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Ultrasound Assessment of the Umbilical Cord

Junichi Hasegawa

ABSTRACT

As a nonreassuring fetal status and intrauterine fetal death are often caused by umbilical cord abnormalities, obtaining an ultrasound diagnosis of umbilical cord abnormalities is required for a safe delivery. We believe that the prenatal detection of umbilical cord abnormalities and appropriate management of the delivery improves perinatal morbidity and mortality rates. In the present review, the protocol for ultrasound diagnosis and management of umbilical cord abnormalities, including abnormalities of cord insertion, hypercoiled cord, nuchal cord and fore-lying umbilical cord, is discussed considering current knowledge regarding the physiological and pathological aspects of these umbilical cord abnormalities.

Keywords: Velamentous cord insertion, Hypercoiled cord, Nuchal cord, Umbilical cord, Ultrasound, Fetal heart rate.

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INTRODUCTION

A nonreassuring fetal status and intrauterine fetal death are often caused by umbilical cord and placental abnormalities. Therefore, conducting ultrasound assessments of the umbilical cord and placenta is important for a safe delivery. We believe that the prenatal detection of umbilical cord and placental abnormalities improves perinatal morbidity and mortality rates.

The causes of intrauterine fetal death after 12 weeks' gestation determined based on clinical data collected from our department are demonstrated in Table 1. In our hospital, two-third of intrauterine fetal deaths are caused by umbilical cord or placental abnormalities, and umbilical cord abnormalities account for 48% of causes of intrauterine fetal death. Among cases involving cord abnormalities, a hypercoiled cord is observed in more than half of patients. Other causes, including velamentous cord insertion, umbilical ring constriction, rupture of the

umbilical cord, a hypocoiled cord and umbilical cord prolapse are also observed.¹ The indications for emergency cesarean section due to a nonreassuring fetal status are also demonstrated in Table 2. Although 15% of emergency cesarean sections were performed based on unexplained abnormal fetal heart rate tracing findings, more than half of these procedures were carried out due to the indications of umbilical cord and placental abnormalities.¹

In the present review, we address umbilical cord abnormalities that should be detected based on an antenatal ultrasound diagnosis and require precise management during pregnancy and delivery, including abnormalities of cord insertion, a hypercoiled cord, multiple nuchal cords and fore-lying umbilical cord, considering current knowledge regarding the physiological and pathological aspects of these umbilical cord abnormalities.

Abnormal Cord Insertion of the Placenta

Abnormal cord insertion of the placenta includes cases of velamentous and marginal cord insertion and vasa previa. Velamentous cord insertion involves abnormal cord insertion in which the umbilical vessels diverge as they traverse between the amnion and chorion before reaching the placenta (Fig. 1). Because velamentous insertion forms as a result of abnormal placental development in early gestation and velamentous vessels of the umbilical cord are easily compressed during uterine contractions or fetal movement due to a lack of Wharton's jelly, this condition is strongly associated with fetal morbidities, such as fetal growth restriction, preterm labor, abnormal intrapartum fetal heart rate patterns, low Apgar scores at one and 5 minutes, neonatal death^{2, 3} and abruption of the placenta.⁴⁻⁷ On the other hand, as marginal cord insertion involves abnormal cord insertion in which a normal umbilical cord with Wharton's jelly inserts into the placental edge, the incidence of the above complications is lower in such cases compared with that observed in patients with velamentous insertion.

Vasa previa is a condition in which the lower velamentous vessels are present over the region of the internal os (Fig. 2). These fetal vessels are easily compressed and can rupture during uterine contractions and/or membrane rupture, thus resulting in fetal exsanguination. The infant survival rate in cases prenatally diagnosed by ultrasonography/color Doppler is 97%, compared



Senior Assistant Professor

Department of Obstetrics and Gynecology, Showa University School of Medicine, Tokyo, Japan

Corresponding Author: Junichi Hasegawa, Senior Assistant Professor, Department of Obstetrics and Gynecology, Showa University School of Medicine, 1-5-8 Hatanodai, Shinagawa-ku Tokyo 142-8666, Japan, Phone: +81-337848551, e-mail: hasejun@ oak.dti.ne.jp

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Table 1: Causes of intrauterine fetal death after 12 weeks' gestation	
Umbilical cord abnormalities	39 (48%)
Hypercoiled cord with umbilical ring constriction	21 (15)
Multiple cord entanglement	10
Velamentous cord insertion	3
Umbilical ring constriction	2
Rupture of umbilical cord	1
Hypocoiled cord	1
Umbilical cord prolapse	1
Placental abnormalities	12 (15%)
Placental abruption	9
Placental dysfunction	1
Twin to twin transfusion	2
Fetal anomalies	16 (20%)
Major morphological or chromosomal abnormalities	15
Amniotic band syndrome	1
Maternal factors	2 (2%)
Uterine infection	2
Unexplained	13 (16%)

A total of 82 cases (0.3%) of intrauterine fetal death were analyzed among 24,446 deliveries at Showa University School of Medicine, between 2001 and 2007

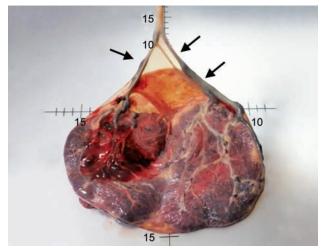


Fig. 1: A placenta with velamentous cord insertion: Velamentous cord insertion involves abnormal cord insertion in which the umbilical vessels diverge as they traverse between the amnion and chorion before reaching the placenta. Velamentous insertion forms during abnormal placental development starting in early gestation. The velamentous vessels of the umbilical cord easily compressed due to a lack of Wharton's jelly (arrows) during uterine contractions or fetal movement

to 44% among cases without a prenatal diagnosis.⁸ Obtaining an antenatal diagnosis and performing elective cesarean section prior to membrane rupture is therefore required to prevent fetal mortality from vasa previa. Even in cases of velamentous cord insertion in which the site of cord insertion is located on the lower uterine segment, without a diagnosis of vasa previa, strong associations with variable decelerations, a

Table 2: Indications for emergency cesarean section due to a nonreassuring fetal status
Placental abnormalities (35%)
Placental abruption, placenta previa
Umbilical cord abnormalities (21%)
Hypercoiled cord, velamentous cord insertion, cord prolapse
Oligohydramnios (9%)
Maternal complications (13%)
Pre-eclampsia, uterine infection
Fetal anomalies (7%)
Fetal growth restriction
Unexplained abnormal heart rate tracing (15%)

A total of 136 cases (3.3%) of a nonreassuring fetal status were analyzed among 4,163 deliveries at Showa University School of Medicine, between 2005 and 2008

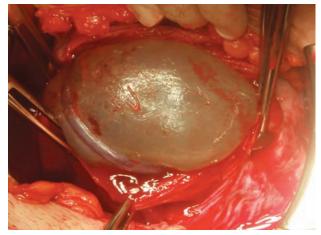


Fig. 2: A velamentous vessel on the membrane during cesarean section in a case of vasa previa



Fig. 3: Ultrasound picture of velamentous cord insertion in the mid gestation period: The placenta and umbilical cord insertion site are located on the anterior uterine wall. The umbilical vessels enter the placental margin parallel to the uterine wall and connect to the superficial placental vessels

non-reassuring fetal status, emergency cesarean section and other perinatal complications have been reported.⁷ Risk factors for vasa previa include an ultrasound diagnosis of low-lying placenta or placenta previa at an early stage of gestation,⁸⁻¹⁰ a bilobed or succenturiate placenta,⁸⁻¹⁰ multiple gestation,¹⁰ suspicion of aberrant vessels,^{10,11} cord insertion into the lower uterine segment ¹¹ and an *in vivo* fertilization pregnancy.⁸

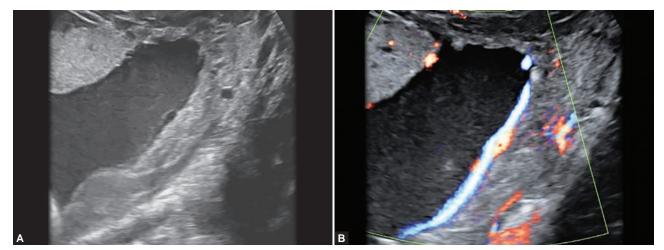
The rate of velamentous insertion ranges from 0.5 to 1.69% of singleton pregnancies; this rate is 10-fold higher in multiple pregnancies.^{2,3} Vasa previa was previously believed to be very rare, with an estimated occurrence of approximately 1 in every 2,000 to 5,000 pregnancies in older studies.¹² However, the rate of vasa previa (1:365) was found to be much higher in our consecutive series of patients assessed using ultrasound starting in the first trimester.¹³ Furthermore, the number of cases of vasa previa is increasing, as recent advances in ultrasonographic screening have enabled clinicians to detect potential cases of vasa previa more precisely. In the past, it was difficult to correctly diagnose vasa previa after delivery, and some patients treated with cesarean section due to indications of a low-lying placenta or who delivered infants with velamentous cord insertion diagnosed after delivery may not have been included in the population counts in previous studies.

Ultrasound Diagnosis of Abnormal Cord Insertion

The intrapartum clinical diagnosis of umbilical cord abnormalities is rare.¹⁴ Since, the ability to visualize the site of placental cord insertion becomes more difficult with advancing gestation, the site should be evaluated in the mid trimester period.¹⁵⁻¹⁷ The criteria for the ultrasound diagnosis of velamentous cord insertion are as follows: the umbilical vessels enter the placental margin parallel to the uterine wall and connect to the superficial placental vessels (Fig. 3); the cord insertion site and velamentous vessels are immobile, even when the uterus is shaken, and the umbilical vessels diverge as they traverse the membrane (Figs 4 and 5). The diagnosis of abnormal cord insertion should be made before delivery, as the relationship between the site of insertion and the location of the placenta within the uterus is consistent after the mid gestation period.¹⁸

With respect to the detection of vasa previa, previous studies have found routine screening of all pregnant females to not be feasible,¹⁹ although transvaginal scanning and color flow Doppler sonography of the cervical region should be employed in patients considered to be at increased risk.^{8,9,19} Importantly, our previous study suggested that ultrasonographic detection of the site of placental cord insertion, focusing on the potential for velamentous vessels and cord insertion in the lower uterine segment, enables physicians to effectively detect vasa previa¹¹ (Figs 6A and B).

Furthermore, we previously demonstrated^{20,21} umbilical cord insertion in the lower uterine segment in the first trimester to be associated with various placental abnormalities at delivery, including velamentous cord insertion, vasa previa, low-lying placenta, abruption of the placenta, umbilical cord prolapse and abnormal placental forms (Fig. 7). These placental abnormalities are similar to risk factors for vasa previa. Because the early placenta develops in association with advancing gestation in order to ensure an adequate blood supply from more richly vascularized areas in the direction of the uterine body, velamentous cord insertion is frequently observed in cases of umbilical cord insertion in the lower uterine segment during the first trimester. Interestingly, a total of 45% of cases of velamentous cord insertion develop in patients with low cord insertion.²¹ Therefore, systematically identifying the site of cord insertion at the time of early screening for fetal abnormalities is a very



Figs 4A and B: Grayscale ultrasound picture (A) and color Doppler image (B) of a long velamentous vessel in the mid gestation period: The placenta is located on the anterior wall, and the long velamentous vessel runs from the anterior wall to the posterior wall. The velamentous vessel is clearly depicted on color Doppler

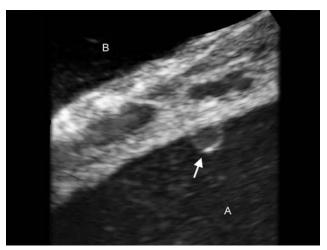


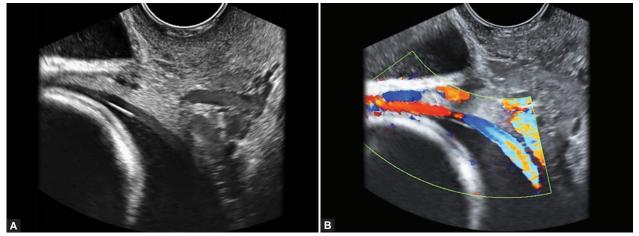
Fig. 5: Cross-sectional velamentous vessel in the anterior wall (Arrow: Velamentous vessel; B: Urinary bladder; A: Amniotic cavity)

simple and useful method for detecting vasa previa and velamentous cord insertion as well as various placental and umbilical cord abnormalities.

Hypercoiled Cord

A coiled umbilical cord with support provided by Wharton's jelly is thought to be more resistant to torsion, stretching and compression.²² However, several studies have shown that the presence of a hypercoiled cord correlates with poor perinatal outcomes, such as a low-birth weight, meconium staining of the amniotic fluid at birth and fetal growth restriction²³⁻²⁸ (Fig. 8). The postnatal umbilical coiling index (UCI) is calculated in order to evaluate the degree of umbilical coiling by dividing the total number of coils by the length of the cord in centimeters after delivery. The diagnosis of a hypercoiled cord after delivery is made in cases involving a postnatal UCI of \geq 0.3 coils/cm.²⁷

Narrow and weak cords near the fetal side are frequently observed in cases of intrauterine fetal death associated with a hypercoiled cord (Fig. 9). It is though that the umbilical cord is weakest on the fetal side and



Figs 6A and B: Grayscale transvaginal ultrasound picture (A) and color Doppler image (B) of vasa previa: A vessel of vasa previa runs along the uterine internal os. It is difficult to distinguish vasa previa from the amniotic membrane without using color Doppler, which can be used to easily make the diagnosis of vasa previa

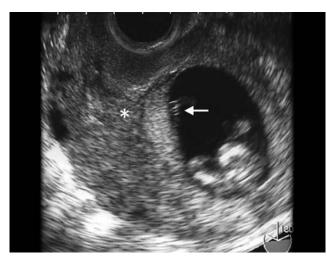


Fig. 7: Ultrasound picture of umbilical cord insertion in the lower uterine segment in the first trimester: Umbilical cord insertion (arrow) on the uterine os (*) in the first trimester is associated with various placental abnormalities at delivery

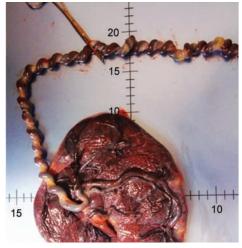


Fig. 8: A placenta with a hypercoiled cord



Fig. 9: A constricted umbilical ring in a case of a hypercoiled cord at mid gestation resulting in fetal death: No abnormalities were found in the stillborn baby, except for umbilical ring constriction

that conditions of a severe hypercoiled cord are associated with sudden fetal death. Alternatively, hypercoiled cords are frequently observed in fetuses exhibiting growth restriction due to the retention of an umbilical blood flow as a result of strong coiling.

Ultrasound Diagnosis of Hypercoiled Cord

The antenatal umbilical coiling index (aUCI) is ultrasonically calculated by measuring the distance between two adjacent coils of the umbilical artery from the right outer surface of the vascular wall to the next twist (antenatal UCI = 1/distance in centimeters), as proposed by Degani et al²² (Fig. 10). It is known that the antenatal UCI value is higher than the postnatal UCI value ($0.44 \pm 0.11 vs 0.28 \pm$ 0.08; p < 0.001).²² In addition, the 90th percentile, mean and 10th percentile antenatal UCI values in the second trimester have been reported to be 0.602, 0.403 and 0.204 respectively.²⁴ It has also been demonstrated that the antenatal UCI decreases as the pregnancy progresses to the third trimester due to elongation of the umbilical cord.²⁹ Therefore, a hypercoiled cord is usually diagnosed in cases in which the antenatal UCI is above 0.6 (Fig. 11).

However, only a few fetuses whose umbilical cord is diagnosed as hypercoiled antenatally are compromised during pregnancy or delivery. Although there are various reports of a hypercoiled cord being associated with the umbilical blood flow,³⁰⁻³² it remains unclear whether ultrasound screening is needed in such cases, as there is no predictable method of preventing a nonreassuring fetal status or intrauterine fetal death.

Skulstad et al³³⁻³⁵ recently showed that the blood velocity is higher in the umbilical vein at the site of the abdominal ring than in the cord. However, since measurements of the vessel diameter at the umbilical ring are too

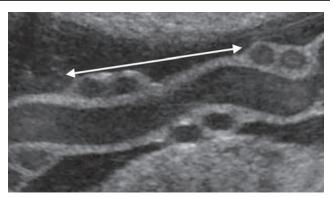


Fig. 10: Measurement of the antenatal coiling index of the umbilical cord: The antenatal umbilical coiling index is calculated by measuring the distance between two adjacent coils of the umbilical artery from the right outer surface of the vascular wall to the next twist (antenatal UCI = 1/distance in centimeters), as proposed by Degani, et al

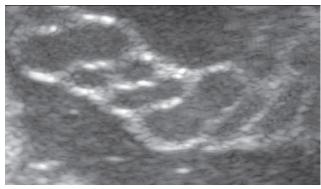


Fig. 11: Ultrasound picture of a hypercoiled cord

small to be valid, the detection of a high venous blood velocity is a superior marker of vascular constriction at the umbilical ring than direct diameter measurements. Furthermore, we previously demonstrated³⁶ that the venous velocity at the umbilical ring is significantly higher in fetuses with umbilical venous pulsation than in those without this feature. Therefore, there is a significant correlation between the venous velocity and the amplitude of pulsation. In fact, we experienced a case of intrauterine fetal death due to umbilical constriction in association with a hypercoiled cord in which the umbilical venous flow increased in correlation with the progression of deep umbilical venous pulsation before death.³⁶ However, to date, there is no way to rescue such fetuses, as the unfavorable conditions resulting from a hypercoiled cord often occur in the early second trimester.

Nuchal Cord

Nuchal cords are most frequently seen in cases involving umbilical cord abnormalities, with a prevalence of 15 to 24% at delivery.^{37,38} Although the presence of a single nuchal cord does not appear to affect clinical



management, patients with multiple nuchal cord entanglement are more likely to exhibit an abnormal fetal heart rate pattern during advanced labor, an umbilical artery pH of 7.10, require low or mid-forceps application and give birth to an infant with meconium and/or a low 1 minute Apgar score compared with those with single or no cord entanglement.³⁹

Ultrasound Diagnosis of Nuchal Cord

Nuchal cords are usually visualized ultrasonically as dimples with umbilical cords at the neck of the fetus on the sagittal view (Figs 12A and B). Such cases are identified by presence of the cord in the transverse and sagittal plane of the neck lying around at least three of the four sides of the neck. Although there is a linear increase in the incidence of single and multiple loops over the period of gestation,⁴⁰⁻⁴² nuchal cords continue to appear and disappear over time.⁴² However, the difficulty encountered in visualizing the nuchal cord at term and prior to the induction of labor may be due to fetal crowding, the low position of the fetal head and/or a reduced amniotic fluid volume.⁴³ Generally, the sensitivity of diagnosis is higher for color Doppler imaging, which may have a particular advantage in the presence of ruptured membranes.43

Prolapse and Fore-lying Umbilical Cord

Although the incidence of umbilical cord prolapse is extremely rare, ranging from 0.12 to 0.62%,⁴⁴⁻⁴⁷ cases of umbilical cord prolapse are strongly associated with poor neonatal outcomes, including intrauterine fetal death, neonatal death and cerebral palsy, as this condition may cause the cord to be compressed between the fetus and the maternal bony pelvis and/or soft-tissue, leading to fetal hypoxia.⁴⁸ Due to the increasing trend to deliver breech babies and cases of multiple pregnancies via cesarean section, the frequency of umbilical cord prolapse has decreased, with a reported incidence of 0.6% in 1932, 44 0.2% in the 1990s 49 and 0.12 in 2003. 44

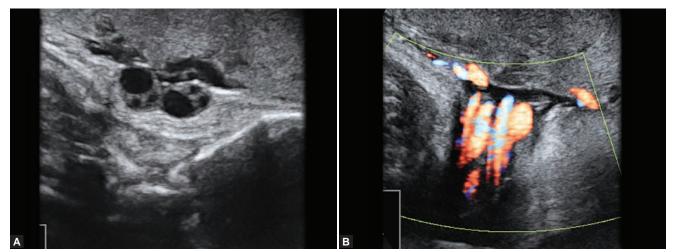
Fore-lying of the umbilical cord is diagnosed in cases in which the fetal membrane is intact and the umbilical cord precedes the presenting part within the intact membrane. In contrast, umbilical cord prolapse is diagnosed in cases in which the fetal membrane is ruptured and the cord protrudes in advance of the fetal presenting part through the cervical os and into or beyond the vagina. Umbilical cord prolapse quickly results in fetal compromise, with resultant long-term disability or death,^{47,50,51} either due to mechanical occlusion resulting from prolonged compression of the umbilical cord under the fetal presenting part or umbilical cord vasospasms triggered by the comparatively cooler temperature in the vagina.⁵²

Umbilical cord prolapse is associated with a high perinatal mortality rate, estimated to be approximately 32 to 47% before the 1950s.⁴⁹ The rate of perinatal mortality has also been reported to have decreased to 10% or less after the 2000s,^{47,49,53} presumably due to the more widespread availability of cesarean delivery.⁴⁸

Several risk factors associated with umbilical cord prolapse have been reported, including fetal anomalies, fetal malpresentation, multiple pregnancies, polyhydramnios, preterm delivery, a birth weight less than 2,500 gm, preterm premature rupture of the membranes, artificial rupture of the membranes, attempted rotation of the fetal head, amnioinfusion, external cephalic version, placement of an intrauterine pressure catheter or fetal scalp electrode and placement of cervical ripening balloon catheters.^{44,47, 48,53}

Ultrasound Diagnosis of Fore-lying Umbilical Cord

Umbilical cord prolapse is usually diagnosed based on an internal examination after rupture of the membranes,



Figs 12A and B: Grayscale ultrasound picture (A) and color Doppler image (B) of two nuchal cords on the sagittal view



Fig. 13: Transvaginal ultrasound picture of fore-lying umbilical cord: A fore-lying umbilical cord located between the internal os and fetal presenting part is depicted

whereas fore-lying of the umbilical cord is diagnosed via transvaginal ultrasonography (Fig. 13), which allows for the detection of the umbilical cord preceding the presenting part within an intact membrane. With respect to achieving a favorable neonatal outcome, it is important to conduct an ultrasound examination before and after amniotomy or spontaneous rupture of the membranes in order to detect an abnormal umbilical cord presentation.

Management of Delivery complicated by Umbilical Cord Abnormalities

Cord occlusion can induce both an increase in afterload and decrease in the fetal arterial oxygen content, both of which result in an activated vagal reflex, thus causing bradycardia.⁵⁴ Since, variable decelerations frequently occur in the second stage, even in cases without cord abnormalities, due to both cord and significant head compression, the frequency of variable decelerations is high in the first stage of labor in the presence of cord abnormalities, although the rate of variable decelerations does not differ from that observed in controls in the second stage of labor.^{55,56} Therefore, the application of continuous fetal heart rate tracing during labor is recommended in cases involving antenatally diagnosed umbilical cord abnormalities.

In particular, frequent fetal heart rate monitoring is recommended in the late third trimester in cases of velamentous cord insertion and vasa previa, as vulnerable velamentous vessels are likely to be compressed by the fetal presenting part, even during weak uterine contractions. Patients should therefore be educated regarding the signs and symptoms of preterm of labor. In cases of vasa previa, elective cesarean section is recommended at approximately 35 weeks of gestation in patients prenatally diagnosed with the condition.⁸

CONCLUSION

The antenatal ultrasound diagnosis of umbilical cord abnormalities is used to triage patients as either being high- or low-risk during the partum period and may alert the physician to an increased risk of a nonreassuring fetal status. In cases of antenatally diagnosed cord abnormalities, intensive monitoring with potential plans for cesarean delivery is indicated. We believe that neonatal outcomes can be improved with a precise antenatal ultrasound diagnosis and appropriate management.

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