Nuchal cord

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INTRODUCTION — A loop of umbilical cord around the fetal neck (nuchal cord) is a common finding at delivery. In most cases, it is not associated with a significant increase in the rate of any clinically important adverse fetal/neonatal outcome. In case reports and small case series, tight nuchal cords have been associated with adverse outcomes, including fetal asphyxia and demise, but causality generally cannot be proven.

This topic will discuss issues related to prenatal diagnosis of nuchal cords, pregnancy and intrapartum management, and potential outcomes. Abnormalities of the umbilical cord are reviewed separately. (See "Umbilical cord abnormalities: Prenatal diagnosis and management".)

CLASSIFICATION — The term nuchal cord describes an umbilical cord that passes 360 degrees around the fetal neck. Nuchal cords can be classified as [1]:

- Single or multiple
- Loose or tight (ie, compressing the fetal neck)
- Type A or B (figure 1)
  - Type A – The placental end crosses over the umbilical end, entangling the neck in an unlocked pattern
  - Type B – The placental end crosses under the umbilical end, entangling the neck in a locked pattern

PATHOGENESIS — The occurrence of a nuchal cord(s) appears to be a random event, with increased risk among fetuses with excessive movement and/or a long umbilical cord [2,3]. In a retrospective study of singleton vaginal deliveries at term, an excessively long cord (≥70 cm in length) was far more common in pregnancies with a nuchal cord than in those with no nuchal cord (403/1451 [28 percent] versus 54/4733 [1 percent]) [4]. A case report described a fetus with an umbilical cord 150 cm in length and 10 loops around its neck [5].

INCIDENCE — The incidence of nuchal cords increases with increasing gestational age. At term, reported incidence ranges from 15 to 34 percent in large series [6-10].

Single nuchal cords are more common than multiple nuchal cords (11 to 28 percent versus 2 to 7 percent) [7,8,11-13]. In one study, the incidence of single, double, triple, and quadruple nuchal cords at delivery was reported to be 10.6, 2.5, 0.5 and 0.1 percent, respectively [13].

PRENATAL DIAGNOSIS — The prenatal diagnosis of a nuchal cord is based on an ultrasound examination documenting that at least 75 percent of the neck is encircled by umbilical cord. This is a pragmatic approach because, near term and depending on the lie of the fetus, imaging 100 percent of the neck is not always possible.

Although ultrasound cannot consistently and reliably distinguish between tight and loose nuchal cords [14,15], indentation of the fetal neck ("divot sign") suggests that the cord is tight [16,17]. Tightness can change during labor as the fetus descends through the birth canal.

Diagnostic performance of ultrasound — Both longitudinal and transverse views of the fetal neck should be obtained (image 1). The presence or absence of a nuchal cord can be determined by color Doppler (image 2), even in the first trimester (movie 1 and movie 2).
The sensitivity of ultrasound for diagnosis of a nuchal cord at term has generally been reported to be approximately 70 percent for gray-scale imaging [14,18], increasing to 83 to 97 percent with color Doppler [14,18-20].

In a study that compared the diagnostic performance of two-dimensional, color Doppler, and three-dimensional ultrasound for predicting a nuchal cord at birth in 120 singleton pregnancies, the overall performance of these techniques was [18]:

- Gray-scale (sensitivity 68.6 percent, specificity 80 percent, accuracy 76.7 percent),
- Color Doppler (sensitivity 82.9 percent, specificity 77.7 percent, accuracy 79.2 percent) and
- Three-dimensional ultrasound (sensitivity 71.4 percent, specificity 82.4 percent, accuracy 79.2 percent)

These differences were not significant, possibly due to the small number of pregnancies with a nuchal cord (n = 35) and the high proportion with multiple nuchal cords (5/35). Nuchal cords are easier to detect when there are multiple loops [15,19]. The authors’ subjective assessment of the ease of visualization of nuchal cord was best with three-dimensional sonography.

**Differential diagnosis** — Sonographic findings that may be misdiagnosed as a nuchal cord include a cord adjacent to but not encircling the fetal neck, posterior cystic neck mass, fetal skin folds, and amniotic fluid pockets [21]. Obtaining multiple real-time images from different angles and Doppler imaging can readily distinguish a nuchal cord encircling the fetal neck from these other entities.

**SCREENING** — We do not screen for the presence of a nuchal cord, given the lack of high-or even moderate-quality evidence that prenatal diagnosis of nuchal cords improves pregnancy outcome (see 'Possible sequelae during pregnancy' below). The American Institute of Ultrasound in Medicine does not consider attempts to visualize a nuchal cord a part of the standard prenatal ultrasound examination [22], and prenatally diagnosed nuchal cords are not routinely reported in ultrasound reports since they can be considered a normal finding.

The potential harms of screening are that it may cause maternal anxiety and lead to unnecessary sonographic follow-up appointments, antepartum fetal assessment, and intervention in the absence of evidence that the nuchal cord significantly increases the risk of an adverse fetal outcome [23].

For patients who ask about a nuchal cord during their ultrasound examination, we reassure them that visualization of nuchal cord is a common incidental finding, often resolves, has not been associated with a markedly increased risk of adverse pregnancy outcome, and does not warrant specific changes in prenatal or intrapartum care based on this finding alone. (See 'Pregnancy management' below.)

**NATURAL HISTORY** — A nuchal cord may persist or resolve, and those that resolve may reform [3,24]. Although formation and resolution appear to be random events, persistence may be more likely at term and with multiple nuchal cords. The type of nuchal cord impacts the course; a type A nuchal cord can become undone with fetal movement, whereas a type B nuchal cord cannot undo itself and can form a true knot when it passes caudally over the fetal body (figure 1).

**POSSIBLE SEQUELAE DURING PREGNANCY** — The body of evidence from observational studies suggests that nuchal cords are not associated with a significant increase in the rate of any clinically important adverse fetal/neonatal event. This evidence is of low quality due to factors such as publication bias, lack of comparison with an adequate control group, limitations of data derived from chart review and discharge coding, and small numbers of subjects and events.

In the largest available data set, in which a tight nuchal cord (defined as inability to manually reduce the loop over the head) was documented in 6.6 percent of 219,337 live births, there was no statistical association with adverse neonatal outcome [25]. However, a small increase in one or more adverse outcomes could not be excluded conclusively. In case reports and some small case series, nuchal cords have been associated with fetal demise, impaired fetal growth, meconium-stained amniotic fluid, perinatal arterial ischemic stroke and an increased frequency of intrapartum fetal heart rate abnormalities, operative delivery, low five-minute Apgar scores, and umbilical artery acidemia. Long-term, an increased risk of neurodevelopmental abnormalities has been reported.

**Fetal demise** — Data from large retrospective studies have not demonstrated an increased risk of stillbirth in fetuses with nuchal cords compared with those without nuchal cords [9,25-27]. However, case reports of stillbirths with one or more nuchal cords, indentation marks in the tissue around the fetal neck, otherwise normal prenatal and postnatal evaluations, and no other explanation for the demise suggest that nuchal cords can rarely be a cause of fetal death [27].

The presence of a nuchal cord alone is insufficient evidence of demise due to strangulation, but causality is supported by the presence of the following findings:
● Petechial hemorrhages of the head and neck, which are characteristic findings in strangulation [28, 29].

● Vascular congestion and thrombosis of umbilical cord vessels [28, 29].

● Cord edema or hemorrhage into the Wharton’s jelly [28, 29].

● Vascular ectasia, vascular thrombosis, and/or thrombotic vasculopathy (avascular villi, villous stromal karyorrhexis) involving the umbilical cord vessels and/or chorionic plate and stem villous vessels [30, 31].

● Absence of other conditions associated with fetal demise.

One potential mechanism for fetal asphyxia is restriction of carotid artery blood flow from tight entanglement around the neck; however, severe venous congestion may be sufficient to cause asphyxia and demise. Another potential mechanism is compression of the umbilical cord vessels themselves when the cord becomes tightly compressed against itself or the fetal neck. Multiple mechanisms may be involved.

Of note, a tight nuchal cord that impeded fetal oxygenation before delivery may not be tight at delivery because loss of fetal tone with increasing cerebral hypoxia may lead to loosening of the nuchal cord if multiple loops are not present.

**Impaired intrauterine growth** — Whether nuchal cords adversely affect fetal growth is unclear. Studies that have not discriminated between single and multiple nuchal cords have not reported a clinically significant impact of nuchal cords on birth weight [6, 32].

Several studies have observed a significant negative correlation between the number of nuchal cord entanglements and birth weight; multiple nuchal cords reduced birth weight by 93 to 180 grams [7, 11, 33]. However, others have reported discordant findings [34, 35].

**Abnormalities in tests for fetal evaluation**

**First-trimester combined test for Down syndrome screening** — A nuchal cord may be detected incidentally during measurement of nuchal translucency and may alter the nuchal translucency (NT) measurement if it is tight, or it may not be detected and incorrectly included in the measurement [36, 37]. In one prospective study including 53 fetuses with a nuchal cord during NT measurement (incidence 4.65 percent), the largest NT measurement overestimated NT in 50 percent, underestimated NT in 48 percent, and estimated NT correctly in 2 percent of cases compared with the true NT measurement after resolution of the nuchal cord [36].

If a nuchal cord is indenting the fetal neck, we suggest obtaining a NT measurement after resolution, which typically occurs within a short period of time. The median time to resolution was 2.21 hours in the study described above [36]. If the cord is loose, NT can be measured accurately as long as correct anatomic landmarks are used. If the nuchal cord does not resolve, measurements of NT above and below the cord will be different; the average of the two measurements is used to calculate risk [38].

**Nonstress test, biophysical profile, Doppler indices** — Information on the effect of nuchal cords on the nonstress test is sparse. Theoretically, vigorous fetal movement can tighten the nuchal cord, resulting in compression of carotid and umbilical blood vessels. The potential fetal heart rate consequences are variable decelerations and possibly late decelerations [2, 39-41]. However, at least one small retrospective study of variable decelerations during reactive nonstress tests found no correlation with the frequency of nuchal cords or other cord entanglements [42] (see "Intrapartum fetal heart rate assessment", section on ‘Physiologic significance of selected FHR characteristics’).

Prospective studies have reported similar Doppler indices in the cerebral arteries of fetuses with and without nuchal cords [19, 43, 44]. Other prospective studies of term and postterm pregnancies have not reported a reduction in prelabor amniotic fluid levels in pregnancies with nuchal cords [15, 45, 46].

These findings in large series do not exclude the possibility that an individual fetus with a tight nuchal cord may become compromised. As an example, a case report described absent fetal movement on a routine ultrasound examination of a 30-week appropriate for gestational age fetus [17]. Doppler velocimetry revealed absent end-diastolic Doppler flow and cardiotocography showed repetitive severe variable decelerations. Emergency cesarean delivery was performed with delivery of a 1200 gram, acidemic (pH 7.10) newborn with a tight double nuchal cord. Laboratory evaluation and placental histopathology were otherwise normal so the metabolic acidosis was attributed to the nuchal cords.

**Intrapartum fetal heart rate** — Prospective studies of newborn outcomes after antenatal identification of nuchal cords have generally not described increased rates of nonreassuring fetal heart rate patterns or operative delivery [15, 45-47]. However, each of these studies included fewer than 400 subjects. In contrast, two large retrospective studies including
approximately 38,000 fetuses with a nuchal cord at birth reported these pregnancies had a 60 to 80 percent increase in the frequency of abnormal fetal heart rate tracings compared with pregnancies with no nuchal cord [6, 32], although the cesarean delivery rate was not increased [6]. Both studies observed a higher rate of labor induction in pregnancies with a nuchal cord, which may account, at least in part, for the increased frequency of fetal heart rate abnormalities. Multiple nuchal cords appear to increase the risk of an abnormal fetal heart rate tracing [8, 27, 48].

A study that used near-infrared spectroscopy to assess the intrapartum effect of nuchal cords on cerebral hemodynamics and oxygenation found that nuchal cords were associated with an increase in cerebral blood volume during contractions, without a significant effect on cerebral oxygenation or neonatal outcome [47]. An increase in variable decelerations was also noted. The authors hypothesized that these findings were due to transient compression of the jugular veins by the cord during contractions.

**PREGNANCY MANAGEMENT**

**Antepartum** — An incidental finding of a nuchal cord on ultrasound examination does not warrant a change in prenatal care, given the lack of evidence of a clinically significant increase in adverse pregnancy outcome. (See 'Possible sequelae during pregnancy' above.)

No randomized trials or prospective studies have compared potential approaches to management of pregnancies with nuchal cords diagnosed prenatally. A retrospective study compared the outcomes of 188 pregnancies with nuchal cords detected by sonography during the second and third trimesters with 115 pregnancies without prenatally detected nuchal cords [49]. At delivery, 37 percent of pregnancies with prenatally diagnosed nuchal cords had nuchal cords versus 15 percent of the control group. Both groups had similar perinatal outcomes and few adverse events. This study of nuchal cords diagnosed remote from delivery, although small and retrospective, affirms previous data of generally good outcomes of nuchal cords diagnosed closer to the time of delivery.

**Breech presenting fetus** — We do not perform cephalic external version and advise against an attempt at vaginal breech delivery if the fetus has a nuchal cord because we believe there may be an increased risk of complications during these procedures. However, due to the possibility that the nuchal cord will resolve before labor, we offer a follow up ultrasound examination a few days later and proceed with version if the cord has disentangled [50].

There are sparse data on the management of breech presentations with a nuchal cord at the time of planned cephalic external version or planned vaginal delivery [51, 52], and no consensus on management of these patients. One study reported a decreased rate of successful external cephalic version in patients with nuchal cords (17.6 versus 53.6 percent) and a significant increase in fetal heart rate decelerations leading to interruption or abandonment of the procedure (82.3 versus 24.3 percent) [51].

**During labor** — The presence of a nuchal cord does not affect our intrapartum management. As discussed above, there is no strong evidence of a clinically significant increase in adverse pregnancy outcome (see 'Possible sequelae during pregnancy' above). In an observational study of pregnant women with fetuses with nuchal cords, antepartum Doppler ultrasound of the nuchal cord had poor sensitivity and specificity for predicting intrapartum fetal distress necessitating cesarean delivery [53].

As the fetal head descends or rotates, changes in the tightness of a nuchal cord may occur and may result in fetal heart rate decelerations, which should be managed as in any labor with fetal heart rate decelerations. (See "Management of intrapartum category I, II, and III fetal heart rate tracings").

**Delivery** — If a loose nuchal cord(s) is palpated after expulsion of the fetal head, it can usually be slipped over the head to free the fetus from the tether. If the cord(s) is too tight to easily slip over the head, it may be possible to slip it back over the shoulders and deliver the body through the loop.

These approaches may be unsuccessful if the cord is too tight. In these cases, it is important to avoid avulsing or tearing the cord while attempting to effect delivery. Occasionally, the body can be delivered without releasing the cord and without compromising the fetus. In most cases, we place a palm on the fetal occiput and push the face into the mother's thigh (or pubic bone), which allows the shoulders, then body, then legs to deliver (called "somersault maneuver") (figure 2) [54, 55]. The cord can then be unwrapped from the neck. In rare cases when this maneuver is not successful, the cord is doubly clamped and transected. However, early clamping and cord cutting decreases the volume of blood transferred from the placenta to the neonate, which may increase neonatal anemia, and may adversely affect the fetal to neonatal transition. (See "Management of normal labor and delivery", section on 'Cord clamping'.)

In monoamniotic twin pregnancies, clamping and cutting a tight nuchal cord on the first twin should be avoided since it may be the umbilical cord of the undelivered twin.
NEONATAL OUTCOMES

Neonatal intensive care unit admission — Short-term outcomes of neonates with a history of a tight nuchal cord are favorable. In a large retrospective study, term neonates with a tight nuchal cord were slightly more likely to be admitted to a neonatal intensive care unit but not more likely to have dopamine administered, have blood hemoglobin measured on the first day, receive a transfusion, or die [25]. Similarly, there were no differences in outcomes between the subset of very low birth weight neonates with a tight nuchal cord compared with those with no nuchal cord.

Long-term neurodevelopmental abnormalities — There is no strong evidence that a nuchal cord increases the risk of neurodevelopmental impairment, but few long-term outcome data are available. One study compared neurodevelopmental performance at one year of age for 66 infants with nuchal cords at birth and 124 infants without a nuchal cord [58]. Although the development scores of both groups were in the normal range and averaged well above the standard means, the nuchal cord group had slightly, but statistically, lower scores, which was attributed to the cases with multiple or tight nuchal cords with intrapartum signs of fetal compromise.

It is not clear whether a nuchal cord increases the risk for developing cerebral palsy [57-61]. The collaborative study of cerebral palsy, a prospective multicenter study including over 12,000 infants, found no increased risk of clinical neurologic impairment at one year of age among the over 4200 infants with nuchal cords at birth [58]. In contrast, a population-based case-control study of 271 singletons with spastic cerebral palsy and 217 controls without developmental disorders reported a 2.8-fold increased risk of spastic cerebral palsy in newborns with a nuchal cord (odds ratio [OR] 2.8, 95% CI 1.31-6.02) [60]. (See “Cerebral palsy: Epidemiology, etiology, and prevention”.)

Some authors have suggested that tightness of the nuchal cord is the key factor increasing the risk of adverse outcome. A retrospective study reported that a tight nuchal cord at delivery increased the odds of developing unexplained spastic quadriplegia (OR 18, 95% CI 6.2-48) [57]. Other presentations of nuchal cords, such as any or multiple entanglements, were not analyzed. However, tightness of the nuchal cord was not significantly associated with cerebral palsy in another retrospective study [60]. A retrospective study analyzing factors preceding neonatal hypoxic-ischemic encephalopathy (HIE) found that a tight nuchal cord was an independent risk factor for HIE (OR 2.8, 95% CI 1.02-7.8) [61].

The possible association of a tight nuchal cord with cerebral palsy in retrospective studies may be due to recording bias: Documentation of a tight nuchal cord may be more likely when the infant is born in a poor condition. In a study comparing elective versus systematic recording of nuchal cords in healthy infants with low Apgar scores <7 at one minute, documentation of a tight nuchal cord was six times more likely with elective reporting than when recording was systematic [59]. Likewise, when the presence or absence of nuchal cords was recorded systematically, a significant association between nuchal cords or tight nuchal cords and cerebral palsy was not observed.

SUMMARY AND RECOMMENDATIONS

- Nuchal cords can occur as single or multiple entanglements around the fetal neck. They may be loose or tight. They have been classified as type A or B (figure 1) (See ‘Classification’ above.)
- The occurrence of a nuchal cord(s) appears to be a random event, with increased risk among fetuses with excessive movement and/or a long umbilical cord. (See ‘Pathogenesis’ above.)
- A nuchal cord can form at any gestational age, but appears to be more common at term. It may persist, disentangle, or reform. Type B nuchal cords are more likely to persist than type A nuchal cords (figure 1). (See ‘Incidence’ above and ‘Natural history’ above.)
- The incidence of nuchal cords at term ranges from 15 to 34 percent of births. Ninety percent are single. (See ‘Incidence’ above and ‘Natural history’ above.)
- The prenatal diagnosis of an incidental nuchal cord is based on an ultrasound examination documenting that at least 75 percent of the neck is encircled by umbilical cord (movie 1 and movie 2). Color Doppler imaging or Doppler flow velocimetry (image 2) helps to confirm a diagnosis made by gray-scale imaging (image 1). The sensitivity of ultrasound for detecting nuchal cords at term has been reported to be approximately 70 percent for gray-scale imaging and 83 to 97 percent with color Doppler. (See ‘Diagnostic performance of ultrasound’ above.)
- Although available data are of low quality, the body of evidence suggests that nuchal cords are not associated with a significant increase in the rate of any clinically important adverse fetal/neonatal event.

However, in case reports and small case series nuchal cords have been associated with serious adverse outcomes, such as fetal demise and impaired fetal growth, as well as an increased rate of intrapartum fetal heart rate...
abnormalities leading to an increased rate of operative delivery. (See 'Possible sequelae during pregnancy' above.)

- We suggest not screening for nuchal cords during pregnancy (Grade 2C). There is no high or even moderate quality evidence that nuchal cords adversely affect pregnancy outcome or that prenatal diagnosis of nuchal cords improves pregnancy outcome. (See 'Screening' above and 'Possible sequelae during pregnancy' above.)

- An incidental finding of a nuchal cord on ultrasound examination does not warrant a change in prenatal or intrapartum care, given the lack of evidence of a clinically significant increase in adverse pregnancy outcome. However, we do not attempt external cephalic version of the breech fetus with a nuchal cord. (See 'Antepartum' above and 'During labor' above.)

- We do not report incidental detection of a nuchal cord, as it can be considered a normal finding. For patients who ask about a nuchal cord, we reassure the patient that a nuchal cord is a common finding that often resolves and that persistent nuchal cords have not been associated with a markedly increased risk of adverse pregnancy outcome. As in all pregnancies, the patient should be instructed to report decreased fetal movement to her provider. (See 'Screening' above.)

- If the cord is around the neck after expulsion of the fetal head, slipping the cord over the head or slipping it over the shoulders and delivering the body through the loop usually successfully frees the fetus from the tether. If delivery of the shoulders and body are compromised by a tight single or multiple nuchal cord that is not reducible, we perform a "somersault maneuver." If this is not successful, the cord may be clamped and cut. (See 'Delivery' above.)

- Fetal demise due to strangulation is rare. The presence of a nuchal cord alone is insufficient evidence of causality. The presence of the following findings supports causality in these rare cases (see 'Fetal demise' above):
  - One or more nuchal cords indenting fetal neck tissue
  - Petechial hemorrhages of the head and neck
  - Vascular congestion and thrombosis of umbilical cord vessels
  - Cord edema or hemorrhage into the Wharton's jelly
  - Vascular ectasia, vascular thrombosis, and/or thrombotic vasculopathy (avascular villi, villous stromal karyorrhexis) involving the umbilical cord vessels and/or chorionic plate and stem villous vessels
  - Absence of other conditions associated with fetal demise

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**GRAPHICS**

**Type A and Type B nuchal cords**

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<th>Type A</th>
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Type A: The placental end crosses over the umbilical end, entangling the neck in an unlocked pattern. This pattern can become undone with fetal movement.

Type B: The placental end crosses under the umbilical end, entangling the neck in a locked pattern. This pattern cannot undo itself and can form a true knot when it passes caudally over the fetal body.


Graphic 108849 Version 1.0
Gray-scale and color Doppler images of a double nuchal cord

(A) Gray-scale ultrasound.
(B) Color Doppler ultrasound.

*Courtesy of Leonhard Shaffer, MD, and Roland Zimmermann, MD.*

Graphic 54917 Version 4.0
Doppler ultrasound showing nuchal cord

Courtesy of Leonhard Shaffer, MD, and Roland Zimmermann, MD.

Graphic 67672 Version 3.0
Somersault maneuver for delivery of the fetus with a tight nuchal cord

Modified from:
2. East Carolina University College of Nursing. Somersault maneuver demo. Available at: https://www.youtube.com/watch?v=WaJ6sZ4fnQ (Accessed on June 29, 2016).

Graphic 107944 Version 1.0
Contributor Disclosures

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