Modern Ultrasonography of the Umbilical Cord: Prenatal Diagnosis of Umbilical Cord Abnormalities and Assessment of Fetal Wellbeing

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Source of support: Departmental sources

The umbilical cord is the only connection between the mother and the fetus, through which it is possible to transport respiratory gases, nutrients, and metabolites. Thanks to the umbilical cord, the fetus has also the ability to move, which is necessary for its proper psychomotor development. The correct structure and function of umbilical vessels and the entire umbilical cord determine the possibility of proper development and survival of the fetus. Umbilical cord anatomy should be assessed in the ultrasound examination in the first trimester. It is of vital importance to confirm the correct number of umbilical vessels and their intra-abdominal course, as well as carefully assessing the abdominal and placental insertion sites. In the latter half of pregnancy, the use of the Doppler imaging enables assessment of the function of the fetal-placental vessels, thus providing valuable information about the condition of the fetus.

MeSH Keywords: Congenital Abnormalities • Fetal Distress • Prenatal Diagnosis • Umbilical Cord

Full-text PDF: https://www.medscimonit.com/abstract/index/idArt/913762
Embryology

The formation of the umbilical cord occurs at a very early stage of the embryonic development. Towards the end of the second week of embryonic life, there is a band of a connective tissue between the amnion and the chorion, referred to as the abdominal stalk. The urachus with umbilical blood vessels delves into the stalk. The increasing amniotic cavity, from the dorsal towards the ventral side, gradually covers the embryo, the yolk sac, and the abdominal stalk. From the part of the yolk sac, the intestine is formed, while its extra-embryonic fragment extends in the form of the omphaloenteric duct. Finally, the omphaloenteric duct together with the abdominal stalk form the umbilical cord, connecting the embryo with the chorion (see also textbooks of embryology, e.g., www.embryology.ch).

In the primary umbilical cord, there are 2 veins that lead oxygenated blood from the placenta to the fetus and 2 arteries, in which deoxygenated blood flows in the opposite direction. In the embryonic period, the umbilical arteries are aortic branches. As a result of the proliferation and differentiation of the fetal vascular system, they are transformed into branches of the common iliac arteries. In the postnatal period, the intra-abdominal section of the umbilical arteries closes above the exit of the upper bladder arteries, forming the medial umbilical ligaments. The veins in the abdomen initially run alongside the liver to the sinus venosus of the heart. Then, in the typical development of the embryo, the right vein fades around the sixth to seventh weeks of pregnancy. The left vein increases and connects to the hepatic veins, losing a direct connection to the heart. A stream of blood in the umbilical vein promotes the growth of a vessel in the hepatic vessel network, which connects directly to the inferior vena cava, which leads to the creation of a ductus venosus. After delivery, the umbilical vein and DV become closed, turning into the liver oblique ligament and venous ligament.

The umbilical vessel is surrounded by Wharton’s jelly, and the entire umbilical cord is covered by amnion on the outside. Wharton’s jelly is made of a connective tissue with a generous amount of extracellular matrix, composed mainly of the hydrophilic proteoglycans and hyaluronic acid. It gives the umbilical cord elasticity and strength, preventing tearing, refraction, or compression of the vessels [1].

The assessment of the umbilical cord should be an indispensable element of an ultrasound examination in every trimester of pregnancy. The umbilical cord may be visualized for 42 days of gestation as a ropelike, echogenic structure between the fetus and trophoblast [2,3]. In advanced pregnancy, it is usually impossible to visualize the whole umbilical cord, and its estimation may be extremely difficult. The diagnostic possibilities are primarily limited by the tortuous course and mobility of the umbilical cord, the size and position of the fetus, the size and location of placenta, and the volume of amniotic fluid. In recent years, the advancement of high-resolution ultrasound, three-dimensional ultrasound (3D US), associated with color and/or power Doppler, have made significant improvement in diagnostics. A prenatal diagnosis of abnormalities of the umbilical cord is rarely encountered; therefore, sonography skills are essential. Complementary methods such as color Doppler and 3D HD flow are reliable for diagnostic and differentiation purposes. Color Doppler imaging is a well-established method for a prenatal diagnosis of fetal cord vascular alterations. Complementary use of 3D-HD-Flow improves the diagnosis, as it is a more sensitive technique than power Doppler, and it also allows the direction of flow to be visualized [4].

Umbilical cord abnormalities can have important prognostic implications for perinatal morbidity and mortality.

The defects in the umbilical cord can be divided into:
1. Abnormal length – too long/too short; abnormal cord twist, coil, thin cord;
2. Cord insertions site abnormalities;
3. Cystic abnormalities: true cysts, pseudocysts;
4. Cord hematomas;
5. Solid or complex malformations: angiomyxoma, teratoma;
6. Knots: true, false;
7. Nuchal cord;
8. Vascular anomalies: single umbilical artery, hypoplasia of 1 umbilical artery, supernumerary vessels, aneurysm and varix, persistent right umbilical vein (PRUV);
9. Funic presentation and prolapse cord.

Changes in Umbilical Cord Length

The umbilical cord is usually approximately 50 cm in length; however, in pregnancies, the value considered to be normal is in the range of 35 to 70 cm. The traction force produced by the fetus in the first trimester is assumed to be the main stimulus for its growth and it stops growing by the end of the second trimester [5].

A short UC is probably associated with reduced fetal movement and all its respective causes (e.g., malformation, myopathies, neuropathies, oligohydramnios). Both longer and shorter cords may be associated with increased rates of complications [6]. A particularly short UC may be associated with the body stalk anomaly. This unusual anomaly occurs in 1: 7500 to 1: 31 000 of pregnancies [7]. It can be detected by ultrasound as early as in the first trimester. The upper half of the fetus lies within the amniotic cavity. In severe cases, it may be regarded as an extreme form of the amniotic band syndrome. Fetal karyotype is usually normal. It quite commonly demonstrates multiple
congenital anomalies like abdominal wall defect, single UA, direct attachment of the fetus to the placenta, lower limb deformities and kyphoscoliosis, cranial defects as additional findings. Nuchal translucency and alpha-fetoprotein (AFP) are usually increased [8].

Long umbilical cords are of greater clinical importance in the third trimester and are associated with an increased risk of knots and cord prolapse. During the outflow of the amniotic fluid, the umbilical cord may unexpectedly fall out below the presenting part. Cord prolapse is associated with an approximate 50% perinatal mortality rate [4]. In addition to the long umbilical cord, the most important risk factors include an abnormal fetal position, prematurity, low birth weight, polyhydramnios, abnormal pelvic structure, or multiple pregnancy. All of these cases require vigilance and mandatory internal obstetric examination with an assessment of the fetal heart rate immediately after the withdrawal of amniotic fluid.

**Abnormal Cord Twist or Coil, Thin Cord**

The diameter of the umbilical cord in the third trimester of pregnancy can reach up to 17 mm. The umbilical vessels do not run in a straight line. Umbilical arteries entangle the vein to varying degrees, resulting in a twist, which is usually left-sided. Coiling of the cord gradually decreases with increasing gestational age. The number of cord spirals per centimeter, expressed as the Umbilical coiling Index (UCI), is 0.9–0.6, which denotes that there is a 0.3–0.5 “rim” created by arteries per 1 cm of the umbilical cord, usually up to 40 helical turns [9,10]. Adequate amniotic fluid volume and fetal movements are necessary to develop normal cord coiling and length. An abnormal vessel course within the umbilical cord is associated with a higher rate of fetal distress, chromosomal abnormalities, IUGR, decelerations on cardiotocography, and even fetal death [9,11]. It may be easily visualized with Color Doppler ultrasonography in sagittal and transverse scans.

A thin cord means that umbilical cord circumference is less than 1 cm and is associated with postdates or small-for-gestational age births. A thin cord can be caused by deficiency of Wharton’s jelly, inappropriate number of vessels, or disproportion between the sizes of umbilical vessels (Figure 1).
Cord Placental Insertion Site Abnormalities

The umbilical cord connects to the placenta, usually in its central part. On the section about 3 cm from the placental attachment, the umbilical artery connects to form the so-called Hyrtl anastomosis. This connection compensates for the difference in pressure and volume between the umbilical cord and placental circulation, which seems to be of great importance during uterine contractions [12]. A placental cord insertion should be tracked at routine obstetric ultrasound. The cord insertion abnormalities are best diagnosed with Doppler US. Visualization of cord placental insertion is particularly important in twin pregnancies (Figure 2).

Various cord insertion variants are:
- Marginal insertion – occurs in 7% of pregnancies [13];
- Furcate insertion (rare) – the umbilical vessels separate from the cord substance before their insertion to the placenta;
- Velamentous insertion occurs in 1% of pregnancies, in which the umbilical vessels separate in the membranes at a distance from the placental margin; therefore, a part of the umbilical cord remains between the 2 amniotic membranes without the protection of Wharton’s jelly [13]. This condition is associated with the fetal growth restriction, congenital anomalies and retained placenta. In case of a sudden rupture or thrombosis of the umbilical vessels, the prematurity or fetal death can occur [14]. In those cases, the ultrasound diagnosis is possible, as well as indispensable;
- Vasa previa occurs in 1: 2, 00 births, in which the velamentous insertion, associated with umbilical vessels, crossing the region of the cervical os. These blood vessels, unprotected by the Wharton’s jelly, may tear when the cervix dilates or the membranes ruptures, resulting in life-threatening bleeding and even fetal death [15]. In cases of maternal obesity, when the maternal bladder is empty, or if there is a maternal abdominal wall scar, the blood vessels may be difficult to assess. In these cases, endovaginal sonography may confirm the pathology. According to a screening algorithm proposed by Rebarber et al., transvaginal ultrasound should be performed in every pregnant woman who satisfies any of the following criteria [16]:
  - Placenta praevia in the current pregnancy that is subsequently no longer demonstrable;
  - Vasa previa in a previous pregnancy;
  - Velamentous cord insertion in the lower uterine segment;
  - Accessory placenta in the lower uterine segment;
  - Multiple pregnancy.

Vasa previa, which is an indication for a planned cesarean section at approximately 35 weeks of gestation, before rupture of fetal membranes, should be diagnosed by ultrasound earlier in pregnancy. However, in 24% of cases diagnosed prior to 26 weeks of gestation, vasa previa does not persist into the third trimester [16]. A correct diagnosis and repeated US examinations increase fetal survival from 44% to 97% [15].

Fetal Umbilical Cord Insertion Abnormalities

The most important abnormalities of the fetal side cord attachment are the abdominal wall defects: gastroschisis and omphalocele. The incidence of both is approximately 3: 10 000 [17].

A physiological umbilical hernia normally regresses before the 12th week, resulting from intestinal rotation. Omphalocele, as an abnormality, must be differentiated from the physiological hernia. It is covered by peritoneum. The rate of associated genetic abnormalities is relatively high. In gastroschisis, the rate of associated abnormalities is approximately 10%, and the karyotype is normal in most cases [18]. The wall defect is placed to the right of the UC, which is attached normally to the abdominal wall. The intestines and the other viscera lie in the amniotic cavity. The best method is to use routine grayscale scans (Figure 3).

Cystic Abnormalities: True Cysts and Pseudocysts

The cysts of the umbilical cord can be identified using ultrasound at all stages of gestation. During the first trimester, the prevalence ranges from 0.4% to 3.4% [19,20]. In a prospective study on cysts occurring between 7 and 13 weeks of pregnancy, in 26% of cases, the cysts of the umbilical cord exert an adverse effect on the outcome of pregnancy, especially when they occur in the proximal part or distal umbilical cord (near the fetus or placenta) [21]. In the second and third trimesters, their presence has been described only in case reports or small series [22]. The prognosis and clinical significance are different.
between the first-trimester and the second-/third-trimester umbilical cord cysts. In the study conducted by Zangen et al., as in other publications, an association was found between the presence of second- and third-trimester umbilical cord cysts and fetal anomalies (trisomy 18, IUGR, heart defect) [20].

Umbilical cord cysts are usually classified as true cysts or pseudocysts. True cysts are derived from the embryological remnants of either the allantois or the omphalomesenteric duct. They are typically located towards the fetal insertion of the cord and range from 4 to 60 mm in size (Figure 4) [23–25]. Pseudocysts are more common than true cysts and can be located anywhere along the cord [20]. Pseudocysts and omphalomesenteric cysts are associated with chromosomal abnormalities, unlike allantoid cysts [20,22]; therefore, accurate prenatal diagnosis is very important. When sonographic abnormalities are detected, the patient should be transferred to a referral center for intensive 3D US monitoring and fetal karyotype testing.

Pseudocysts appear due to degeneration or from edema in Wharton’s jelly, and they are not covered by epithelium. Allantoid cysts usually rupture spontaneously in the third trimester. A patent allantois can be diagnosed with gray-scale ultrasound. Lately, a B-flow ultrasound technique has been used to illustrate erythrocytes in the flowing blood by a subtraction mode. B-flow does not use the Doppler principle; therefore, this technique is not angle-dependent. B-flow is useful in visualizing blood vessels with a winding course, similar to the vessels in the umbilical cord [26].

Cord Hematomas

Cord hematomas is another rare abnormality, with a prevalence rate of 1: 55 000 [27]. The rate of perinatal loss associated with this condition is above 50% [28]. The causes of the development of umbilical cord hematoma are mainly unexplained. In approximately 90% of cases, the cause is umbilical vein rupture [28]. Iatrogenic causes result from cordocentesis, amniocentesis, or in utero transfusions. Other similar causes (e.g., twisting and traction of cord, true knots, vessel wall abnormalities, umbilical cord cysts, trauma, post term) can also be observed [29]. Cord hematomas may lead to cardiotocographic (CTG) abnormalities, hypoxic ischemic encephalopathy, or even a fetal death. In cases identified on ultrasound, they are usually located at the fetal cord insertion site.
The differential diagnosis of umbilical cord tumors should comprise umbilical cord teratoma, hemangioma, and angiomyxoma. The management in the third trimester is not clearly defined.

Angiomyxoma is a rare tumor of the umbilical cord associated with increased perinatal morbidity and mortality. Therefore, it should be considered when using prenatal ultrasound for detection of cystic lesion. Color Doppler imaging can easily detect perfusion through the umbilical vessels, so by using high-resolution ultrasound and color Doppler, the umbilical cord tumor can be suspected to be an angiomyxoma without malformations in the fetus. The clinical significance common to all anomalies is determined by their size, which can potentially cause a vascular compromise and affect fetal growth [30].

Teratomas are the most frequent congenital tumors and may occur at any unusual location in the fetus and placenta [31,32]. Umbilical cord teratomas derive from ectopic germ cells that migrate out of the wall of the invaginated gut into the connective tissue of the cord. Umbilical cord teratomas have a very polymorphic presentation. They are observed along the whole length of the cord. Umbilical cord teratomas have both solid and cystic natures, and are covered with skin. The associated anomalies can be observed in nearly half of the cases. Despite the large volume of some tumors, few obstetrical complications have been reported. Umbilical cord teratomas tend to grow rapidly and may cause cardiac failure and fetal hydrops. If the teratoma is associated with the umbilical vessels or omphalocele, it may lead to rupture of the fetal cord [33].

Hemangiomas are extremely rare umbilical tumors. They consist of an angiomatous nodule, encompassed by edema and myxomatous degeneration of Wharton’s jelly. The placental end of the cord is the most frequent location [34]. Hemangiomas usually originate from the umbilical artery. They may cause non-immune fetal hydrops, IUGR, and severe fetal hemorrhage. The morbidity and mortality rate of umbilical cord hemangiomas have been reported to be approximately 35% [35]. To decrease the rate of intraterine and postnatal complications, an early diagnosis is necessary. The ultrasound examination with color Doppler is recommended [36,37]. With the advancement of technology and the use of ultrasound and maternal serum alpha-fetoprotein evaluation in the second trimester, it may be possible to diagnose hemangiomas of the umbilical cord in early gestation [30].

**Knots: True and False**

Another cord abnormality is a true knot. It is observed in approximately 0.3–2.1% of births [38]. Although its incidence is rare, this finding is associated with serious consequences. The fetal mortality rate is significantly higher than in the general obstetric population [4]. It can also be associated with a non-reassuring fetal heart rate pattern during labor and a higher incidence of the cesarean section. Some obstetric factors (e.g., polyhydramnios, gestational diabetes, fetuses that are small for the gestational age, long umbilical cord, male fetus, and genetic amnioentes) are correlated with a true knot [38]. The prenatal diagnosis of a true knot is possible with US gray scale, color Doppler, two- and three-dimensional power Doppler, and 3D US with HD flow. The suspected area on gray-scale ultrasound is visualized as a “loop”. A confirmation can be achieved by acquiring a color Doppler three-dimensional volume and rotating the volume along the x- and y-axes until a full view of the knot has been displayed [39,40].

True knots should be distinguished from false knots, which are not clinically significant. However, the diagnosis is rarely given prenatally. In a 5-year study by Bohiltea, 133 (0.71%) of 18 500 deliveries were diagnosed with a true umbilical cord knot, and only 16 (0.08%) of the cases were diagnosed by antepartum
ultrasound [41]. Nevertheless, it is unusual for a knot to tighten before the onset of labor, so the decision to perform an earlier delivery in these cases should be taken individually.

**Body Cord and Nuchal Cord**

The body cord is recognized when the umbilical cord encircles any fetal body part. Most often the loop is located on the fetus’s neck, less often around the trunk or limbs. It is estimated that the umbilical cord loop appears in every third pregnancy. A nuchal cord occurs when the umbilical cord becomes wrapped around the fetal neck 360 degrees (Figure 5). The prevalence of a nuchal cord at delivery has been reported to range between 6% and 37% [42]. The risk factors for a nuchal cord are: reduced amniotic fluid, increased fetal activity, and advanced gestational age [41,42]. The prevalence of a single, double, triple, and quadruple nuchal cord was described by Sherer et al. to be 10.6%, 2.5%, 0.5%, and 0.1%, respectively [43]. In the vast majority of cases, the cord loop does not exert any negative effects, but it can cause serious consequences such as intra-antenatal hypoxia or even an intrauterine death. It has been proved that the risk of serious complications increases only if there are 3 or more loops.

The prenatal diagnosis of a nuchal cord is not routinely recommended, although there is a correlation with an abnormal fetal heart rate pattern during labor [42]. The visualization of the sagittal and transverse section of the fetal neck with the use of color Doppler and/or 3D US allows precise diagnosis [40]. In certain cases, it is possible to observe the “divot sign” described by Ranzim et al. [44].

The complications described above are linked to clamping or a significant constriction of the umbilical vessels and multiple monoamniotic pregnancy. A common fluid space, in which fetuses and umbilical cords float freely, promotes the creation of mutual loops and nodes. Perinatal mortality reaches 40% in these pregnancies. Perinatal mortality can be decreased using only strict biophysical monitoring of fetuses and planned caesarean sections in the third trimester after prior induction of fetal lung maturation.

**Inappropriate Number of Umbilical Cord Vessels**

Initially there are 4 vessels in the umbilical cord. The right umbilical vein usually disappears at around 6–7 weeks of pregnancy, which leads to the formation of the 3-vessel
umbilical cord. The presence of 4 vessels in the umbilical cord is an extremely rare anomaly [45]. The most common anomaly, present in 0.2–1% of pregnancies, is the umbilical cord containing only 2 vessels: 1 artery and 1 vein (SUA – single umbilical artery) [46]. Usually, the left arterial vessel is missing [47]. The umbilical artery, which is a branch of the right or the left common iliac artery, is accompanied by the left umbilical vein. This is the most common type of SUA, found in 98% of cases. In only about 1.5% of cases, the umbilical artery goes away from the superior mesenteric artery. The rarest and the least promising variant is the SUA coexisting with 2 umbilical veins or the right umbilical vein. In this case, the artery is usually twice as large as the umbilical cord, while in the proper 3-vein umbilical cord, this ratio is reversed [48].

SUA may be an isolated development anomaly; however, in approximately 30% of cases, it is accompanied by other structural disorders, and in approximately 10% of cases by genetic abnormalities [46]. The detection in pre-natal examinations of a single umbilical artery should therefore lead to a detailed ultrasound assessment of all organs and systems. SUA is primarily associated with the impaired musculoskeletal, cardiovascular, and the urogenital systems, and less frequently with the gastrointestinal and the central nervous systems [49,50]. SUA increases the risk of restricting intrauterine growth in fetuses. The observations of fetuses in multiple pregnancies indicated a lower birth weight in the majority of fetuses with SUA. Moreover, the analyses of fetuses coming from single pregnancies confirm that, in 20–30% of cases, SUA is conducive to the development of IUGR [49].

Prenatally, the intra-abdominal course of umbilical arteries, which surround the bladder from both sides, can be visualized as early as the first trimester of pregnancy by using Color Doppler techniques (Figure 6). It is necessary to follow the entire intra-abdominal course of the arteries from the navel upwards to avoid confusion with femoral arteries, which may be extremely important in early pregnancy. The detection of SUA in the first trimester requires a confirmation of the diagnosis at a later stage of pregnancy. Then, the number of veins in the umbilical cord can be determined on the cross-section of a free loop, at a suitable distance from the placenta insertion; in the cross-section, the appearance is that of “Mickey Mouse” (Figure 6). In the vicinity of the insertion, there is a possibility of connecting the arterial blood vessels (Hyrtl anastomosis) and making a wrong diagnosis.

**Persistent Right Umbilical Vein (PRUV)**

PRUV is a rare development anomaly occurring in 0.08–0.5% of pregnancies [51]. There are 2 types of PRUV: with the intrahepatic course (PRUV-I) and with the extrahepatic ones (PRUV-E). PRUV-I is diagnosed in 95% cases [52]. It usually coexists with a proper venous duct, and has a good prognosis. If the venous duct runs extrahepatically, the DV is usually not formed, and the venous blood vessel is directly connected with the main lower limb vein or the right vestibule, which contributes to serious hemodynamic disturbances in the fetus, including generalized edema.

PRUV may be an isolated phenomenon or it can coexist with other malformations, especially with regard to PRUV-E [53]. PRUV is usually connected with the defects of the cardiovascular system (7.9%), placenta and umbilical cord defects (7%), defects of the urogenital system (6.3%), and defects of the central nervous system (3.8%). Genetic disorders occur very rarely since they are present in only 1.3% fetuses with PRUV [54].

In a typical pattern, the intra-abdominal course of the left vein may be shown on the cross section of the abdomen, slightly below the level at which the abdomen circuit is measured (AC), in the 2nd and 3rd trimester. This vessel runs to the left of the gallbladder, tangent to the greater sac, and turns to the right, being connected to the hepatic portal system. In case of PRUV, the vessel curves to the left, so that the gallbladder is placed in the middle, in between the vessel and the stomach. PRUV may therefore be visualized in B-mode; however, in diagnosis, Color-Doppler (Figure 7) and 3D imaging (Figure 8) are helpful. These techniques are particularly useful in confirming
the presence of the venous duct, which affects further prognosis [51].

**Fetal Intra-Abdominal Vein Varix (FIUVV)**

Fetal intra-abdominal vein varix is an extremely rare phenomenon, usually diagnosed during a routine ultrasound examination, in the 2nd and 3rd trimester. It is estimated that this phenomenon takes place in 0.4–1.1 in 1000 pregnancies [55].

Ultrasound varicose veins appear as oval elongated fluid spaces which run obliquely between the front wall of the abdomen and the fetal liver. Umbilical cord varix can be recognized when the vein diameter inside the abdominal cavity is by 50% larger than in the abdominal insertion place of the umbilical cord, or when it exceeds 9 mm [56,57]. For identification, it is sufficient to conduct the examination in the gray scale, although the color flow, power Doppler, or pulsed wave Doppler techniques may prove useful in the diagnosis and monitoring of varicose veins, particularly with regard to thrombosis.

Based on the analyses of the described few clinical cases, it can be concluded that FIUVV significantly increases the risk of an intrauterine death (44%) associated with thrombosis, especially within the dilated vessels [58,59]. They may also lead to IUGR and fetal edema, as well as being associated with improper genomes [58,59]. It is estimated that in approximately 19% of varicose veins cases, other development anomalies coexist [60].

The diagnosis of FIUVV should lead to increased concern for the wellbeing of the fetus, taking into account the ultrasound evaluation of veins with regard to thrombosis. Dilation of the vessel to above 12 mm, as well as a turbulent flow, are considered to be key risk factors of fetal death. In view of the significant risk of intrauterine death, most authors are of the opinion that, in the case of FIUVV, it is necessary to induce labor after 37 weeks of pregnancy [60].

**Ultrasonography of the Umbilical Cord in Prenatal Diagnostic Testing and Fetal Growth Abnormalities**

**Technical aspects of UA blood flow assessment**

Doppler US of UA demonstrates a high resistance flow until 20 weeks. With advancing gestational age, it decreases and results in an increasing diastolic flow. This knowledge helps with the use of S/D and PI in the clinical practice. Measurements of UA Doppler waveforms should be made in a free cord loop, during the absence of fetal breathing and movements. However, an absent or reversed end-diastolic flow is likely to be seen first at the fetal end of the UA, where the impedance is the highest. In accordance with the international regulations, the insonation angle should be adjusted to as close to 0° as possible [61].

Umbilical artery Doppler is a widely used method for evaluating fetuses with IUGR. It provides important information on placental dysfunction, which manifests itself as rising vascular resistance. In severe cases there is absent end-diastolic velocity (UA-AEDV) and reverse end-diastolic velocity (UA-REDV). Hidaka et al. observed a significant positive correlation between the UA pH and the last measured DV-PI. [62]. It is also well known that, especially in cases of an early onset of IUGR with either UA or DV absent or reversed end-diastolic velocities, there is an increased risk of a fetal death [63]. An assessment of the umbilical artery blood flow is used for the management of fetuses with early IUGR. The decision about the time of delivery should be made taking into account the gestational age, Doppler of UA, MCA, DV, cardiotocography, the biophysical profile, and the umbilical venous blood flow. Gill and Jouppila were
the first to suggest the clinical use of the umbilical venous blood flow [64,65]. Some investigators consider that the umbilical vein blood flow is a more direct measurement of the vascular placental function than the umbilical artery is. It seems to be an extremely useful parameter for determining the quantity of oxygen that reaches the fetus [66]. During the UV Doppler assessment in the physiological state, the blood flow should be continuous and non-pulsatile throughout the cardiac cycle. However, in the first trimester, pulsations may appear. Some authors suggest the use of a combination of the umbilical artery and the vein blood flow assessments for the identification of fetuses at an increased risk of the adversary perinatal outcome [67]. Parra-Saaerda et al. found that the evaluation of the uterine artery PI and the umbilical vein blood flow reflect the degree of placental underperfusion, which can remain undetected by umbilical artery blood flow [67]. Rigano et al. concluded that a reduction of the umbilical vein blood flow (due to a reduced UV velocity) is an early finding in intrauterine growth-restricted fetuses [66].

Umbilical Cord and Invasive Procedures

Cordocentesis is a crucial prenatal diagnostic procedure, which has been performed for 35 years. The percutaneous umbilical blood sampling is a procedure which provides direct access to the fetal circulation. Apart from all the medical procedures, it is associated with some complications, preterm birth, and fetal loss in 0.9–3.2% of cases. The most common indication for cordocentesis is diagnosing and treating fetal severe anemia. A number of historical indications for cordocentesis has been replaced by safer tests and procedures [Society for Maternal-Fetal Medicine (SMFM)]. Srisupundit et al. concluded that cordocentesis negatively affects fetal hemodynamics, as indicated by temporary worsening of the Tei index [68].

Conclusions

The examination of the umbilical cord is an important part of prenatal ultrasonography and it provides information on fetal development and wellbeing. The cord abnormalities may be associated with fetal structural (e.g., cardiovascular) and chromosomal anomalies. The detection of cord abnormalities, especially fetal or placental umbilical insertions, should prompt a thorough fetal US examination. Recent studies prove that modern 3-D and Doppler techniques allow a more precise diagnosis of abnormalities, as well as an evaluation of conditions that could result in fetal growth restriction and even fetal death. Early diagnosis and surveillance can minimize fetal mortality and help make decisions. The isolated cord abnormalities usually have a favorable prognosis.

Conflict of interest

None.

References:

1. Ferguson VL, Dodson RB: Bioengineering aspects of the umbilical cord. Eur J Obstet Gynecol Reprod Biol, 2009; 144: 108–13
2. Dudiak CM, Salomon CG, Posniak HV et al: Sonography of the umbilical cord. Radiographics, 1995; 15(5): 1035–50
3. Mosliri M, Zaidi SF, Robinson TJ et al: Comprehensive imaging review of abnormalities of the umbilical cord. Radiographics, 2014; 34(1): 179–96
4. Rodriguez N, Angarita AM, Casasbuenas A, Sarmiento A: Three-dimensional high-definition flow imaging in prenatal diagnosis of a true umbilical cord knot. Ultrasound Obstet Gynecol, 2012; 39(2): 245–46
5. Hill LM, DiNofrio DM, Guzick D: Sonographic determination of first trimester umbilical cord length. J Clin Ultrasound, 1994; 22: 435–38
6. Rayburn WF, Beynen A, Brinkman DL: Umbilical cord length and intraintrum complications. Obstet Gynecol, 1981; 57(4): 450–52
7. Murphy A, Platt LD: First-trimester diagnosis of body stalk anomaly using 2- and 3-dimensional sonography. J Ultrasound Med, 2011; 30(12): 1739–43
8. Smrecik JM, Germer U, Krokowski M et al: Prenatal ultrasound diagnosis and management of body stalk anomaly: Analysis of nine singleton and two multiple pregnancies. Ultrasound Obstet Gynecol, 2003; 21(4): 322–28
9. Mittal A, Nanda S, Sen J: Antenatal umbilical imaging as a predictor of perinatal outcome. Arch Gynecol Obstet, 2015; 291: 763–68
10. Narayan R, Saaid R, Pedersen L, Hyett J: Ultrasonography of umbilical cord morphology in the first trimester: A feasibility study. Fetal Diagn Ther, 2015; 38(3): 212–17
11. Verkleij CP, van Oppen AC, Mulder EJ et al: Evaluation of antenatal umbilical imaging index at 16–21 weeks of gestation as a predictor of trisomy 21 and other chromosomal defects. Ultrasound Obstet Gynecol, 2013; 42: 545–52
12. Gordon Z, Eytan Q, Jaffa A, Elad D: Hemodynamic analysis of Hyrtl anastomosis in human placenta. Am J Physiol Regul Integr Comp Physiol, 2007; 292: 977–82
13. Benirschke K, Kaufmann P: Placental shape aberrations. In: Baergen RN (ed.), Pathology of the Human Placenta. 4th ed. New York, Springer-Verlag, 2000: 399–414
14. Sepulveda W, Rojas I, Robert JA et al: Prenatal detection of velamentous insertion of the umbilical cord: A prospective color Doppler ultrasound study. Ultrasound Obstet Gynecol, 2003; 21(6): 564–69
15. Catanzarite V, Oyelese Y: Diagnosis and management of vasa previa. Am J Obstet Gynecol, 2016; 214(6): 764
16. Rebarber A, Dolin C, Fox NS et al: Natural history of vasa previa across gestation using a screening protocol. J Ultrasound Med, 2014; 33(1): 141–47
17. Prefumo F, IzzI C: Fetal abdominal wall defects. Best Pract Res Clin Obstet Gynaecol, 2014; 28: 391–402
18. Mastroliacono P, Lisi A, Castilla EE et al: Gastrochisis and associated defects: An international study. Am J Med Genet A, 2007; 143A: 660–71
19. Bonilla F Jr., Raga F, Villalaz E et al: Umbilical cord cysts: evaluation with different 3-dimensional sonographic modes. J Ultrasound Med, 2010; 29: 821–85
20. Zangen R, Boldes R, Yaffe H et al: Umbilical cord cysts in the second and third trimesters: Significance and prenatal approach. Ultrasound Obstet Gynecol, 2010; 36: 296–301
21. Ross JA, Jurkovic D, Zosmer N et al: Umbilical cord cysts in early pregnancy. Obstet Gynecol, 1997; 89: 442–45
22. Szepanjkowski K, Guzik P, Chechliński P et al: Pseudocyst of the umbilical cord-case report. Przegl Lek, 2015; 72(7): 394–96
23. Hellfetz SA, Rueda-Pedraza ME: Omphalomesenteric duct cysts of the umbilical cord. Pediatr Pathol, 1983; 1: 325–35
24. Rosenberg JC, Chevenak FA, Walker BA et al: Antenatal sonographic appearance of omphalomesenteric duct cyst. J Ultrasound Med, 1986; 5: 719–20
25. Sachs L, Fourcroy JL, Wenzel DJ et al: Prenatal detection of umbilical cord allantoic cyst. Radiology, 1982; 145: 445–46
26. Eggebo TM, Dirdal HU, von Brandis P: Allantoid cyst in the umbilical cord diagnosed with B-flow ultrasound. BMJ Case Rep, 2012; 27: 2012
27. Gualandri G, Rivasi F, Santunione AL et al: Spontaneous umbilical cord hematoma: An unusual cause of fetal mortality: A report of 3 cases and review of the literature. Am J Forensic Med Pathol, 2008; 29(2): 185–90
28. Clare NM, Hayashi R, Khodr G: Intrauterine death from umbilical cord hematoma. Arch Pathol Lab Med, 1979; 103(1): 46–28
29. Abraham A, Ratheore S, Gupta M, Santosh JB: Umbilical cord haematoma causing still birth – a case report. J Clin Diagn Res, 2015; 9(12): QD01–2
30. Göksever H, Celiloğlu M, Küpelioğlu A: Angiomyxoma: A rare tumor of the umbilical cord. J Turk Ger Gynecol Assoc, 2010; 11(1): 58–60
31. Takagi MM, Bussamra LC, Araujo Júnior E et al: Prenatal diagnosis of a true knot of the umbilical cord. J Obstet Gynecol Reprod Biol, 2014; 43: 360–64
32. Gualandri G, Rivasi F, Santunione AL et al: Umbilical cord: stenotic change of the umbilical vessels. Fetal Diagn Ther, 1999; 14(1): 318–23
33. Sendzegaard G: Hemangioma of the umbilical cord. Acta Obstet Gynecol Scand, 1994; 73(5): 434–36
34. Caldarella A, Buccoliero AM, Taddei A et al: Hemangioma of the umbilical cord: report of a case. Pathol Res Pract, 2003; 199(1): 51–55
35. Camitomo M, Sueyoshi K, Matsukita S et al: Hemangioma of the umbilical cord: stenotic change of the umbilical vessels. Fetal Diagn Ther, 1999; 14(6): 328–31
36. Hershkowitz R, Silberstein T, Shemer E et al: Risk factors associated with true knots of the umbilical cord. Eur J Obstet Gynecol Reprod Biol, 2001; 98: 36–39
37. Abraham A: Three-dimensional ultrasound with color Doppler imaging of the umbilical cord true knot. Ultrasound Obstet Gynecol, 2014; 43: 360
40. de Castro Rezende G, Araujo Júnior E: Prenatal diagnosis of placenta and umbilical cord pathologies by three-dimensional ultrasound: Pictorial essay. Med Ultrason, 2015; 17(4): 545–49
41. Bohilțea RE, Turcan N, Cirstoiu M: Prenatal ultrasound diagnosis and pregancy outcome in term small-for-gestational-age fetuses. Ultraschall Med, 2018 [Epub ahead of print]
42. Geipel A, Germer U, Welp T et al: Prenatal diagnosis of single umbilical artery: Determination of the absent side, associated anomalies, Doppler findings and perinatal outcome. Ultrasound Obstet Gynecol, 2000, 15(2): 114–17
43. Blackburn W, Cooley W: Umbilical cord. In: Stevenson RE, Hall JG (eds.). Human Malformations and Related Anomalies 2nd ed. New York, Oxford University Press, 1993; 1275–1350
44. Friebre-Hoffmann U, Hiltmann A, Friedl TWP et al: Prenatally diagnosed single umbilical artery (SUA) – retrospective analysis of 1169 fetuses. Ultraschall Med, 2018 [Epub ahead of print]
45. Geipel A, Germer U, Welp T et al: Prenatal diagnosis of single umbilical artery: Determination of the absent side, associated anomalies, Doppler findings and perinatal outcome. Ultrasound Obstet Gynecol, 2000, 15(2): 114–17
46. Boggs E, Meeks M, Welsch J et al: Anomalies of the umbilical cord. Ultrasound Obstet Gynecol, 2009; 36: 93–111
47. Blazer S, Zimmerman EZ, Bronstein M: Persistent infrahepatic right umbilical vein in the fetus: A benign anatomic variant. Obstet Gynecol, 2000; 95: 433–36
48. Lide B, Lindsley W, Foster MI et al: Intrahepatic persistent right umbilical vein and associated outcomes: A systematic review of the literature. J Ultrasound Med, 2016; 35: 1–5
49. Beraud E, Rozel C, Milon J, Darnault P: Umbilical vein varix: Importance of ante- and post-natal monitoring by ultrasound. Diagn Interv Imaging, 2015; 96(1): 21–26
50. Allen SI, Bagnall C, Roberts AB, Teele RL: Thrombosing umbilical vein varix. J Ultrasound Med, 1998; 17: 189–92
51. Sepulveda W, Mackenna A, Sanchez J et al: Fetal prognosis in varix of the intrafetal umbilical cord. J Ultrasound Med, 1998; 17: 171–75
52. Fung TY, Leung TN, Leung TY, Lau TK: Fetal intra-abdominal umbilical vein varix: What is the clinical significance? Ultrasound Obstet Gynecol, 2005; 25(2): 149–54
53. Mahony BS, McGahan JP, Nyberg DA, Reisner DP: Varix of the fetal intra-abdominal umbilical vein: Comparison with normal. J Ultrasound Med, 1992; 11(2): 73–76
54. di Pasqua E, Kuleva M, O’Gorman N et al: Fetal intra-abdominal umbilical vein varix: Retrospective cohort study and systematic review and meta-analysis. Ultrasound Obstet Gynecol, 2018; 51(5): 580–85
55. Bhide A, Acharya G, Bilardo CM et al: ISUG practice guidelines: Use of Doppler ultrasonography in obstetrics. Ultrasound Obstet Gynecol, 2013; 41(2): 233–39
56. Hidaka N, Sato Y, Kido S et al: Ductus venosus Doppler and the postnatal outcomes of growth restricted fetuses with absent end-diastolic blood flow in the umbilical arteries. Taiwan J Obstet Gynecol, 2017; 56(5): 642–47
57. Caradeux J, Martinez-Portilla RJ, Basuki TR et al: Risk of fetal death in growth-restricted fetuses with umbilical and/or ductus venosus absent or reversed end-diastolic velocities before 34 weeks of gestation: A systematic review and meta-analysis. Am J Obstet Gynecol, 2018; 218(25): 774–82
58. Gill RW, Trudinger BI, Garrett WJ et al: Fetal umbilical venous flow measured in utero by pulsed Doppler and B-mode ultrasound. I. Normal pregnancies. Am J Obstet Gynecol, 1981; 139(6): 720–25
59. Jopplia P, Kirkpin P: Umbilical venous blood flow as an indicator of fetal hydrops. Br J Obstet Gynaecol, 1984; 91(2): 107–10
60. Ferrazzi E, Rigano S, Bozzo M et al: Umbilical venous blood flow in growth-restricted fetuses. Ultrasound Obstet Gynecol, 2000; 16(5): 432–38
61. Parra-Saavedra M, Crovetto F, Triunfo S et al: Added value of umbilical vein Doppler in women with previous cesarean delivery. J Hum Hypertens, 2012; 26(8): 567–71
62. Parra-Saavedra M, Crovetto F, Triunfo S et al: Added value of umbilical vein Doppler in women with previous cesarean delivery. J Hum Hypertens, 2012; 26(8): 567–71
63. Caradeux J, Martinez-Portilla RJ, Basuki TR et al: Risk of fetal death in growth-restricted fetuses with umbilical and/or ductus venosus absent or reversed end-diastolic velocities before 34 weeks of gestation: A systematic review and meta-analysis. Am J Obstet Gynecol, 2018; 218(25): 774–82
64. Parra-Saavedra M, Crovetto F, Triunfo S et al: Added value of umbilical vein Doppler in women with previous cesarean delivery. J Hum Hypertens, 2012; 26(8): 567–71