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**Case report**

# FATAL TRUE UMBILICAL CORD KNOT: A CASE REPORT.

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**ABSTRACT**

A true umbilical cord knot can be defined as an entwining of a segment of the umbilical cord. These knots are usually due to fetal slippage through a loop of the cord. A true knot is a rare event with a rate ranging from 0.1% to 2.1% of all umbilical cords at delivery. However, in 1.4% to 2.7% of such pregnancies, a true knot causes intrauterine fetal death (IUFD). Wharton's jelly and hydrostatic pressure of the umbilical vessels usually do not allow knots to completely tighten, but just before or during labor, a previously loose knot can tighten and cause asphyxia. Prenatal diagnosis of such an alteration is difficult and, even if diagnosed, there is no consensus on the therapy. We describe a case of a true umbilical cord knot that led to IUFD of a female fetus at the 40th week of gestation.



## 1. Introduction

A true umbilical cord knot can be defined as an entwining of an umbilical cord segment, which usually does not impede the fetal circulation. True umbilical cord knots are usually due to fetal slippage through a loop of the cord [1,2]. The rate of true knots ranges from 0.1% to 2.1% of all umbilical cords at delivery [3]. Identified risk factors of these knots are: a short umbilical cord, anemia, polyhydramnios, a small fetus, a male fetus, gestational diabetes mellitus, monoamniotic twins, amniocentesis, previous abortion and multiparity [4,5,6]. Intrauterine fetal death (IUFD) due to a true knot is a rare event (1.4%–2.7% of such pregnancies) because Wharton's jelly and hydrostatic pressure of the umbilical vessels do not allow knots to completely tighten [7]. However, a previously loose knot can tighten just before or during labor and cause asphyxia [3]. We report a case of a true umbilical cord knot that led to IUFD of a female fetus at the 40th week of gestation.

## 2. Case presentation

A 41-year old woman, gravida 4, para 2, at 40 weeks of gestation was admitted to the local hospital for initial labor. Throughout her pregnancy she suffered from anemia and bacterial vaginitis, and both conditions were successfully treated. No abnormalities were detected by ultrasonography with color Doppler, which was performed monthly during pregnancy, or by a cardiotocographic examination, which was performed during the last week. At admission, an obstetric clinical examination showed that the cephalic end of the fetus was facing down, with the fetal back to the right and the small parts to the left. The fetal heartbeat was barely perceptible, the uterine neck was shortened and impervious, and the amniochorionic membrane was intact. The cardiotocographic examination practiced at admission was reported during the only contraction, which reached the 40% of the "toco" scale, a "late" deceleration of mild entity. At the next obstetric clinical examination, the fetal heartbeat was regular (129 bpm), and was associated with the absence of uterine contractile activity.

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According to F.I.G.O. Guidelines[8], the following day, early in the morning, the woman was taken to the labor room to perform cardiotocography, which detected the absence of a fetal heartbeat. This was subsequently confirmed by ultrasound, which verified IUFD. Oxytocin was then administered with induction of spontaneous delivery of the fetus. At delivery, the presence of a bandolier cord around the fetal body was observed, associated with the presence of a true umbilical cord knot (Figure 1).

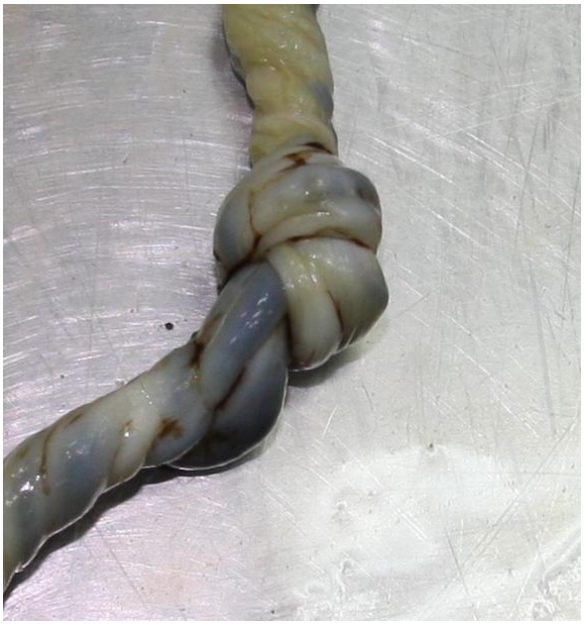

**Figure 1. Macroscopic aspect of a true umbilical knot.**

Delivery was followed by an autopsy, including fetal and placental gross examinations and histology. Histology of the placenta included analysis of the maternal and fetal plates, membranes, and umbilical cord (samples near placental insertion, near fetal insertion, and in the central area, with extensive analysis of the area of the knot) according to the Italian Group of Embryo Fetal and Placental Pathology and Archie et. Al. published protocol [8,9,10]. On gross examination, the fetal weight was 3100, heart weight was 23.1 g, lung weight was 45.2 g, liver weight was 123.0 g, spleen weight was 8.7 g, and brain weight was 302.0 g (< 9 percentile). The crown-heel length was 46.0 cm, crown-rump length was 34.3 cm, fronto-occipital circumference was 29.0 cm (=0 percentile), femur length was 7.0 cm, and foot length was 7.0 cm. Other fetal biometric parameters were normal for gestational age. A histological examination detected intense pulmonary and cerebral edema, and major poly-visceral congestion. The placenta showed a truncated form, was monochorionic-monoamniotic, and had a diameter of 19 cm × 11 cm, with maximum and minimum thicknesses of 6.3 and 2.5 cm, respectively. The placenta weighed 820 g. The umbilical cord was trivascular and 57-cm long, with a swollen central insertion. The samplings made in correspondence of the knot showed a reduction of Wharton's jelly, and hypertrophy of the muscular tunic with narrowing of the caliber of one of the umbilical arteries (Figures 2, 3). The lumen of the other umbilical artery was occupied by a thrombus, which occupied 90% of the lumen (Figure 4).

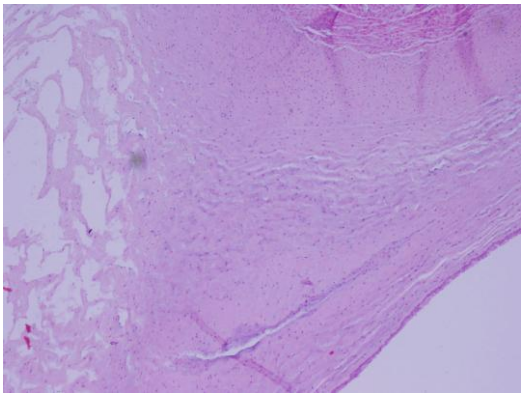

**Figure 2. Microscopic aspect of the cord wall and area of the knot, characterized by fibrous thickening of the chorion (hematoxylin and eosin stain, ×2.5).**

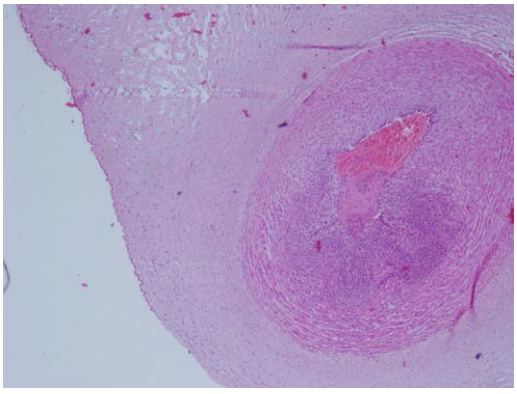

**Figure 3. Umbilical artery with thickening of the wall and hypertrophy of the muscular tunic (hematoxylin and eosin stain, ×2.5).**

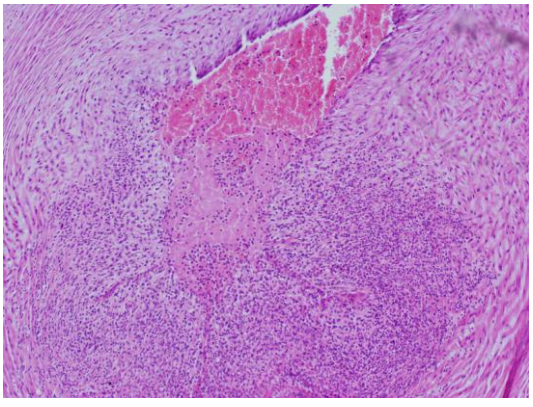

**Figure 4. A clear reduction of the lumen can be seen because of the presence of fibrin-granulocytic material indicating a thrombus (hematoxylin and eosin stain, ×10).**

## 3. Discussion

True knots are hypothesized to form between the 9th and 28th weeks of gestation, but knotting of the cord can also occur during labor [11,12,13]. A true knot is formed when the fetus, with its active movements, enters a loop formed by the cord. This is usually a simple knot, but sometimes three and four knots can occur [12,13]. A true knot leading to death of the fetus is rare because Wharton's jelly and hydrostatic pressure of the umbilical vessels prevent the knot from being completely tightened. When this occurs, the hemodynamic disorder is similar to that described for funicular compression in cases of cord rides, even if the consequences are more serious. This is because reactive movement of the fetus accentuates compression instead of decreasing it, further tightening the knot. Therefore, death of the fetus is inevitable and sudden [12]. An obstacle to blood circulation in the umbilical vessels, caused by compression of the cord at the knot, is serious because if Wharton's jelly is scarce then there is a lack of spiraling of the funicular arteries; or if there is an obstacle, such as a thrombus, as in the present case [3]. From a diagnostic-instrumental point of view, the appearance of repeated variable decelerations on cardiotocographic traces during labor is relatively typical with a circulatory obstacle at the level of the umbilical cord. These decelerations can be observed, even before the onset of labor, during sporadic spontaneous contractions that occur spontaneously, or when a test is performed with uterine contractions [14]. The obstetric scientific literature considers death of a fetus as inevitable and sudden in the case of a true knot of the cord that becomes tight due to reactive movements of the fetus [12,15]. The pregnancy presented in our report showed some of the antenatal risk factors such as male gender fetus, multiparity, previous abortion and maternal anemia. Autopsy and histological findings highlighted the morphological scenario of a particularly acute and rapid death, such as intense pulmonary and cerebral edema and major polivisceral congestion, but also revealed an important microcephaly ( brain weight < 9° percentile, head circumference = 0 percentile ) These findings confirmed critical role of maternal–fetal blood flow determining symmetric and regular growth of the fetus. An interesting finding was Wharton's jelly reduction, confirming its role in preventing obstacles to blood circulation [15-17]. Diagnosing or suspecting the condition of a true cord knot prenatally is difficult because of a limitation of the methods available. In particular, an ultrasonographic image of a true umbilical knot in the uterus has no characteristic signs. Therefore, in a routine ultrasonographic examination, a true umbilical knot is easily missed [15]. However, use of pulsed Doppler velocimetry of the umbilical vessels may display abnormal findings, which are correlated with cord compression in a constricted knot [16]. When there is suspicion of a true cord knot, complementary use of 3D-HD-flow improves diagnosis because it is more sensitive than power Doppler and allows the direction of flow to be visualized [17-20]. According to Eizenberg [21], antepartum fetal death due to umbilical cord abnormalities is still difficult to prevent because detecting distress in time for appropriate intervention is often impossible. However, prompt action to deliver the neonate after an abnormal cardiotocograph appears to be a necessary step to prevent such deaths. In such cases, forensic experts should have an understanding of this type of alteration of the umbilical cord that is associated with intrauterine growth retardation, fetal distress, fetal hypoxia, perinatal mortality, and long-term neurological damage. This is because this situation can be a source of claim for medical liability [22 - 24].

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