ABSTRACT

The umbilical cord is not an inert structure, suspended between the fetus and placenta, but it plays an active role and it is involved in several processes affecting the fetoplacental unit.

Its study has to be regarded not only its morphology and morphometry, and the impedance of blood flow by Doppler waveform analysis, but it includes also an analysis of the coiling type and the amount of the Wharton Jelly. The umbilical cord has been considered like an important and huge source of informations, useful to assess the well-being of the fetus and the outcome of pregnancy.

The standardization of ultrasound techniques is the first step to speak the same language and make the study of this structure a fundamental part of well-being fetus assessment.

This article is carefully focused on morphologic, morphometric and functional ultrasound examination of umbilical cord and suggests that any anomaly detected should provide an indication for an intensive fetal follow-up, useful for early helpful therapy, preventing serious complication for the pregnancy.

Keywords: Umbilical cord, Wharton’s Jelly, Coiling index, Hyrtl anastomosis, Reynolds’ hypothesis.

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INTRODUCTION

For several decades, the morphological and morphometric aspects of the umbilical cord have been studied and retrospectively correlated with the perinatal outcome by pathologists after delivery. The advent of ultrasound has increased our knowledge and added a dynamic form of information in particular on the development of the fetus and its supporting structures such as the placenta and the umbilical cord. However, at the beginning, the umbilical cord has received only little interest mainly due to the limited resolution of the initial ultrasound machines. Indeed, the prenatal sonographic morphologic investigation of the umbilical cord has for long time been limited to the assessment of the number of vessels and later to the evaluation of the impedance to blood flow by Doppler waveform analysis.

However, an increasing body of clinical and experimental evidences show that both prenatal morphology and morphometry of the umbilical cord and its vessels may help in understanding the physiology of development as well as adaptive processes of the fetoplacental unit to pathologic insults. Moreover, studying the umbilical cord may in some circumstances help in the prediction of adverse pregnancy outcome. In the last decade, a considerable amount of scientific work has been published on this topic. We have learned that the umbilical cord is not an inert structure which is suspended between the fetus and placenta but is actively involved in important processes, such as fetal growth restriction, preeclampsia, diabetes, stillbirth and chromosomal defect or genetic syndromes.1-5 The aim of this chapter is to evaluate the role of the sonographic assessment of the umbilical cord during fetal life.

MORPHOLOGY

A normal umbilical cord at term is about 50 to 60 cm long and its surface is covered by a single layer of amniotic epithelium. The ground substance, in which three vessels—two arteries and one vein—are embedded, is called Wharton’s jelly. The characteristic structure of the umbilical cord is determined by the helical course of the arteries around the vein. Between the fetal umbilical ring and the placental insertion, the vessels fulfill usually 10 to 11 coils. This structure is very dynamic, as its morphology is influenced by a number of factors including gestational age, amount of amniotic fluid and its composition, fetoplacental hemodynamics as well as maternal complications during pregnancy. The evaluation of the umbilical cord can be accomplished either from the long-axis view or from a cross-sectional view. Probably the latter method is more appropriate because it allows quantification not only of the umbilical vessels’ size but also of the amount of the Wharton’s jelly.

What can be Observed in an Umbilical Cord?

How can we Read each Sign?

Basically, the morphology and morphometry of the umbilical cord is influenced by external factors and/or by factors which are inherent to the cord itself. Nomograms for the diameter of the umbilical vessels have been reported by Weissman and Raio,6,7 showing that the diameter of the umbilical arteries increases from 1.2 ± 0.4 mm at 16 weeks to 4.2 ± 0.4 mm at term of gestation and the umbilical vein diameter varies from 2.0 ± 0.6 mm at 16 weeks of gestation to 8.2 ± 0.8 mm at the term of gestation. These nomograms have showed that the diameter increases as a function of gestational age, progressively up to 32 weeks of gestation followed by a plateau toward the end of the pregnancy due to a reduction of water content of the Wharton’s jelly.
Moreover, a significant relationship between umbilical cord diameter and cross-sectional area and fetal anthropometric parameters (biparietal diameter, femur length, abdominal circumference) has been described. Experimental and clinical evidence suggest that Wharton’s jelly plays a metabolically active role throughout pregnancy; Vizza et al reported that the collagen fibrillar network of the Wharton’s jelly, studied by scanning electron microscopy, shows the presence of a wide system of interconnected cavities consisting of canalicular-like structures as well as cavernous and perivascular spaces. Considering that the Wharton’s jelly lack of a proper vasculature, this system of cavities may have an important role facilitating a bidirectional transfer of water and metabolites between amniotic fluid and umbilical cord vessels through the Wharton’s jelly. Moreover, Wharton’s jelly cushions umbilical blood vessels, preventing disruption of flow due to compression or bending caused by fetal movements and uterine contraction, i.e. at delivery. Modifications in the amount and composition of Wharton’s jelly of three-vessel cords have been described in a number of pathological conditions, usually associated with a modification of the amniotic fluid volume and composition, occurring in pregnancy (i.e. hypertensive disorders, gestational diabetes). The reduction of the amount of Wharton’s jelly may be the consequence of either an extracellular dehydration or a reduction in extracellular matrix component.

The sonographic cross-sectional area of Wharton’s jelly can be computed by subtracting the vessels area from the cross-sectional area of umbilical cord. The umbilical vein and arteries areas are to be computed at the maximal magnification using the software of the ultrasound machine. A reference range for the total vascular area has been generated by Weissman and colleagues.

The umbilical cord can be defined as:

- **LEAN**, if its sonographic cross-sectional area is below the 10th percentile for gestational age (Fig. 1), and
- **LARGE** if its sonographic cross-sectional area is above the 90th percentile for gestational age.

### ‘LEAN’ UMBILICAL CORD

Just over 40 years ago, observing two cases of macroscopically thin umbilical cord (UC) associated with stillbirth and fetal distress, Hall stated that ‘The thin cord is a dangerous cord and a fat cord is a safe cord, all other factors being equal’.

Pathologic studies and case reports demonstrated that a lean umbilical cord is associated with adverse pregnancy outcome, oligohydramnios and fetal distress. Raio et al found an association between the presence of a ‘lean’ umbilical cord and the delivery of a small-for-gestational-age infant. Patients with a ‘lean’ UC after 20 weeks of gestation had a 4.4-fold higher risk (95% confidence interval, 2.16-8.85) of having an SGA infant than those with a normal umbilical cord. Wharton’s jelly appears to serve the function of adventitia, which the UC lacks, binding and encasing the umbilical vessels. It has been speculated that the cells of Wharton’s jelly appear to possess contractility comparable to that of smooth muscle cells and participate in the regulation of umbilical blood flow and that, at least in some cases, the reduction in fetal growth could be the consequence of Wharton’s jelly decrease leading to hypoplasia of the umbilical vessels. In fact, a reduction of wall thickness of umbilical cord arteries and vein has been found in intrauterine growth retardation (IUGR) infants with abnormal umbilical artery flow when compared to IUGR infants without increased umbilical artery resistance. Cumulative evidence suggests that an umbilical cord less than 10th centile for gestational age is a simple and early marker SGA infant and the occurrence of intrapartum complication. Moreover, a lean UC is frequently associated with signs of fetal distress at the time of delivery (oligohydramnios, low Apgar score and meconium-stained amniotic fluid).

### LARGE UMBILICAL CORD

Several reports in the literature have described a large UC associated with other fetal structural anomalies, such as umbilical cord tumor, urachal cysts, umbilical cord mucoid degeneration and omphalomesenteric cyst. Generally, in these conditions, the morphology is altered in a limited portion of the umbilical cord (Figs 2A and B).

However, a consistent association between an ultrasonographic large UC and the presence of a gestational diabetes mellitus has been reported. A large UC can be considered as an additional parameter useful to identify fetuses of a mother with some kind of glucose intolerance during pregnancy. Fetuses of patients with gestational diabetes have a larger UC and this is mainly due to a higher content of Wharton’s jelly. Weissman and Jakobi found an
alteration in the distribution of Wharton’s jelly fibers with large empty spaces among them and speculated that this could be caused by an abnormal accumulation of fluid and plasma proteins within the Wharton’s jelly, resulting in an increased surface area and in an increased permeability and hemorrhages due to an increased oncotic pressure in the interstitial spaces of the Wharton’s jelly\(^{18,19}\) (Figs 3A and B). This modification can be observed at 24 gestational weeks, suggesting that the involvement of the umbilical cord in fetuses of diabetic mothers is a phenomenon that occurs early in pregnancy.

In addition, it has been shown that sonographic assessment of umbilical cord area may improve the prediction of fetal macrosomia; although ultrasound remains imprecise in the recognition of fetal macrosomia, obstetrician should not shun the use of biometric methodology to assist in the management of suspected macrosomia, but rather should look forward to further improvements that will enhance its accuracy as a diagnostic tool. Umbilical cord area is an easily obtained sonographic measurement, with highly reliable intraobserver and interobserver reproducibility. The time required to obtain an adequate and satisfactory image of cross-section of UC is about 2 minutes. A large cross-sectional area of the UC performs poorly by itself as a predictor of fetal macrosomia. A sonographic large UC can be used in addition to estimated fetal weight (EFW) as a further marker that may facilitate the detection of fetal overgrowth, potentially improving the performance of ultrasound-based policies for the management of suspected macrosomia.\(^5\)

**DISCORDANT UMBILICAL ARTERY**

Discordance between the umbilical arteries is considered to be present when the difference between the diameter of the two arteries is at least 1 mm in three different portions of the UC in both transverse and longitudinal section\(^ {20,21}\) (Figs 4A to D).

Moreover, these arteries are also characterized by differences in the impedance to blood flow measured by Doppler flow methods with usually a higher resistance index measured in the smaller artery.\(^21\)

Therefore, the information provided by Doppler velocimetry of the smaller umbilical artery should be taken with caution, because the significance of high-resistance patterns observed in other populations seems to represent a more benign condition in patients with discordant umbilical arteries. Therefore, from a clinical point of view, the presence of discordant umbilical artery seems to be a benign condition that does not affect the development of the fetus.

However, cases with discordant arteries are associated with a higher incidence of morphologic placental alterations (placenta bipartite, placenta succenturiata, absence of Hyrtl anastomosis) and anomalous placental insertion (marginal, velamentous). These placental anomalies are similar to those frequently seen in cases of single umbilical artery (SUA) supporting the theory that the presence of a single artery represents the greatest expression of umbilical artery discordance.\(^{22,23}\) The presence of the Hyrtl anastomosis is a common feature of the vascular system in the human placenta, present in at least 95% of all placentae. This anastomosis is the only vessel that connects the umbilical arteries or their branches on the placental surface, close to
the site of cord insertion, playing an active role in equalizing the blood pressure between the territories supplied by each umbilical artery; in fact, although the areas supplied by each of the umbilical arteries may show great discrepancy, the corresponding UC arteries are usually of equal caliber (Fig. 5). This equalizing effect might be of utmost importance in particular during uterine contractions when the blood pressure and resistance in the corresponding portion of the intervillous space and cotyledons may differ in different parts of the placenta (Figs 6 and 7). Discordance in calibers of the umbilical arteries has been postulated to be the consequence of a failure of the Hyrtl anastomosis to develop anatomically or to function fully. With the advent of more sophisticated ultrasound equipment, the morphologic and functional evaluation of this vessel has now become possible. The absence of a Hyrtl anastomosis has recently been associated with the presence of discordant umbilical arteries; similarly, abnormal umbilical cord insertion, such as marginal or velamentous cord insertion has also been associated with a missing Hyrtl anastomosis and discordant umbilical cord arteries.24,25

**SINGLE UMBILICAL ARTERY (SUA)**

The incidence of SUA is reported to be 0.5 to 2.5% in uncomplicated neonates, but is higher in aborted (1.5-7%) and aneuploid fetuses (9-11%). Multiple gestations have a three to seven-fold increased risk of SUA. Fetuses whose umbilical cord has a single artery are at increased risk of intrauterine and intrapartum death, regardless of the

![Fig. 5: Doppler ultrasound demonstration of Hyrtl anastomosis](image)

![Fig. 6: Hyrtl anastomosis](image)

![Figs 7A and B: 3D view of Hyrtl anastomosis](image)
presence or not of congenital or chromosomal malformations. Most cases of SUA are diagnosed in the late second trimester (Figs 8A and B). Despite the apparently easy recognition of SUA, a low sensitivity of ultrasound is reported. Color Doppler imaging allows earlier and more confident diagnosis of SUA, but its apparent efficacy has to be proven. The patent artery is usually larger than normal and it may approximate to the vein diameter. It has been estimated that the risk of anomalies is seven times greater than in infant with three-vessels cord. The list of anomalies identified to be associated with SUA is long. Persutte and Hobbins22 divided the reported abnormalities into three groups:

1. To be identified with prenatal ultrasonography
2. To be difficult to be identified prenatally
3. To be unidentifiable prenatally.

Using these criteria, they conclude that prenatal ultrasonography can consistently identify only 37% of fetal anomalies associated with SUA. This low accuracy should well be kept in mind when counseling a patient with a fetus affected by SUA. The prognosis of SUA infants is mainly related to associated fetal structural or chromosomal anomalies and the frequently present intrauterine growth retardation. The lower amount of Wharton’s jelly present in two-vessels cord could be responsible of a higher vulnerability of the UC during the third trimester of pregnancy and during labor. The elevated incidence of stillbirth at the end of pregnancy in patients with a SUA may be in part explained by the cumulative effect of the relative Wharton’s jelly reduction that occurs physiologically in the third trimester of pregnancy, acting on a constitutional deficiency of jelly in umbilical cord with a single artery.26 It is likely that the amount of Wharton’s jelly at earlier stages of pregnancy exerts a sufficient protection to the vessels without affecting blood flow and therefore fetal growth. The SUA fetuses have a lack of the safety warranted by the presence of Hyrtl’s anastomosis, a safety valve and this fact partially explains the increased rate of unexplained intrauterine fetal demise in the third trimester of gestation and during labor.

**UMBILICAL CORD ANGIOARCHITECTURE**

Although the origin and significance of the umbilical cord angioarchitecture has been the subject of extensive research, the developmental process and functional importance of this vascular coiling are not fully understood.

Regardless of its origin, data from both pathologic and ultrasonographic investigations suggest that umbilical coiling is well established as early as 8 weeks of gestation, and the total number of coils at the end of the first trimester is similar to that observed in fully term cords. Moreover, the direction of twist is not randomly determined, since several investigators have found a clear prevalence of left-twisted umbilical cords.

Color flow mapping could be used to enhance the definition of the umbilical cord vascular architecture. The sonographic assessment of the coiling pattern is performed in different UC segments in order to exclude segmental morphologic anomalies (i.e. false knots, varices). The length of one complete umbilical vascular coil (distance between the right outer surface of consecutive arterial coils) is measured in a longitudinal midsection of the UC and a mean of three measurements is used for analysis. The sonographic umbilical coiling index (UCI) is defined as the reciprocal value of that measurement and it represents the number of vascular coils in a given cord. According to the umbilical coiling pattern the UC can be classified as:

- **Normal**
- **Uncoiled** (two straight umbilical arteries with an umbilical coiling angle equal to zero)
- **Hypocoiled**, if the UCI is below the 10th percentile for gestational age
- **Hypercoiled**, if the UCI is above the 90th percentile for gestational age
- **Atypical coiling** (Figs 9A to D):
  - Uncoordinated coiling or bizarre, or aperiodic coiling pattern, if there is an atypical coiling, in which the absence of a repetitive pattern does not allow the measurement of the UCI
  - **Supercoiling**, in the presence of a spring spatial configuration of the UC (Figs 10 to 12).

The only reference in the pathologic literature on anomalous helical patterns dates back to Hyrtl and Malpas and Symonds,27,28 which described in their postnatal series some ‘complicated’ cords with different directions of twists, occurring in different segments or a combination of coiled and uncoiled portions.

Compelling evidence has demonstrated a correlation between abnormal umbilical coiling pattern and suboptimal pregnancy outcome in singleton pregnancy. The
mechanisms leading to the coiling of the UC are largely unknown and whether it represents a genetically determined or an acquired phenomenon is still the subject of debate. Since the etiology of the vascular coil in normal UC is still an enigma, it is even more intriguing to find a plausible pathophysiologic explanation for an abnormal umbilical cord angioarchitecture. A variety of hypotheses have been advanced to explain the origin of umbilical vascular coiling, including fetal movements, unequal umbilical vascular growth rate (Roach), fetal hemodynamic forces, umbilical vascular wall mechanism and genetics factors. Since the helical course of umbilical vessels is established by 9 weeks of gestation, a hemodynamic imbalance leading to unequal cord morphology is supposed to occur very early in gestation. As this typical repetitive vascular pattern of the umbilical cord is fully established at the end of the first trimester, uncoordinated coiling may be the result of an abnormal coiling process, which takes place during early pregnancy (Figs 13A and B). It has been postulated that coiling is determined by an interaction between the intrinsic properties of the fibers in the umbilical cord vessels wall and hemodynamic forces acting on it during development.

Data from clinical and experimental studies show that the umbilical cord is a dynamic structure where both hemodynamic factors and gestational age influence the vascular umbilical cord pattern. Before the fetal kidneys start the excretion of significant amount of urine, fetal membranes are involved in fetal fluid accumulation and regulation by transmembranous mechanism. In early gestation, the fluid exchange between umbilical vessels and amniotic fluid is facilitated by the limited amount of Wharton’s jelly wrapped around the umbilical cord vessels. Reynolds et al postulated that the UC is a pistonless pulsometer pumping system acting as a cardiac assist pump to sustain the venous return from the placenta. The fetal
blood flows through the umbilical vein pumped by slight but definite decreases and increases in venous pressure that are generated from the force of the rising limb of the arterial pressure pulse. The presence of arterial coils that surround the vein along the length of the cord provide multiple variations in an additive fashion; the presence of vascular coils plays a central role in determining the blood flow from the placenta to the fetus. This mechanism is of utmost importance in early gestation when the placental resistance to blood flow is particularly elevated. Therefore, a reduced number of coils in lean umbilical cords could be responsible for a reduced umbilical blood flow which in turn leads to a fetal growth impairment.

Reynolds’ hypothesis, according to which the umbilical coils serve as a peristaltic pump mechanism enhancing the venous return to the fetus, has been advocated by several authors to explain how an abnormal coiling could influence the perinatal outcome.

Alterations in morphology and ultrastructure of UC components have been described in pregnancy complications that affect fetoplacental hemodynamics (Figs 14A to D). In particular, a high frequency of uncoiled and hypocooiled cords has been reported in intrauterine growth restriction and maternal hypertensive disorders. According to Poiseuille’s law, the three factors that might influence blood flow are the caliber of the vessel, the blood flow velocity and the viscosity of the blood. Di Naro stated that the vein blood flow is lower in fetuses with an umbilical cord cross-sectional area below the 10th centile than in those with an umbilical cord of normal caliber and the risk to
have an IUGR is very high. The alterations of vein blood flow can be detected earlier than arteries. The umbilical vein area and the umbilical vein blood flow are significantly reduced in growth restricted fetuses compared to normally grown fetuses. The discrepancy in the umbilical vein size might represent an adaptive response to venous overload on the one hand and chronic hypovolemia on the other hand. Supercoiling can be associated with pathologic fetal intra-abdominal process and may be explained by a relative increase in resistance at the level of the umbilical ring, which in turn induces a venous congestion of the extra-abdominal umbilical vein (Fig. 15). Kilavuz and Skulstad were able to show that blood velocity in the umbilical vein at the abdominal wall is higher than that in the extra-abdominal portion of the umbilical cord and that this increase in velocity is due to a progressive tightening of the fetal umbilical ring starting after that the physiologic midgut herniation is completed at 12 weeks of gestation.

UMBILICAL CORD AND ANEUPLOIDIES

A number of studies have reported a higher incidence of umbilical cords with decreased coiling index or even absence of coils in fetuses with aneuploidy. The UC extracellular matrix is a tissue composed by a high amount of glycosaminoglycans. The Wharton’s jelly is composed of an insoluble fibrillar network of different collagen types within which soluble open-coil polysaccharides are held. Hyaluronan, the most represented glycosaminoglycan in the Wharton’s Jelly, is known to influence cell behavior and to play a crucial role in angiogenesis, morphogenesis and tissue remodeling especially during embryogenesis. Hyaluronic acid can entrap large amount of water. A smaller part of the Wharton’s jelly extracellular matrix is formed by sulfated glycosaminoglycans, which, in turn, are linked to proteins to form proteoglycans. An alteration of the extracellular matrix has been indicated as one of the possible causes of increased nuchal translucency in trisomy 21 human fetuses. The variation in the amount of hyaluronan found in the skin of trisomy 21 fetuses may be present in the extracellular matrix of umbilical cords influencing their macroscopic appearance. There is evidence that fibroblast synthesis of hyaluronic acid is not different between healthy and trisomy 21 fetuses. Fibroblast of fetuses with trisomy 21 overexpress collagen type VI and experimental evidence has been provided that an inverse correlation exists between collagen synthesis and hyaluronan degradation. An increase in collagen type VI may contribute to hyaluronan accumulation. There is evidence that a reduced turnover of hyaluronan could also influence the growth of the umbilical cord vessels and coiling formation.

CONCLUSION

The antenatal measurement of umbilical cord area is probably a better parameter than determination of umbilical cord diameter to identify fetuses at risk of being small for...
gestational age at delivery or of having distress in labor, or to identify macrosomic fetuses born diabetic mothers. Since the UC area is easy to measure and nomograms are available, its measurement should be part of a routine scan and should prompt a careful and thorough evaluation whenever, there is a discrepancy between the observed and the normal values.

REFERENCES


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