes longer than the crown-rump length.

![Figure 1. First trimester - amniotic cavity diameter equivalent to the crown-rump length. Click for larger image.](image-url)
UMBILICAL CORD CYSTS

Umbilical cord cysts (Fig. 3) are derived from either the allantois (i.e. urachus), the omphalomesenteric duct, or focal edema of Wharton's jelly (pseudocysts). The former two have an epithelial lining and the latter does not. Pseudocysts are far more common than umbilical cord cysts. In general, an allantoic cyst is centrally located and will, therefore, widely separate the umbilical arteries. Since a pseudocyst results from edema of Wharton's jelly, the umbilical vessels may course through the cyst\(^9\). Because of their etiology, allantoic and omphalomesenteric duct cysts are usually found near the fetal end of the umbilical cord. While umbilical cord cysts are usually small, cysts of > 5 cm have been reported\(^10\). If an umbilical cord cyst is large enough, it may compress the umbilical arteries and result in intrauterine growth restriction\(^11\) or fetal distress\(^12\).
The prevalence of umbilical cord cysts between 7 and 13 weeks' gestation is 3%. First trimester umbilical cord cysts that subsequently resolve prior to the second trimester are probably pseudocysts. It has been hypothesized that their formation may be due to increased hydrostatic pressure associated with the initiation of cord coiling and physiologic mid-gut herniation. They have not been associated with an increase in karyotypic abnormalities. However, the presence of multiple first trimester umbilical cord cysts is associated with an increased rate of adverse pregnancy outcome. Ghezzi et al reported only 1 of 6 pregnancies with multiple umbilical cysts resulted in a normal term pregnancy. Of the remaining 5 cases, there were 2 miscarriages; 2 fetuses with trisomy 18; and a fetus with an obstructive uropathy.

Allantoic cysts of the umbilical cord have been associated with omphalocele and a patent urachus. Pseudocysts that persist into the second trimester have been associated with an increased risk of karyotypic abnormalities (specifically trisomy 18); an increase in structural abnormalities have also been reported.

HYRTL ANASTOMOSIS

The hyrtl anastomosis is an interarterial anastomosis between the umbilical arteries, approximately 3 cm from the placenta cord insertion. The effect of this anastomosis is to equalize pressure between the placental lobes. The hyrtl anastomosis permits perfusion of the entire placenta even if one of the umbilical arteries is obstructed. Pregnancies with a single umbilical artery do not have this protective mechanism, possibly explaining the associated increase prevalence of fetal distress and intrauterine fetal demise. There is a higher prevalence of umbilical artery fusion at the placental cord insertion with velamentous or marginal placental cord insertions - both conditions in which the placentas supplied by each umbilical artery is vastly different. If the anastomosis does not equalize the blood flow between the arteries, the placental lobes will be asymmetric in size.

UMBILICAL CORD TWIST

Umbilical cord coiling results from intrinsic properties within the umbilical cord (i.e., growth of the vessels and the differential blood flow within the umbilical arteries) and fetal movement. Left twists outnumber right twists by 7 to 1. The right umbilical artery is usually larger than the left, explaining the increased frequency of left twists. The smooth muscle in the umbilical artery wall that coils around the vessel and may also play a role in the coiling of the umbilical cord. If the smooth muscle fibers are absent, or if the intravascular pressure is decreased, coiling of the umbilical cord will be reduced or not occur.

Umbilical cord coiling is typically established by the end of the first trimester. However, 30% of women with a non-coiling umbilical cord at 20 weeks' gestation were found to have a normally coiled umbilical cord latter in gestation. Five percent of 3-vessel umbilical cords and 15% of cords with a single umbilical artery do not have any twists.

The coiling of the umbilical cord increases its turgor making it more resistant to compression. Non-coiled umbilical cords (Fig. 4) have been associated with an increased risk of intrapartum fetal heart rate decelerations, fetal distress, operative delivery, and intrauterine death. It has not yet been determined if a reduction in the number of coils, as well as a complete absence of coils, places a fetus at risk.
Figure 4. Straight umbilical cord. Click for larger image.

Over coiling of the umbilical cord can result in a slowing of blood flow with the potential for venous thrombosis. Over coiling is associated with an increase in intrauterine growth restriction, an increase in fetal demise, and twice the frequency of velamentous cord insertions. It has been hypothesized that stillbirths of unknown etiology may be caused by abnormal coiling of the umbilical cord.

DISCORDANT UMBILICAL ARTERIES

A 1 mm difference in the diameter of umbilical arteries has been considered discordant (Fig. 5). This degree of discrepancy occurs once in approximately every 72 pregnancies. The resistance index is higher in the smaller vessel. A discrepancy in the size of the umbilical arteries is associated with abnormal placental cord insertions (marginal and velamentous) and an increase in placental abnormalities (succenturiate, bipartate placentas, and placental infarcts). An absence of the hyrtl anastomosis between umbilical arteries may prevent the different pressures within the arteries from being equalized. Differences in the S/D ratio between umbilical arteries of 29% to 10% have been recorded between the second trimester and term, respectively. The decreasing differences in resistance between umbilical arteries as gestation advances may be due to the maturation of the hyrtl anastomosis.
TWO VESSEL UMBILICAL CORD

The umbilical arteries originate from the left and right common iliac arteries. The incidence of a single umbilical artery is 0.1% in embryos and 0.6% to 1% at term. Autopsy series generally have a two-fold higher incidence. This discrepancy is consistent with the theory of umbilical artery atrophy resulting in a single umbilical artery. The rate of a single umbilical artery in twins is 4.6%.

The transabdominal sonographic visualization rate of the number of vessels in the umbilical cord increases significantly from 15 to 17 weeks' gestation (74.1% to 97.6%; p< 0.001). In addition to gestational age, maternal body habitus, a reduction in amniotic fluid volume, and the position of the umbilical cord within the amniotic cavity affect the success of identifying the number of vessels in the umbilical cord.

Umbilical artery fusion may occur along the umbilical cord. Hence, both a 2-vessel, as well as a 3-vessel, umbilical cord may be visualized in the same patient. Intermittent fusion of the umbilical arteries does not have the same affect on neonatal outcome as a uniform single umbilical artery.

The number of vessels around the fetal bladder (Fig. 6) does not reflect the number of vessels in a free loop of umbilical cord in every case.
Approximately 18.4% to 30% of fetuses with a single umbilical artery will have other structural or karyotypic abnormalities (Table I)\textsuperscript{31,32}. When there is a single umbilical artery, there is not a predominance of congenital anomalies from a particular organ system\textsuperscript{28,29}. However, a single umbilical artery is usually associated with acardiac twins and sirenomelia\textsuperscript{29}. In the latter congenital anomaly, the single umbilical artery is a persistent vitelline artery (Type II single umbilical artery) and is not due to the atrophy of one of the umbilical arteries (Type I)\textsuperscript{33}.

Table I. Two Vessel Cord Associations\textsuperscript{29,31,34}

- Preterm delivery
- Small-for-gestational age
- Intrauterine growth restriction
- Stillbirth
- Karyotypic abnormalities
  - Trisomy 18
  - Trisomy 13
- Structural anomalies
  - Cardiovascular
  - Central nervous system
  - Gastrointestinal
  - Genitourinary
  - Respiratory
  - Musculoskeletal
- Placental abnormalities
Velamentous cord insertion
Circumvallat

Associated malformations result in the higher perinatal losses with a single umbilical artery. However, the rate of stillbirth is even increased in otherwise structurally normal fetuses with a single umbilical artery. The overall mortality with a single umbilical artery is approximately four times a control group. If the fetus appears to be structurally normal on a detailed ultrasound examination, it has been reported that from 0 to 7% may be found to have an anomaly at birth.

If there are associated structural anomalies or symmetric intrauterine growth restriction, the likelihood of a karyotypic abnormality is increased. Trisomy 18 is the most common aneuploidy associated with a 2-vessel umbilical cord. When polyhydramnios is present with a single umbilical artery, esophageal atresia or a tracheoesophageal fistula should be considered. Compensatory dilatation of a single umbilical artery may prevent the fetus from developing intrauterine growth restriction (Fig. 7).

Figure 7. Two-vessel umbilical cord. Click for larger image.

There is less Wharton's jelly surrounding a single umbilical artery. This may result in the higher vulnerability of the umbilical cord to compression in the third trimester.

If a single umbilical artery is the only abnormality detected, body weight, length, head circumference, and IQ are comparable to controls with 2 umbilical arteries at 4 years of age.

UMBILICAL CORD HEMATOMA
Umbilical cord hematomas may occur spontaneously or iatrogenically after an intrauterine procedure (i.e.
amniocentesis or percutaneous umbilical blood sampling\textsuperscript{49}). Spontaneous hematomas are estimated to occur one in every 500 deliveries. They are usually located at the fetal end of the umbilical cord and are due to the rupture of the umbilical vein\textsuperscript{41}. While most umbilical cord hematomas are small, larger hematomas have been associated with an increase in perinatal mortality\textsuperscript{42,43,44}.

ANEURYSM OF THE UMBILICAL ARTERY AND VEIN
An umbilical artery aneurysm is a rare, but potentially lethal anomaly. The turbulent flow within the aneurysm can be appreciated with color Doppler. Vascular compression by an umbilical cord aneurysm may result in a sudden intrauterine fetal demise. Once fetal viability has been attained, an early delivery should, therefore, be planned when fetal lung maturity has been confirmed, or if there are signs of distress\textsuperscript{45}.

An aneurysm of the umbilical vein may result in a cord hematoma, fetal anemia, non-immune hydrops and sudden intrauterine fetal death\textsuperscript{46}.

CORD HEMANGIOMA
The two primary tumors of the umbilical cord are hemangiomas and teratomas. Both are quite rare. Umbilical cord hemangiomas are echogenic. As a result, the differential diagnosis would include a cord hematoma or a teratoma. Umbilical cord edema is characteristic for a hemangioma; it may extend for quite a distance beyond the region of the hemangioma. The presence of calcifications within an echogenic cord mass would suggest a diagnosis of a teratoma rather than a hemangioma. Mechanical compression of the umbilical vessels by the hemangioma may result in fetal compromise. A cord hemangioma may bleed, resulting in fetal anemia and secondary hydrops\textsuperscript{47,48}.

TRUE KNOT OF THE UMBILICAL CORD
True knots in the umbilical cord are reported in 0.3 - 2.1\% of deliveries. Antepartum fetal death is increased over four-fold with this particular cord complication. The "hanging noose sign" is a transverse section of umbilical cord surrounded by a loop of cord\textsuperscript{49}. A "clover-leaf" pattern of a true knot has also been described\textsuperscript{50}. With sufficient cord constriction, umbilical venous flow is pulsatile in the post-stenotic segment of cord\textsuperscript{51}. As the degree of cord tightening increases, a systolic notch will become apparent on the umbilical artery waveform\textsuperscript{52}.

VELAMENTOUS CORD INSERTION (VCI)
Velamentous cord insertion occurs when the umbilical vessels enter the membranes before reaching the placenta (Fig. 8). Since the fetal vessels are only surrounded by amnion and devoid of Wharton’s jelly, the risk of compression, thrombosis, and rupture are increased. VCI occurs in 0.48\% of singletons; 5\% of dichorionic twins; 9\% of monochorionic twins\textsuperscript{49}; and 28\% of triplets\textsuperscript{53}. A velamentous cord insertion has been associated with an increased prevalence of fetal heart rate abnormalities in labor, emergency cesarean section, low birth weight, and preterm delivery.
The detection of the placental cord insertion is influenced by placental location and gestational age. Pretorius and co-workers\textsuperscript{44} detected 67% of cord insertions at 15 to 20 weeks' gestation and 30% between 36 and 40 weeks (Fig. 9). Hasegawa et al\textsuperscript{56} reported a 62.5% sensitivity for detecting VCI with a 100% positive predictive value at 18 weeks' gestation. VCI in the lower uterine segment has a significantly higher rate (p < 0.01) of non-reassuring fetal heart rate patterns. In addition, the length of the abnormal vessel is longer in the lower uterine segment. It has been hypothesized that this is due to the development of the lower uterine segment as gestational age advances. In one series the length of the velamentous vessel was 10.6 cm in the lower uterine segment and 4.7 cm in the upper third of the uterus (p = 0.024)\textsuperscript{55}. 

Figure 8. Fundal velamentous cord insertion of a bilobate placenta. Click for larger image.

Figure 9. Placental cord insertion in the third trimester. Click for larger image.
VASA PREVIA

Vasa previa occurs when fetal vessels extend over the region of the internal cervical os (Fig. 10). Its incidence is between once in every 1200 to 5000 deliveries\textsuperscript{56}. A velamentous cord insertion, succenturiate placental lobe or bilobate placenta have fetal vessels traversing the membranes and may result in a vasa previa. A marginal placenta previa, a low-lying placenta, multiple gestations, and pregnancies resulting from in-vitro fertilization (IVF) have also been associated with vasa previa\textsuperscript{57}.

Figure 10. Color Doppler of a vasa previa. Click for larger image.

A velamentous cord insertion occurs in 14\% of IVF pregnancies, in contrast to 1\% of spontaneous pregnancy\textsuperscript{58}. In one study the incidence of vasa previa after IVF was 1:293 deliveries, in contrast to a rate of 1:6068 deliveries in the general population\textsuperscript{58}. It has been hypothesized that the increase in the placental abnormalities with IVF pregnancies may be related to the improper orientation of the blastocyst at the time of implantation\textsuperscript{57}.

When there is a vasa previa, spontaneous rupture of the membranes may lacerate the fetal vessels, resulting in rapid fetal exsanguination. In the past, vasa previa has been associated with a substantial perinatal mortality. Several series have shown that the antenatal detection of vasa previa is possible\textsuperscript{56,59}. In one series the perinatal mortality decreased from 56\% without, to 3\% with prenatal diagnosis\textsuperscript{60}.

Catanzarite et al\textsuperscript{61} classified vasa previa into two types. In Type I there is a single placental lobe with a velamentous cord insertion. In Type II fetal vessels cross the internal cervical os connecting two separate placental lobes (Figs. 11 & 12). The sonographic determination of the umbilical cord insertion into the placenta during the routine second trimester ultrasound examination will exclude a Type I vasa previa\textsuperscript{52}. In order to detect Type II vasa previa, the area over the cervix must be evaluated with color Doppler and transvaginal sonography when necessary.
Lee and co-workers\textsuperscript{59} have reported that aberrant vessels extending over the cervix in the second or early third trimester may regress by term. While the umbilical cord insertion is generally fixed, aberrant vessels within the membranes could move because of the differential growth between the lower uterine segment and placenta.

While three-dimensional power Doppler\textsuperscript{61} may help to appreciate the spatial relationship between the aberrant fetal vessels and the internal cervical os, two-dimensional transvaginal sonography is sufficient to make a diagnosis of vasa previa.

REFERENCES


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