

# Four True Umbilical Cord Knots

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## Abstract

**Background:** True umbilical cord knots are rare, but may be associated with intrapartum fetal distress and perinatal complications.

**Case:** This report describes a pregnancy complicated by intrauterine growth restriction and oligohydramnios. The labour was complicated by a non-reassuring fetal heart rate tracing. Four umbilical cord knots were discovered at the time of delivery. The baby's neonatal course was uncomplicated.

**Conclusion:** There is currently no reliable method of detecting umbilical cord knots using ultrasound or colour Doppler flow studies. Cord knots may be responsible for fetal compromise during labour and delivery.

## Résumé

**Historique :** Les nœuds vrais du cordon ombilical sont rares, mais peuvent entraîner la détresse fœtale intra-partum et des complications périnatales.

**Cas :** Ce rapport décrit une grossesse accompagnée de complications causées par un retard de croissance intra-utérin et un oligohydramnios. On a obtenu un tracé non rassurant de la fréquence cardiaque fœtale pendant le travail. Au moment de l'accouchement, on a constaté la présence de quatre nœuds dans le cordon ombilical. La période néonatale s'est passée sans complication.

**Conclusion :** Il n'existe présentement aucune méthode fiable de détection de nœuds dans le cordon ombilical, soit par échographie, soit par flux Doppler couleur. L'atteinte fœtale pendant le travail et l'accouchement peut être attribuable à la présence de nœuds dans le cordon ombilical.

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## INTRODUCTION

The clinical significance of true umbilical cord knots is unclear.<sup>1,2</sup> The incidence varies from 0.04% to 3% of all deliveries,<sup>3,4</sup> with an associated perinatal morbidity reaching as high as 11%.<sup>4</sup>

True knots should be distinguished from false knots, which are caused by the umbilical vein twisting around the artery, leading to localized thickening of Wharton's jelly. A true

knot, when untied, shows compression with a groove at the knot site, loss of Wharton's jelly, and persistence of structural change after being untied. There may also be edema, venous drainage, and vascular congestion distal to the knot.<sup>1</sup>

The following is a case report of a baby with intrauterine growth restriction (IUGR) and oligohydramnios who was born with four true umbilical cord knots.

## THE CASE

The patient, a 40-year-old, gravida 3, para 2 Sri Lankan woman, had undergone first trimester serum screening that suggested a possible trisomy 18 karyotype. Subsequently, amniocentesis done at 15 weeks and one day of gestation (15 + 1 weeks) showed a normal male karyotype (46, XY). An initial ultrasound at 15 + 1 weeks revealed a placenta previa. Follow-up ultrasound at 20 + 5 weeks showed an unchanged placenta previa and noted mild oligohydramnios, IUGR (estimated fetal weight [EFW] at the 2nd percentile), and echogenic bowel. Maternal screening tests for toxoplasmosis, hepatitis B, syphilis, herpes zoster, rubella, cytomegalovirus, herpes simplex, and thalassemia were negative.

An ultrasound examination at 23 weeks' gestation showed worsening oligohydramnios, with an amniotic fluid index (AFI) of 4 cm. The EFW remained at the 2nd percentile. Perinatology consultation suggested early onset oligohydramnios secondary to placental insufficiency or an undetected fetal syndrome; the patient was offered pregnancy termination, which she declined.

Ultrasound examinations and Doppler flow studies were performed weekly from this point onwards. At 27 + 2 weeks, the AFI had increased to 15 cm, but there was evidence of resistance in the umbilical artery demonstrated by a Doppler systolic to diastolic (S/D) flow ratio of 95%. The AFI improved from this point, but the Doppler S/D flow ratio remained high at 98% at 29 + 5 weeks and 98% at 31 + 6 weeks, with high resistance in the right umbilical artery and normal flow in the left umbilical and middle cerebral arteries. Weekly biophysical profiles and Doppler S/D flow

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**Prenatal ultrasounds and Doppler flow studies**

Gestational age	EFW (percentile)	Placenta	Amniotic fluid	Doppler S/D	BPP	Other
15 + 1 weeks	NA	Placenta previa	Normal, amniocentesis done	NA	NA	3.2 cm anterior wall fibroid
20 + 5 weeks	2nd	Placenta previa	Mild oligohydramnios, AFI = 7 cm	60%	NA	Echogenic bowel
23 weeks	2nd	Placenta previa	Moderate oligohydramnios AFI = 4 cm	NA	NA	Echogenic bowel
27 + 2 weeks	2nd	Marginal previa	Normal	95%, MCA Zone B	NA	
29 + 5 weeks	2nd	Marginal previa	Normal, AFI = 19 cm	98%	8/8	
30 + 5 weeks	NA	Marginal previa	Normal	35%	8/8	
31 + 6 weeks	2nd	Low lying	Normal	98%, High Doppler in right umbilical artery, normal on left. Normal MCA	8/8	
33 + 2 weeks	2nd	Low lying	Normal	Normal MCA	8/8	
33 + 6 weeks	NA	Marginal previa	Normal	80%, MCA Doppler normal	8/8	
34 + 2 weeks	NA	Low lying	Normal	90%	8/8	
35 + 2 weeks	2nd	Low lying	Normal, AFI = 10 cm	90%	NA	
35 + 6 weeks	NA	Low lying	Normal, AFI stable at 10 cm	85%	8/8	
36 + 2 weeks	NA	Low lying	Normal, AFI stable	90%, stable	8/8	
36 + 6 weeks	NA	Low lying	Mildly decreased	85%, MCA 70.8 cm/s	8/8	
37 + 2 weeks	2nd	High	Normal	NA	N/A	

EFW: estimated fetal weight; Doppler S/D: systolic to diastolic flow; BPP: biophysical profile; NA: not available; AFI: amniotic fluid index; MCA: middle cerebral artery.

ratios from 29 + 5 weeks onward were normal. The EFW remained at the 2nd percentile. By 37 + 2 weeks, the placental edge was 3.8 cm from the internal os. Induction of labour, originally scheduled for 38 weeks' gestation, was brought forward by one day following a non-reassuring non-stress test. The findings in prenatal ultrasound examinations and Doppler flow studies are shown in the Table.

The cervix was ripened mechanically with a Foley catheter placed intracervically on the day before delivery. An intravenous oxytocin infusion was begun on the following morning, and intravenous penicillin G (for group B streptococcus prophylaxis) was given at the same time. Three hours after starting the infusion, the patient spontaneously ruptured membranes, with cervical examination showing 2.5 cm of dilatation and 0% effacement, and the presenting part was 2 cm above the ischial spines.

Fetal cardiotocography subsequently showed deep variable decelerations of the fetal heart rate (FHR) with good recovery and minimal variability. With continuous external fetal monitoring, prolonged fetal bradycardia was noted at seven hours after beginning induction of labour. At this point the

cervix was found to be 7 cm dilated and 100% effaced, and the presenting part was at the level of the ischial spines. The cervix was fully dilated within 10 minutes, and delivery was expedited using low vacuum extraction. Four true knots in the umbilical cord were identified (see Figure). The placenta subsequently delivered spontaneously. The length of the umbilical cord was 127 cm.

The male newborn had Apgar scores of 8 and 9 at one and five minutes respectively, and had a birth weight of 2060 g (below the 5th percentile). He required free flow oxygen at birth for mild transient respiratory problems. Umbilical arterial cord pH was 7.32. The baby was admitted to the neonatal intensive care unit for one week because of low birth weight, but his course was uncomplicated. An evaluation for other causes of IUGR was negative.

## **DISCUSSION**

It is unclear when umbilical cord knots are formed during the antenatal period. In medical publications from as early as 1875, it has been postulated that knots form at between 9 and 12 weeks' gestation, when amniotic fluid volume is

**Four umbilical cord knots (photograph taken shortly after delivery).**



relatively large.<sup>1</sup> Subsequent theories propose that knots might actually be formed during labour, explaining the difficulty of prenatal ultrasonographic diagnosis.<sup>2</sup>

Various factors predisposing to cord knot formation have been described, including a long cord (> 80 cm), polyhydramnios, and a small fetus.<sup>1,5</sup> Hershkovitz<sup>2</sup> aimed to identify risk factors associated with true knots of the umbilical cord and found the following independently associated factors: genetic amniocentesis, a male fetus, chronic hypertension, grand multiparity, and other cord accidents such as cord encirclements and prolapse. Other studies have noted similar associations with cord knots, including a male fetus and multiparous mothers.<sup>3</sup> In the present case, we note that the baby was male and had a long umbilical cord (127 cm), and the mother was multiparous and had undergone genetic amniocentesis.

Associations can be found between true cord knots and intrapartum fetal distress. Hershkovitz<sup>2</sup> found higher rates of non-reassuring FHR tracings, meconium-stained amniotic fluid, Caesarean section, and antepartum death in babies with knotted cords. Rates of intrapartum and postpartum deaths were the same in both groups. However, other studies have noted a 4-fold to 10-fold increase in the risk of fetal death in association with cord knots.<sup>3</sup> The mechanism by which cord knots can affect the fetus is

unclear but could involve a shortening of the cord or alteration in cord positioning, causing a disruption in placental blood flow.<sup>1,5</sup>

Prenatal diagnosis of umbilical cord knots is extremely challenging and not always possible. Collins<sup>6</sup> reported the first prenatal diagnosis of a true knot in a pregnancy of 32 weeks' gestation, and described a characteristic "clover-leaf" pattern on ultrasound, but others have not been able to identify this.

If the theory that umbilical cord knots cause disturbances in uteroplacental blood flow is correct, then one should be able to detect abnormalities on prenatal Doppler flow studies.<sup>4</sup> Sherer<sup>5</sup> has described a case in which a true cord knot remained undetected prenatally, but in which an increased S/D flow velocity ratio and a systolic notch in the umbilical artery Doppler flow velocity waveforms were identified in retrospect. The author postulated that the knot could have caused a narrowing in the arterial lumen, leading to a rapid rise in resistance and reduction in flow with high S/D flow velocity ratio.

Some case studies report an association between cord knots and IUGR. For example, Ulm<sup>4</sup> described a pregnancy with severe symmetrical IUGR, oligohydramnios, and fetal distress during labour, in which the only potential cause identified was the presence of two cord knots found at delivery.

Although the knots remained undetected during the course of the pregnancy, as with the present case, the author noted that there were recurrent abnormal Doppler flow values in the umbilical arteries that were probably caused by the cord knots. In the present case, we also noted a high Doppler S/D ratio (95% and above) at 27 + 2 weeks and 31 + 6 weeks. It is possible that this was caused by a transient stenosing effect of the cord knot, since no other cause for flow abnormalities was found at delivery.

### **CONCLUSION**

Umbilical cord knots can be associated with a non-reassuring FHR tracing during labour and fetal compromise in utero. In the present case, it is possible that the cord knots were the underlying cause of IUGR and the non-reassuring intrapartum FHR tracing, since no other cause was apparent. However, prenatal diagnosis remains a challenge, as there is still no reliable method of detecting cord knots with ultrasound or colour Doppler flow studies. Cord knots may have a transient stenosing effect on the umbilical vessels in utero, causing disturbances in blood flow and higher resistances seen on waveform Doppler

studies. The use of waveform Doppler studies may help in screening for cord knots where these are suspected.

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