Introduction

The umbilical cord (UC) is the essential life-sustaining connection between fetus and placenta. It constitutes a stable connection to the fetomaternal interface, while allowing fetal mobility that is of great importance for fetal development in general and fetal neuromotor development in particular. This combination of mechanical stability and flexibility is due to the architecture of the UC. There is however a range of umbilical cord complications that may be life threatening to the fetus and these too can be explained to a large extent by the cord’s structural characteristics. This review article discusses clinically relevant aspects of UC ultrasound.

 development and Architecture of the Umbilical Cord

In its embryonic stage the UC develops in the region of the body stalk to become the embryo’s connection to the fetal portion of the placenta (fetal placenta). The amniotic cavity expands from dorsal to ventral while the chorionic cavity shrinks in volume. During cephalo-caudal and lateral folding the early UC arises as it is “enveloped” by the expanding amnion (Fig. 1) (see also textbooks of embryology, www.embryology.ch). In this early stage at around 7–8 weeks postmenstrual age the UC contains the body stalk with umbilical vessels as well as other structures that will later regress and disappear entirely: the allantoic diverticulum (an outpouching from the endoderm connected to the (future) urinary bladder, later the urachus), as well as the extra-embryonic coelom that at this early stage still forms a connection to the cho-
The Hyrtl anastomosis have been described in association with absence of the Hyrtl anastomosis [4]. Anatomical variants of described in association with cord insertion pathologies (see below) appears to be of particular importance during uterine contractions. This pressure regulation mechanism ap-

duced. Normal values for UC diameter and cross-sectional area of fetoplacental circulation. They share a close anatomical rela-
tionship within the UC, their lumens being connected by the Hyrtl anastomosis. Named after Josef Hyrtl who first described it, this anastomosis is usually located within the last 3 cm of the cord before its placental insertion and serves to compensate for gestational age at diagnosis is discussed in detail below.

**The First Trimester**

**Umbilical cord length, body stalk anomaly**

Umbilical cord length varies, with a range of 35–70 cm at birth being regarded as normal. Both shorter and longer cords are associated with increased rates of intrapartum complications [13]. UC length is thus a significant factor when assessing fetal risk. Antenatal estimation of cord length is however extremely diffi-
cult. Traction force produced by fetal movements in the first tri-
mester is assumed to be the crucial growth stimulus of the cord [14]. A short UC is thus associated with reduced fetal movement and all its respective causes (e.g. malformation, myopathies, neuropathies, oligohydramnios).

The body stalk anomaly, with an incidence between 1:7500–1:31,000 depending on gestational age at diagnosis, refers to a...
particularly short UC associated with a high spontaneous abortion rate [15]. It can be detected by ultrasound as early as the first trimester (Fig. 2). It is characterised by multiple congenital anomalies including abdominal wall defect, absent or extremely short UC with possible single UA and direct attachment of the fetus to the placenta, and lower limb deformities. The term “limb bodywall complex” (LBWC), which is also used in literature, is probably best considered as a variant of the body stalk anomaly [16]. Body stalk anomaly quite commonly presents with fetal spinal deformity (kyphoscoliosis) and cranial defects as additional findings. Nuchal translucency and alpha-fetoprotein (AFP) in maternal serum are typically increased, while fetal karyotype is usually normal [17]. Anomalous development of the body stalk causes the syndrome, which explains the typical location of the lower limbs and abdominal organs in the extraembryonic coelom (chorionic cavity). The upper half of the fetus is located within the amniotic cavity. Some authors compare the pathology to an extreme form of amniotic band sequence, although various theories on its pathogenesis exist.

The differential diagnosis includes other syndromes associated with ventral body wall defects (Table 1).

Table 1  Differential diagnosis of abdominal wall defects.

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Structural abnormalities</th>
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<tbody>
<tr>
<td>Body stalk anomaly</td>
<td>Ventral clefts, short or absent umbilical cord, fetal position partially in extraembryonic coelom, with or without other malformations (cranial, vertebral)</td>
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<tr>
<td>Limb Body Wall Complex</td>
<td>As above with lower limb abnormalities</td>
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<tr>
<td>Pentalogy of Cantrell</td>
<td>Abdominal wall defects (omphalocele), lower sternal defect/celeft, anterior diaphragmatic hernia, pericardial defect/ectopia cordis, cardiovascular malformations</td>
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<tr>
<td>OES complex</td>
<td>Omphalocele, bladder extrophy, Imperforate anus, caudal neural tube defects (spina bifida)</td>
</tr>
<tr>
<td>Gastrochisis</td>
<td>Defect usually to the right of the umbilicus, UC insertion normal, intestinal loops float freely in the amniotic cavity.</td>
</tr>
<tr>
<td>Bladder or cloacal extrophy</td>
<td>Defect at the umbilicus, parietal peritoneum forms hernial sac, various herniated abdominal organs.</td>
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Prognosis depends on the severity of the various abnormalities and is very poor to infaust [17]. Longer than average umbilical cords are associated with increased risk of UC loops, knots and cord prolapse. These complications are of greater clinical importance in the third trimester and will be discussed later.

**Fetal and placental insertion of the umbilical cord**

The ends of the UC are usually easy to demonstrate by ultrasound in the first trimester, given the relative abundance of space in utero at this stage. As gestational age increases, however, visualisation becomes increasingly difficult and may be impossible, especially when the placenta is posterior. Both ends of the UC may show abnormalities that have an impact on the subsequent management of pregnancy.

**Placental umbilical cord insertion**

In addition to the “normal” central and paracentral umbilical cord attachments, insertion anomalies with more peripheral cord attachment occur. A cord insertion of less than 2 cm from the edge of the placenta is clinically significant. In marginal cord insertion (5–7% of pregnancies) the UC arises directly from the edge of the placenta [11, 18]. Anomalous cord insertion is associated with an increased rate of intrauterine growth restriction so that fetal monitoring should be intensified if it is detected antenataly [19]. The UC insertion can be found sonographically using colour Doppler by following the chorionic plate vessels. If demonstrated, the UC insertion is no longer possible, which may be the case as gestation advances, tangential visualisation of the placental surface may be helpful to obtain an overview (Fig. 3). Using this technique the chorionic plate vessels can be demonstrated by colour Doppler as they approach the UC insertion, creating a star-like pattern.

It is particularly important to diagnose velamentous cord insertion, a pathological insertion of the cord seen in 1–2% of pregnancies [11]. Here the origin of the actual UC lies outside the placenta (Fig. 4a) and consists of velamentous vessels (enveloped → here: between the amniotic membranes i.e. between
amnion and chorion). In contrast to normal UC vessels, velamentous vessels are not protected by the stabilising effect of Wharton’s jelly. There is an increased risk of cord compression, fetal growth restriction, premature labour, placental abruption, CTG abnormalities and low Apgar scores [11]. It is important to be aware that velamentous cord insertion is associated with an increased rate of vasa praevia.

The term vasa praevia describes velamentous vessels that lie in direct proximity to the cervix and below the fetal “presenting part” (Figs. 4b and d); the prevalence of this anomaly is 0.04% [20]. Risk factors for vasa praevia, apart from velamentous cord insertion (vasa praevia type I), include accessory placenta (vasa praevia type II) [21], and multiple pregnancy and pregnancy in the setting of assisted reproduction [22]. The closer the vessels are to the internal os (which exposes them to traction and shearing forces during birthing efforts, especially in the context of premature rupture of membranes) the greater the danger of vessel rupture and massive fetal haemorrhage. Prenatal diagnosis of vasa praevia followed by delivery by primary caesarean section prior to the onset of labour can increase survival from 44 to 97% [20]. Depending on gestational age RDS prophylaxis should be considered since increased rates of preterm delivery are probable [21, 23]. A current systematic review describes antenatal detection rates of vasa praevia of between 53 and 100% [24]. Transvaginal colour Doppler ultrasound is best for diagnosis. Detection rates may be much lower in the abdominal plane. According to a screening algorithm proposed by Rebarber et al. transvaginal ultrasound should be performed in every pregnant woman who presents with any of the following criteria [25]:

- Placenta praevia in the current pregnancy that is subsequently no longer demonstrable
- Vasa praevia in a previous pregnancy
- Velamentous cord insertion in the lower uterine segment
- Accessory placenta in the lower uterine segment
- Multiple pregnancy
In a large cohort of patients (n = 27,573 with 31 cases of vasa praevia) the authors showed that when diagnosis was made before 26 weeks gestation, vasa praevia was subsequently no longer demonstrable in 24% of cases, whereas if diagnosis was made in the third trimester it persisted.

**Fetal umbilical cord insertion**

Abnormalities of cord attachment can also occur on the fetal side. Abdominal wall defects at the UC insertion (gastroschisis and omphalocele) are the most important. The incidence of both entities is around 3:10000 although that of gastroschisis has increased in recent years for reasons as yet unknown [16]. Omphalocele must be differentiated from physiological umbilical hernia that occurs in the first trimester during intestinal rotation and normally regresses before the 12th week. The UC and its fetal insertion are anatomical landmarks, important criteria to be visualised to differentiate abdominal wall defects. Omphalocele occurs at the umbilicus. The defect is covered by peritoneum with the UC inserting into this hernial sac. The relatively high rate of associated genetic abnormalities is an important aspect requiring consideration. In gastroschisis the UC inserts normally into the abdominal wall, in most cases to the left of the wall defect. The viscera lie freely in the amniotic cavity. While the karyotype is usually normal, the rate of associated abnormalities is approximately 10% [26]. A detailed discussion is beyond the scope of this text [16].

**Umbilical cord cysts**

Cysts can occur anywhere along the entire length of the umbilical cord (Figs. 5 and 6); the incidence in the first trimester is between 0.4 and 3.4% [27, 28]. Histologically they are differentiated according to their origins into either true (epithelialized) cysts or pseudocysts (lacunar expansions/protuberances of Wharton’s jelly). True cysts can occur as remnants of the allantois or the omphaloenteric duct, or as vascular malformations. They may be associ-
ated with malformations (e.g., omphalocele, urachus fistula) and chromosomal abnormalities (especially trisomy 18). As most of the available literature on the subject is in the form of case reports and small case series, data on complication rates and clinical course are limited.

Cases of UC cysts should be followed up by detailed investigation for associated malformations. Many first trimester cysts disappear by the second trimester (ca. 80% [11]), a development associated with a good prognosis [29]. UC cysts in the second and third trimester have a poorer prognosis due to the above-mentioned complications [28].

Number of vessels/single umbilical artery

Early in its development the umbilical cord is formed from four vessels. At this stage there are two umbilical arteries (UA) and two umbilical veins (UV) of which the right UV typically atrophies by the 6th week of gestation. Thus the normal umbilical cord consists of two UAs and one UV. While a persistent four- vessel umbilical cord rarely occurs [30], the most common cord variant, with an incidence of approx. 0.5% in the second trimester [31], is a single UA (two vessels: one vein and only one artery; SUA = single umbilical artery). Visualisation of the umbilical arteries is done either in the free umbilical cord or preferably at the level of the fetal urinary bladder by demonstrating the cord’s perivesical course using colour Doppler ultrasound (Fig. 7).

Using colour Doppler and high definition ultrasound, SUA can be diagnosed at this level as early as the first trimester. A low pulse repetition frequency (PRF) and a high colour gain are recommended, as comparatively small vessels (compared to the opposite side) may otherwise be overlooked (Fig. 7c and d). The perivesical course of the UAs should be visualised all the way to the umbilicus as there is a danger of confusion with fetal femoral arteries in early gestation (Fig. 8c and d).

If single umbilical artery is suspected in the first trimester the diagnosis should be confirmed at a later stage (Fig. 8a and b). The existing (single) UA compensates and its lumen is usually bigger than that of a three-vessel cord, with a diameter ratio of UV/UA ≤ 2 (in three-vessel cord UV/UA the ratio is > 2) [32]. The main clinical relevance of SUA is the association with structural abnormalities of the fetal cardiovascular and genitourinary systems, as well as more rare gastrointestinal and CNS abnormalities [11]. Approximately one third of fetuses with SUA have structural abnormalities, and chromosomal anomalies are found in 10% [31], although according to current data isolated occurrence of SUA does not significantly increase the risk of chromosomal anomalies [31,33]. SUA is regarded as a marker for the development of fetal growth restriction and premature birth [34].

The demonstration of an SUA should always prompt a detailed anatomical ultrasound examination with particular focus on the cardiovascular and genitourinary systems. Increased pregnancy monitoring because of the higher risk of IUGR is currently controversial [11,31].

Structural changes of the umbilical vein

There are two structural UV variants which are of clinical importance, both involving its intraabdominal course.

Persistent right umbilical vein (PRUV)

By the 7th week of gestation one of the initially paired UVs (usually the right one) has typically atrophied. The left UV initially runs dorsally in the abdomen and joins the portal system. In abdominal transverse section (at a level slightly caudal of the bony ilium) the umbilicalportal vessels arch to the right appearing to “encircle” the gallbladder while passing the stomach tangentially (Fig. 9a).

Portal vessels branch off along its course, as does the ductus venousus approximately at the peak of the dorsal arch (Fig. 9b). Chaoui et al. recently published a detailed description of the hepatic veins as a CME article in German and English [35].
If the left UV atrophies, the right UV persists (PRUV). The incidence is reported to be around 0.5–3:1000 [11, 36]. In ideal imaging circumstances the ultrasound shows a mirror image course of the vessel arching left towards the stomach. The gallbladder lies medially and thus also within the vascular arch. Demonstration as in Fig. 10 is usually more difficult since the gallbladder is more caudal to the UV. Persistent right umbilical vein should be regarded as a normal variant of no clinical significance when it connects to the portal system intrahepatically (intrahepatic variant of PRUV giving rise to the ductus venosus). However, during embryonic development, extrahepatic communications between the UVs and the sinus venosus (later communicating with the right atrium) develop, which later obliterate while an anastomosis between one of the UVs and the sinusoids of the liver develops [36]. If the right UV persists it is more common for this anastomosis not to take place, with the extrahepatic connection to the right atrium remaining patent (without development of the ductus venosus). This extrahepatic connection causes an increased preload as the entire umbilical venous blood volume bypasses the liver to enter the right atrium directly. Significant right heart strain and subsequent hydrops fetalis can develop. Thus if a PRUV is diagnosed, further cardiovascular anomalies should be excluded. The clinical approach is dictated if concurrent malformations are present. Where PRUV is isolated, and an intrahepatic connection to the portal system exists, the prognosis is excellent [36].

Umbilical vein varix
Dilation of the UV in its extrahepatic course behind the fetal cord insertion is diagnosed with an incidence of 2.8:1000 and is termed umbilical vein varix [37]. Morphologically on ultrasound a circular vessel dilation ≥9 mm [38] or more than 50% over the diameter of the intrahepatic UV is seen [39] (Fig. 11). Due to its association with chromosomal anomalies (especially trisomy 21) and other abnormalities, if umbilical vein varix is diagnosed, further focused examination of the fetus should always
be done [37,40]. Increased rates of IUGR and intrauterine death have also been described. The optimal timing of delivery (recommendations start from 34 completed weeks of gestation) is currently controversial since cases of intrauterine death have been described despite intensified monitoring even in the presence of isolated umbilical vein varix [41,42].

**The Third Trimester**

**Umbilical cord loops and prolapse**

Umbilical cord loops (the UC is wrapped at least once around the fetus’s neck, more rarely around the body) are increasingly common as gestational age increases with an incidence of up to 33% at term [43]. Although in most cases UC loops have no negative effect on perinatal outcome, the possibility of higher rates of UC compression as a cause of fetal distress or even asphyxia and intrauterine death is currently discussed. Although some of the above-mentioned UC complications are existentially relevant, current data do not show increased risk in the presence of less than 3 loops [43]. Nevertheless the diagnosis often triggers significant uncertainty and anxiety in pregnant patients and this must be taken into account when counselling the patient.

A unique form of UC loop occurs in monoamniotic twin pregnancies (Fig. 12). Since both fetuses are freely mobile in their common amniotic cavity, UC loops, knots and a thoroughly convoluted/tangled cord is the rule [44]. There is currently no consensus on optimal antenatal monitoring or timing of delivery of monoamniotic twins, which are generally high risk (mortality 8–42% [44]). The finding of a convoluted UC in monoamniotic twin pregnancies does not appear to have a significant effect on overall risk [45].

The term umbilical cord prolapse describes when the UC, as the presenting part, prolapses into the cervix or vagina after rupture of membranes. This must be distinguished from cord presentation outside of labour. UC prolapse has a mortality of up to 50%
[11], although current data show a reduction in mortality to under 10% in the context of rising caesarean section rates [46]. The main risk factor for cord prolapse necessitating emergent delivery, is abnormal fetal lie. Numerous other factors are also associated with increased risk including prematurity and low birth weight, abnormalities of the maternal pelvis, polyhydramnios, UC length $>80$ cm and others (male sex, multiple pregnancy, placenta praevia and obstetric interventions) [46]. Risk factors such as multiple cord loops, very low placental cord insertion or polyhydramnios should be known to the obstetric team as this knowledge is essential for the correct interpretation of the progress of labour and particularly the CTG performed during labour, with labour management adjusted to take account of this information.

**True and false knots**

Umbilical cord compression can also be caused by tightening of a true UC knot. So-called false knots, in contrast, are varicose dilatations of umbilical vessels with an appearance sometimes imitating true knots. Their course is snaked but the UC does not form an actual knot that can tighten. UC knots are usually diagnosed postnatally. Antenatal diagnosis of true knots is possible, also causing major maternal anxiety. Well-defined diagnostic criteria and management standards are currently lacking and misdiagnosis is relatively common. Nevertheless, diagnosis of a UC knot should be followed by detailed Doppler ultrasound of the UC and stringent pregnancy monitoring [47, 48].

**Umbilical cord tumours**

Umbilical cord tumours are very rare entities. In most cases they are haemangiomas or teratomas. As in the case of UC haematomas (see below), tumours can cause umbilical vessel compression. Antenatally haemangiomas appear as echogenic structures with demonstrable perfusion on Doppler ultrasound. Multicystic variants make the differentiation from teratomas and haematomas particularly difficult [11]. Mortality is estimated to be 40%. Associated fetal malformations are described without any specific preponderance [49].

Teratomas of the UC are usually benign tumours. Their appearance on ultrasound is heterogeneous often with both cystic and solid components. Concurrent malformations are described in about half of published cases. Teratomas can reach considerable size and are a cause of cardiac failure in the fetus (“high-output heart failure”). Rupture of cord vessels is a particular danger associated with UC teratomas [50].

**Special Considerations**

**Doppler ultrasound**

Doppler ultrasound of the UC is relatively easy to carry out even in the later weeks of pregnancy thanks to the length and variable course of the UC. Due to the flexibility of the cord, vessels can usually be examined at the appropriate angle for good quality spectral curves. Nevertheless the cord’s length and winding course make standardisation and the exact definition of the examined segment and precise Doppler measurement at a specific angle difficult.

The umbilical arteries are the most commonly examined vessels in the assessment of fetal circulation. They provide information on the fetoplacental capillary bed; where resistance is increased, investigation of the UAs allows early detection of risk to the fetus. Assessment of UC blood flow is done particularly in the setting of fetal growth restriction to differentiate between compensated and decompensated placental insufficiency. Various (semi-) quantitative and qualitative measures can be assessed. The umbilical vein also provides information on the fetal status, allowing assessment of the fetal central venous vasculature, e.g. increased preload in the setting of right heart failure. The examination of other fetal and/or maternal vessels is often necessary for a complete assessment. We refer the reader to textbooks on the application of Doppler ultrasound in obstetrics for a complete description of Doppler ultrasound of the UC.

**Invasive procedures and umbilical cord haematoma**

Fetal blood sampling for the investigation of fetal anaemia, congenital infections and specialised genetic investigations is possible through UC puncture. Ultrasound-guided puncture of the UV allows both blood sampling and, via the same route, blood transfusion for the correction of fetal anaemia as indicated. The proce-
dural risk of abortion is 1% (fetus without malformations) [51]. A very rare complication is the development of a UC haematoma that, if large enough, can compress UC vessels and impair fetal circulation [52]. Spontaneous formation of a haematoma is also possible and the following risk factors have been reported: short UC, cord prolapse, velamentous cord insertion [53] and fetal clotting disorders [54]. UC haematomas are associated with a high rate of asphyxia and fetal mortality.

**Conclusion**

Detailed examination of the umbilical cord provides information on different aspects of fetal development. Targeted examination of this organ contributes to a better understanding of mechanisms underlying cord-related disorders. UC haematomas are associated with a high perinatal risk of adverse outcome. A very rare complication is the development of a UC haematoma during pregnancy. The diagnosis is usually made antenatally and is associated with a high risk of morbidity and mortality.

**Conflict of Interest**

None.

**References**


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