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REVIEW ARTICLE

Umbilical Cord Entanglement: Diagnostic and Clinical Repercussions

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ABSTRACT

The use of three-dimensional (3D) ultrasonography since the first trimester of pregnancy allows the diagnosis and follow-up of cord entanglement throughout the entire pregnancy and delivery.

This technology permits a more accurate diagnosis when compared with two-dimensional (2D) ultrasonography or Doppler.

The vast majority of cord entanglements observed at the end of the first-trimester will persist during the entire pregnancy; delivery outcome is usually not affected by this finding except for cases in which multiple cord entanglement is diagnosed.

Keywords: Doppler, 2D, 3D/4D umbilical cord, Entanglement control through all pregnancy.

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INTRODUCTION

The umbilical cord can develop a considerable number of abnormalities, accidents and pathological conditions that may have very different obstetric and fetal repercussions.[1-4] The most common event is nuchal cord, but cord entanglement can take place within the cord itself or around any part of the fetus. This event is seen in 16 to 30% of all deliveries[5,6] and a frequency of cord entanglement of up to 38% has been reported[7] (Fig. 1).

Up to 2% of newborn babies have a double loop nuchal chord[8-15] and in 0.5% of newborn babies the cord is wrapped around one of the extremities (Fig. 2).

Cord entanglements can be classified according to the following:[48]

- Their nature (tight or loose)
- Number of loops (simple, double, triple or multiple)
- Location (neck, body, extremities or mixed).

A loose nuchal cord is one that can be reduced during delivery by sliding the loop over the head or body of the baby. In contrast, a tight nuchal cord must be clamped and cut to allow delivery of the baby.

Cord entanglement results from active fetal mobility and is facilitated by abundant amniotic fluid (especially if there is polyhydramnios), a long umbilical cord or monoamniotic multifetal pregnancy. As a consequence there is shortening of the free segment of cord, the extent of which can be estimated by calculating that the segment around the neck in a case of nuchal cord measures about 32 cm, if it is around the leg it measures about 15 and 10 cm, if it is around an

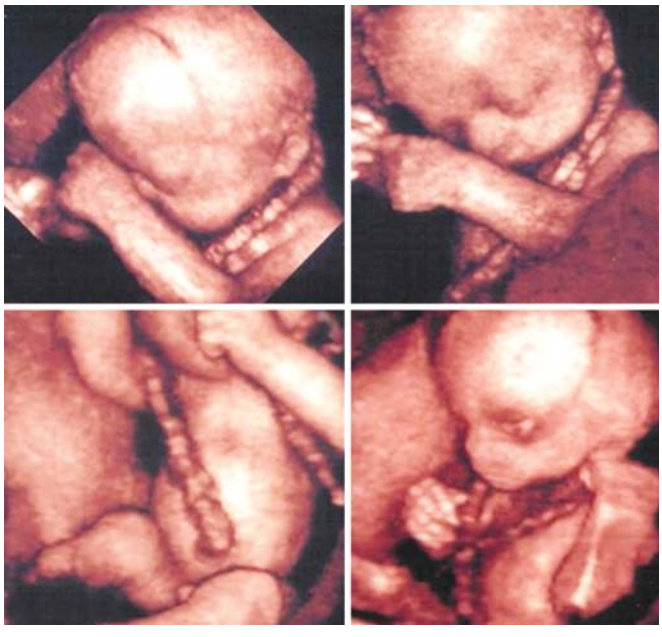

Fig. 1: Four cases of fetuses between weeks 18 and 22 with nuchal cord entanglement around the neck. The below right fetus is a hydrocephalus. The above right fetus shows two loops

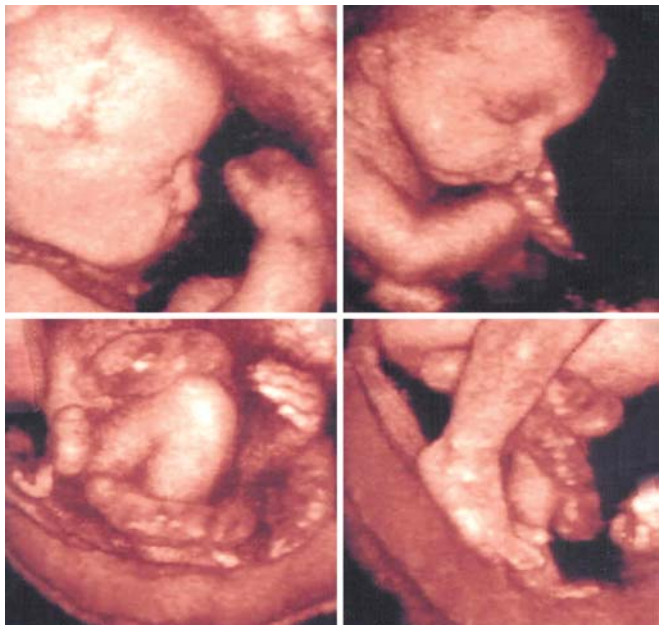

Fig. 2: Third trimester. Fetuses with loops around the neck (above left), hand (above right), foot (below left) and two loops around the leg (below right)

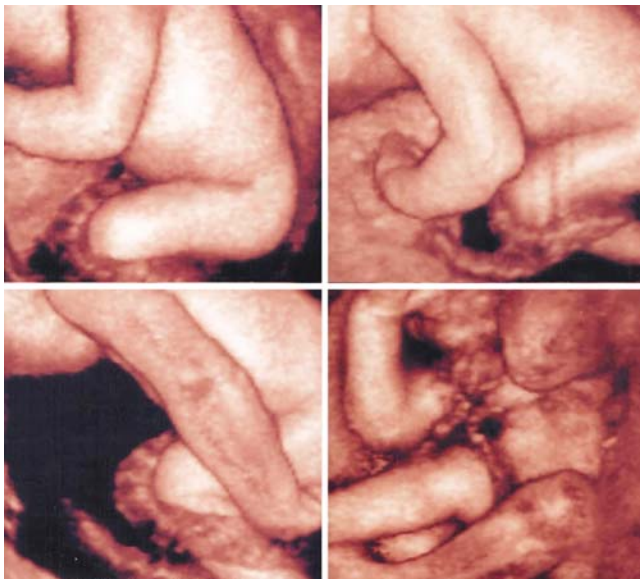

Fig. 3: Umbilical cord images showing loops around the thigh and legs. Almost 30% of the fetuses are born with nuchal entanglement, thus, it must be considered a physiological finding

arm.[16,17] These events can be clearly observed with three-dimensional (3D) ultrasonography[16,18,19] (Fig. 3).

There is still controversy about the clinical significance of cord entanglement, but given the frequency with which it is seen, it must be considered as a physiologic event. Cord entanglement is not usually a problem during pregnancy or during labor and delivery. However, in rare case multiple, tight loops may interfere with blood flow that results in fetal distress or even fetal death.[20,21] Cord accidents, blood flow impairment problems, amputations, death by strangulation, premature placental separation, etc. have been described in relation to cord entanglement. Cord entanglement has also been associated with an increase in the number of pathologic monitor tracings, fetal acidosis, meconium-stained amniotic fluid, emergency cesarean sections, admission to neonatal intensive care units (NICU) and possibly with higher rates of perinatal mortality.[22,23] The presence of severe variable decelerations antepartum or intrapartum on the monitor tracing should lead obstetricians to consider a possibility of cord entanglement, but this would be a late diagnostic sign.

Diagnostic Means

2D Ultrasound

Loops of cord involved in entanglement can be seen with two-dimensional (2D) ultrasonography, but it is not easy diagnose with this mode. Toward the end of pregnancy it is difficult, if not impossible, to see the entire neck of the fetus. There are many reports of nuchal cord diagnosis with 2D.[22] Nevertheless, this event may be missed due to the poor sonic reflection of umbilical cord blood vessels. At times it is difficult to observe the entire trajectory of the cord. This problem may occur even with the most sophisticated machines.[27]

Color Doppler and Doppler Energy

These modes are exceptionally useful for detecting cord entanglement and should be the diagnostic 'gold standard'[24-26] (Fig. 4).

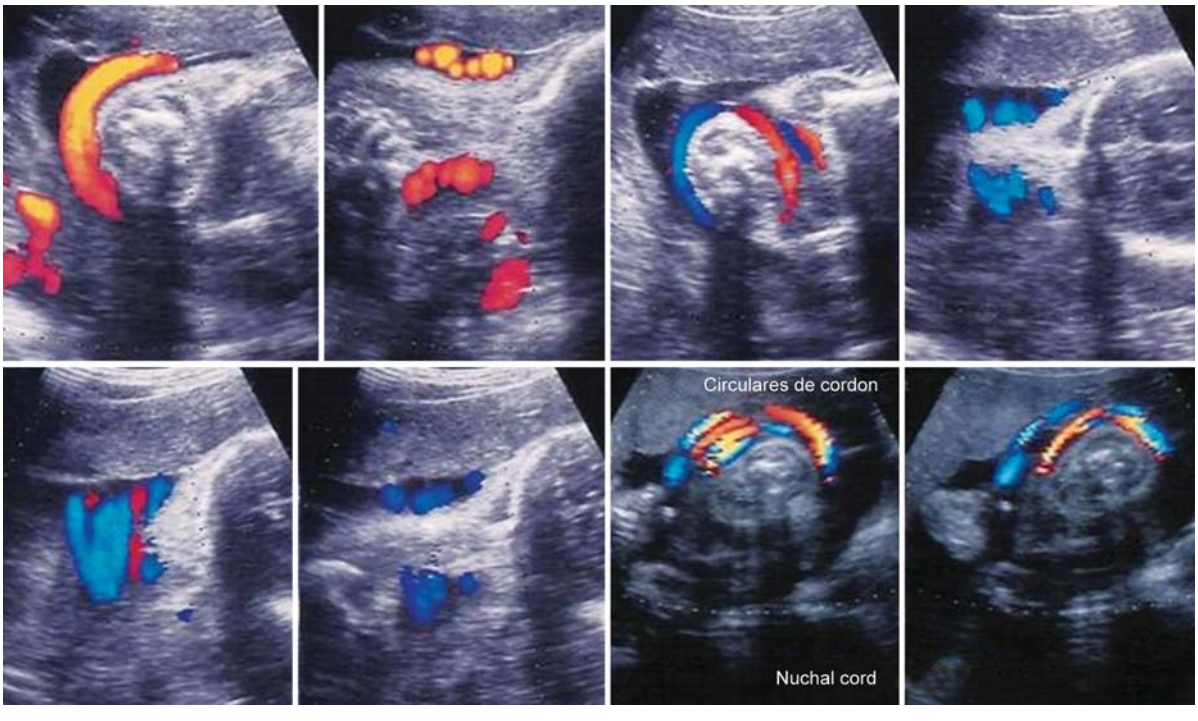

Fig. 4: Color Doppler showing a cord loop around the neck. Although this image is a typical finding with 2D, it is not always seen clearly. Sometimes it is difficult to be suspected if this technology (Doppler) is not used. The below right picture shows two loops around the neck

Examination with Doppler can at times be long and tedious due to the scant sonic reflection of blood vessels, and at times it is difficult to see the entire trajectory of the cord.[27]

There are many reports that confirm the diagnostic importance of these Doppler modes.[24,26,28] They facilitate detection and provide a sensitivity of 79%[29] that increases to 93% and to 97% at 32 and 36 gestational weeks[24,30,31] with a positive predictive value (PPV) of 89% , a negative predictive value (NPV) of 96%, and an efficiency of 93% during labor,[6,29] which is superior to the 33% efficiency obtained with 2D. Global sensitivity would be of 95% with a specificity of 92% for a PPV of 82% and a NPV of 98%.[32]

Tomographic Ultrasound Imaging

With this mode, multiple serial cuts that are separated by millimeters are carried out. When combined with Doppler, diagnosis of cord entanglement can be completed since loops can be observed millimeter by millimeter for the entire cord extension (Fig. 5).

Three-dimensional/Four-dimensional Ultrasound Imaging

Three-dimensional (3D) systems in real time and multiplanar mode as well as four-dimensional (4D) have facilitated diagnosis of cord entanglement as well as of other cord pathologies, such as knots, tumors and cysts.[1-4,30,33] A comparative study 2D/Doppler/3D of pregnancies at term[30] revealed the following diagnostic differences for each of these modes (Table 1).

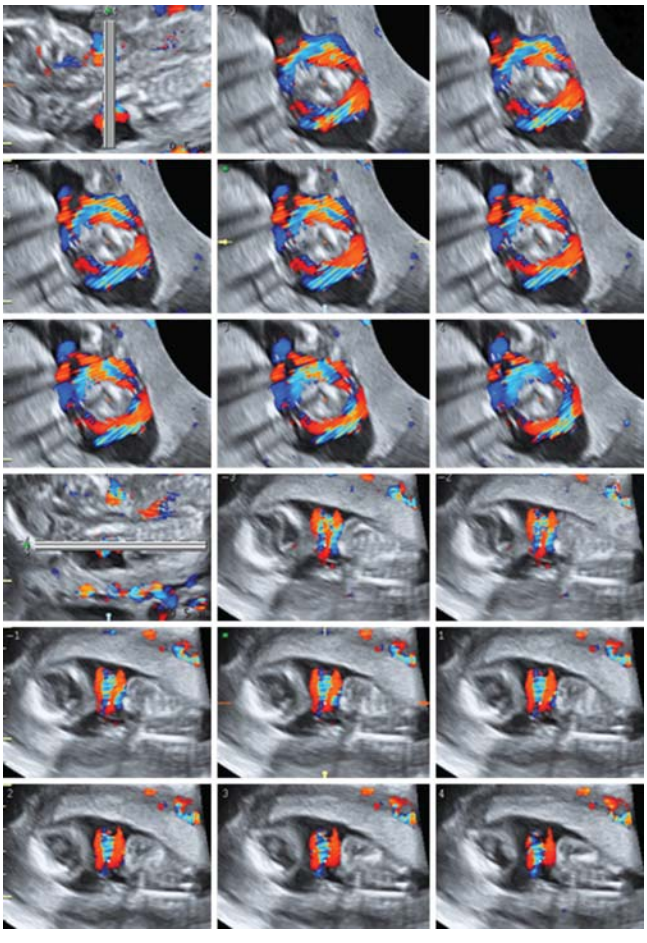

Fig. 5: Above TUI. Multiple tomographic sections of a single loop in sagittal view. Below in longitudinal view but showing a double loop

Table 1: Ultrasound technique (diagnostic differences)

Parameter	*2D*	*Doppler*	*3D*
Sensitivity	68.5	71.4	82.9
Specificity	80	82.4	77.7
PPV	58.5	62.5	60.4
NPV	86.1	87.5	91.7
False negative index	31.4	28.6	17.1
False positive index	20	17.7	22.4
Reliability	76.7	79.2	79.2

Sensitivity, specificity, PPV and NPV according to the diagnostic technique used (From Hanaoka U et al[30])

Fig. 6: Umbilical cord showing two loops around the leg (right) and then going up to the face (left)

The ability to see the loops was greater using 3D than 2D or Doppler, improving diagnostic security indexes.[1-4,7,17,21,26]

3D real-time allows the loops visualization from the gestational week when appeared facilitating the early diagnosis and follow-up (Fig. 6).

Gestational Age

Practically, all studies about cord entanglement have been carried out in the last weeks of gestation. However, it is advisable to try to diagnose this event at the end of the first-trimester during 'prenatal diagnosis.' This is an ideal moment to carry out the study because the whole fetus can be observed with 3D (Fig. 7). Visualization of the cord should be a part of the morphological study, as is done with nuchal translucency and nasal bone. In this way it is possible to follow the events that take place during the remainder of the pregnancy, since, as we will show further ahead, most cord entanglements persist up to the time of delivery.

Cord assessment at the end of the first trimester represents an important diagnostic step that can help avoid complications further ahead in the gestation, since multiple loop entanglements can have repercussions during pregnancy and during labor and delivery (Fig. 7).

Fig. 7: Above, 3D of a nuchal cord loop in week 14th. In the middle, the same loop in frontal and sagittal 3D view. The combination of such images allows to show how the cord ascend from the umbilicus, producing a loop and goes down to the abdomen making like a 'scarf' or a 'shoulder belt'. Below, loop located between the thorax and the mandible. The cord coiling is clearly seen

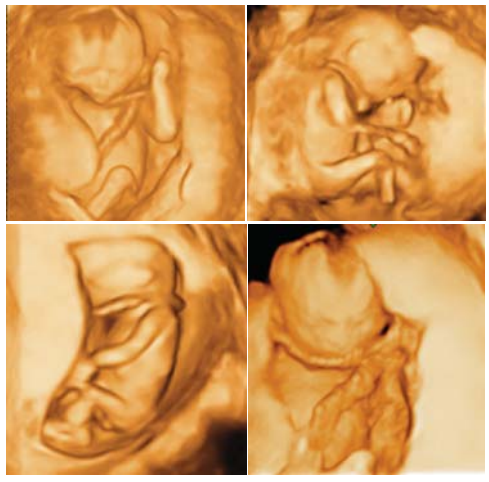

Fig. 8: Four 3D images of a cord ascending to surround the neck showed in different views

Early diagnosis will also allow obstetricians to distinguish between true entanglements from less significant events, such as loops around the abdomen, knots, cysts, etc. We will show further ahead that there are entanglements that are present in the first-trimester and then disappear, while others appear in the second-trimester, or more so, in the third-trimester. It is not difficult to search for them during the anatomic survey that is carried out between the 18 and 24 weeks of gestation or in the third-trimester.

Number of Loops

The new ultrasound techniques allow the determination of the number of loops involved in cord entanglement from the first-trimester on. The risk of an adverse outcome for the fetus increases as the number of loops increase (Figs 8 and 9).

We have observed the following incidence of loops in nuchal cords:

- Overall in the first-trimester, 25.3%
  - One loop in 18.7% of cases
  - Two loops in 6% of cases
  - Three loops in 0.7% of cases
- Persistent loops at 20 weeks gestation, 16.8%
  - One loop in 13.4%
  - Two loops in 3.4%
- Loops present at 32 weeks gestation, 18.2%
  - One loop in 15.2%
  - Two loops in 2.7%
- During delivery, 29.3% had nuchal cords
  - 22.7% with one loop
  - 5.3% with two loops
  - 1.3% with three loops

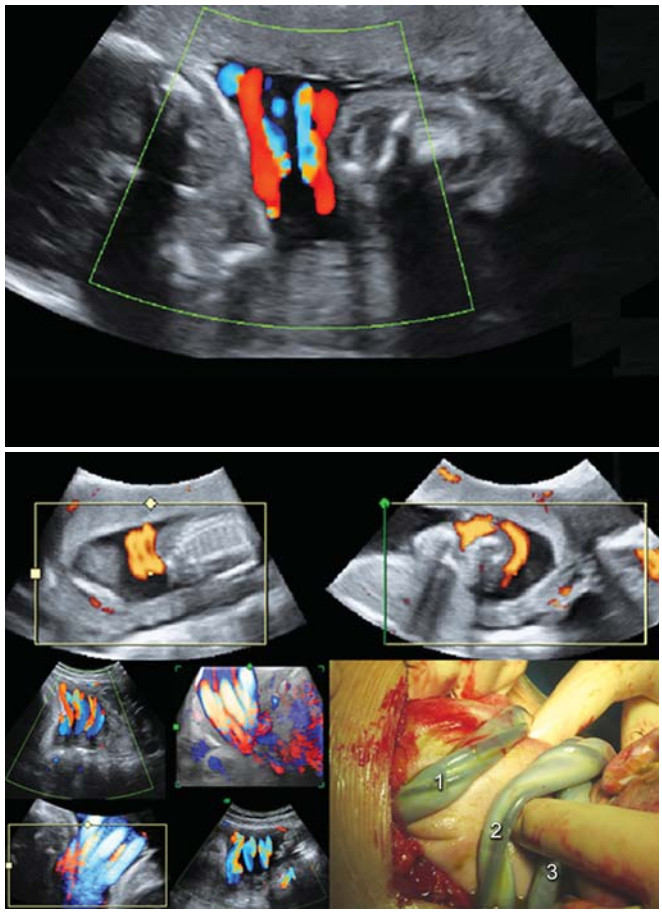

Fig. 9: Multiple cord loops

Most nuchal cords persisted throughout gestation, although the number of nuchal cords increase at the end of gestation.

Repercussions and Clinical Interest

Clinical interest about cord entanglement centers about the possible reduction in venous circulation that would result with tight loop compression. The result would be immediate

fetal hypoxia and central hypovolemia. Data from serial studies[22] have shown that cord compression for more than 10 minutes results in a reduction of cerebral vascular flow resistance along with a fall in partial pO_2 pressure.

These alterations only take place if there is nuchal cord compression. This is why these manifestations are more common during the active phase of the second stage of labor and why there are only a few newborn babies with nuchal cords which are severely affected at the time of birth. Since, identification of babies at risk can lead to appropriate and timely intervention, there is interest in early diagnosis of umbilical cord entanglement.[34-36]

Classical reviews have described a higher frequency of meconium-stained amniotic fluid (two to six times, between 10 and 20% of cases), a higher rate of inductions, slow labor progress, shoulder dystocia, low blood pH (20 to 30%), higher cesarean section rates, a greater need for immediate neonatal resuscitation and a higher rate of NICU admissions.[37-39] It seems that cord entanglements that restrict fetal movement are the ones that are at increased risk of adverse perinatal outcome.[2,37]

Management of nuchal cord cases with multiple loops is more debatable. In cases of multiple loops compared with a single loop[40] a greater tendency to cardiac frequency alterations during labor was observed, more instrumentation was required, there were more low Apgar cases and more cases of blood pH below 7.10; but curiously, the cesarean section rates were not significantly different nor were the Apgar scores worse.[41] The authors conclude that prospective decisions should not be made based on ultrasound multiple-loop nuchal cord diagnosis.[40,42]

It is of interest that there are reports of nuchal cord with transitorily increased nuchal translucency that resolves after the 14th week of gestation, and with normal karyotypes and ultrasound findings at 18 to 20 weeks gestation. These findings are important for the prevention of decisions on false positive nuchal translucency results.[8]

In our study of 150 cases observed from the first trimester until the end of gestation there were only to fetal deaths (1.3% of the sample), both of which had tight double loops, with fetal demise at 14 and 20 weeks gestation (2 of 9 cases, 22%). Evidence suggests that risk increases with the number of loops involved (Fig. 10). But these data, as we will show further ahead, do not justify cesarean section based on this finding.

In our series, 82.6% ended pregnancy with a vaginal delivery. Spontaneous vaginal delivery occurred in 63.3% of gestations and instrument-assisted vaginal delivery (forceps or vacuum extraction) in 19.3%. Of the 16% gestations that had a cesarean delivery, only 1.3% were sectioned because of a suspicion of loss of fetal well-being.

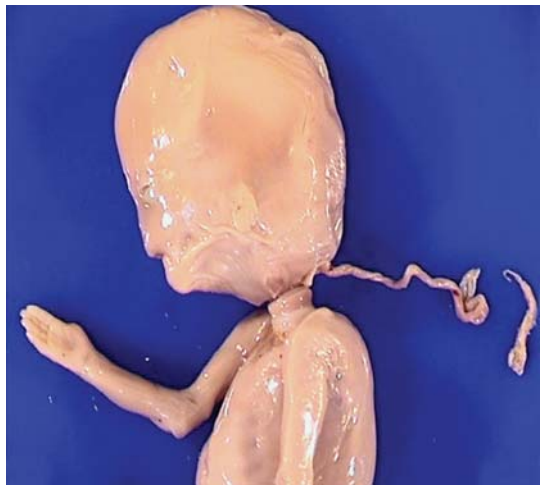

Fig. 10: Fetal obitus in week 14 due to two tight cord loops

The fetal monitor graphs were normal in 77.7% of cases. There were variable decelerations in 13.5%, early decelerations in 8.1% and only 0.7% had late decelerations. There were 8 cases with cord pH $\leq$ 7.20.

When we compared nuchal cords diagnosed at 12 to 14 weeks with those seen on week 20, week 32, and at delivery, we observed that there was a correlation, but there was no relation with Apgar scores or with blood pH. There was an inverse correlation between the number of loops seen at the 20th week of gestation and subsequently with the lowest pH values.

COMMENTS

Umbilical cord entanglement is a frequent finding in deliveries. Its prevalence at birth varies between 15 and 25% and is considered a physiologic event. It has been reported that the presence of two or more loops around the fetal neck occurs in between 2.5 and 8.3% of pregnancies.[9,10,41,42] Cord entanglement can be diagnosed with 2D ultrasonography and Doppler, although more recently and especially in the early stages of pregnancy 3D is recommended.

The presence of nuchal cord or other entanglement is not usually associated with alterations in fetal oxygenation. In very few cases is fetal health compromised and vaginal birth is contraindicated. Nevertheless, at times during labor a loop can become tight and affect blood flow and fetal oxygenation as the presenting part descends through the birth canal. This condition can be detected with proper monitoring of labor progress and with appropriate determination of blood pH.[6,42]

Although there seems to be a linear increase in the number of nuchal cords with gestational age, it is also true that these may appear and disappear with time.[11] A greater incidence of nuchal cord has been reported in white women and in male fetuses.[11,32,40]

There are multiple reports in the world literature that address the implications of cord entanglement during labor, delivery, and in newborn babies, but only a few, like this one, follow this event throughout the entire gestation. We have used for this study 3D/4D ultrasound assisted with Doppler and 3D angiography, which are aids that afford great diagnostic precision.

In nuchal cord cases we have not observed changes in flow velocimetry of the middle cerebral artery nor in the umbilical cord, since if there are interruptions of flow, they are only intermittent. If cord compression is not prolonged, flow does not cease in any significant way and fetal reserve can maintain adequate oxygenation.[32] Nevertheless, the numer of coils was reduced in nuchal cords and there was an inverse relationship between the number of coils and susceptibility to cord occlusion.[2] Maybe nuchal cords predispose to cord compression that can be associated with a higher rate of bradycardias, variable decelerations, umbilical cord acidemia, an increase in free oxygen radicals, and although rare, fetal demise.[12,32]

When there is cord occlusion, the most frequent change in fetal heart rate observed is variable deceleration.[11] For this reason a significant increase in alterations of intrapartum fetal heart rate patterns has been described.[11,40] A larger number of loops has also been associated with the most severe cases of growth restriction.[13,40]

When comparing newborn babies who had no nuchal cord with those born with one loop, two loops and multiple loops, we found no differences in birth weight, in heart rate alterations, in incidence of operative vaginal delivery or in Apgar scores. There was only a significant difference in the rates of cesarean birth, which resulted from evidences of nonreassuring fetal heart rate patterns or of cord compression during labor that were associated with low Apgar scores, but there was no correlation with the worst perinatal outcomes.[14,15,39] Retrospective studies also conclude that an association of nuchal cord with significant neonatal morbidity and mortality rarely occurs.

Studies about the incidence of nuchal cord in term and post-term pregnancies have established incidences of 33.7% at term (5.8% with multiple loops) and of 35.1% post-term (5.5% with multiple loops) with an increase rate of meconium staining and more severe cases of low pH ocurring in post-term babies with multiple loops. There were no differences with other variables. The authors concluded that the presence of nuchal cord should not influence the clinical management of labor and delivery and that primary neonatal adaptation was not affected.[42-45]

We conclude that since nuchal cords in the first-trimester do not imply in themselves poor perinatal outcomes, they should only be observed during gestation without a need for increased surveillance.[46,47] It is important to realize that if present in the first-trimester, there is an increased probability that they will persist throughout pregnancy, and that if they persist, especially if multiple loops are involved, they are correlated with deliveries that are managed more aggressively and with a higher rate of complications.

CONCLUSION

- The incidence of nuchal cord diagnosed in the first trimester with 3D/4D and Doppler ultrasonography is of about 25.3% (one loop in 18.7%, two loops in 6%, and three loops in 0.7% of cases).
- These percentages remain with small variations throughout pregnancy (they can increase or decrease), with 16.8% in week 20 and 18.2% in week 32. At birth, the number increases to 29.3%.
- Analysis of the type of delivery indicates that most babies (82.6%) are delivered vaginally. The cesarean section rate was 16%, but only 1.3% were sectioned because of a suspicion of fetal distress.
- The incidence of fetal demise was very low (1.3%), and fetal death was always early in gestation and associated with multiple loops.
- There is a correlation between the presence of nuchal cord in the first-trimester and its presence later on in pregnancy. However, there is no correlation with adverse pregnancy outcome. The presence of nuchal cord at weeks 20 and 32 of gestation only correlated with the worst cases of fetal acidosis with an increasing number of loops in the nuchal cord.
- The new ultrasound modes [Doppler, 3D/4D, tomographic ultrasound imaging (TUI)], are diagnostic instruments of great precision.

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