Fetal demise due to cord entanglement in the early second trimester

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In this report, we describe a rare cause of in utero fetal death, a complex entanglement of the umbilical cord around the fetal neck. At the 16th gestational week of pregnancy, routine fetal ultrasonography showed no fetal heartbeat. Thereafter, the fetus was delivered vaginally in the breech presentation. The neck was found to be encircled by multiple tight loops of the umbilical cord. Other than a thin and elongated neck, there were no dysmorphic features and no chromosomal abnormality on cytogenetic analysis.

Cord entanglement is a common finding in utero; however, fetal demise resulting from nuchal cord entanglement is rare (1–8). This report describes such a case.

CASE PRESENTATION

The 37-year-old patient had no history of familial genetic disease and no systemic disease. In her previous pregnancy, she delivered a healthy baby girl vaginally at the age of 31 years. The present pregnancy was conceived naturally. Screening completed at the 12⁺³ gestational week, which included fetal nuchal translucency measurement, maternal serum pregnancy-associated plasma protein A levels, and free beta-human chorionic gonadotropin levels, showed no increased risk for trisomies 21, 18, and 13. Doppler evaluation of maternal uterine arteries was within normal limits. At the last routine visit at the 16th gestational week, there was no fetal heartbeat. The fetus was delivered vaginally and was in the breech presentation. The fetus's neck was encircled by multiple tight loops of the umbilical cord and had a reduced diameter (*Figure*). The umbilical cord was 35 cm long, with a sectional diameter of 0.6 cm. There were no dysmorphic features macroscopically and no congenital anomalies. Cytogenetic analysis showed 46, XY normal pattern.

DISCUSSION

The *Table* lists studies related to the incidence of nuchal cord and two case reports with fetal demise due to nuchal cord (1–8). The reported incidence of nuchal cord varies, with the highest rates of 43% in the 13th to 16th gestational week and 8.3% in newborns (1, 6). The varying rates of incidence depend mainly on the definition of cord entanglement. These studies

Figure. A fetus with a thinned and elongated neck, encircled by multiple tight loops of the umbilical cord.

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Table. Previously reported cases of nuchal cord entanglement

Reference number	First author	Year of publication	Number of cases	Study design	Frequency	Study outcome
1	Lipitz S	1993	1016 newborns	Retrospective + prospective	8%	No effect on term birth weight
2	Larson JD	1995	326 newborns	Retrospective	4%	No effect on neonatal outcome. Increased risk of abnormal fetal heart rate pattern, operative vaginal delivery
3	Larson JD	1997	4029 newborns	Retrospective	29%	No effect on stillbirth rate
4	Singh V	2003	1 intrauterine fetal death (IUFD)	Prospective	8%	Cord constriction, coiling, hemorrhage, thrombosis, amniotic bands as frequent causes of IUFD
5	Ghi T	2007	5 newborns	Prospective	3%	No effect on rate of vaginal delivery after induction
6	Tepper R	2009	64 fetuses	Prospective	43%	A 63% rate of any cord entanglement in early fetal life
7	Gambhir PS	2011	3 cord entanglements	Case report	—	Died (3), one due to nuchal cord
8	Dodds M	2012	3 nuchal cord entanglements	Case report	—	Died (1)

generally support that nuchal cord is a common finding in fetal life, with decreasing rates in the newborn (1, 2, 5, 6). However, rather than showing decreasing rates, one study showed that nuchal cord rates increased linearly from 6% in the 20th gestational week to 29% in the 42nd gestational week, regardless of whether the entanglement involved a single loop or multiple loops (3).

The presence of nuchal cord seems to have no effect on the outcome of pregnancy in terms of birth weight, rate of stillbirth, or rate of vaginal delivery after induction (1–3, 5). Compared to no entanglement or only one cord entanglement, two or more cord entanglements have been associated with an abnormal fetal heart rate pattern, requiring more low or midforceps application with a lower 1-minute Apgar score and an umbilical artery pH ≤ 7.10 without any adverse neonatal outcome (2). Umbilical cord abnormalities, mainly umbilical cord constriction and coiling, have been shown to be related to 11% of intrauterine fetal deaths within 16 gestational weeks (4). Fetal demise due to nuchal cord entanglement has been reported to occur in the first or second trimester in two case reports (7, 8).

A decreasing incidence of cord entanglement around the neck seems to be a normal phenomenon during fetal uterine development, but rare instances result in fetal demise. An autopsy study of 139 mostly second-trimester fetuses that died due to umbilical cord stricture and overcoiling revealed the absence of Wharton's jelly as an intrinsic cord pathology (9). Likewise, a not-yet-clarified intrinsic umbilical cord anomaly associated with the physical compression of entanglement may

explain why some rare cases of nuchal cord end in fetal demise. To date, nuchal cords have been shown to have significantly lower vascular coiling than ones without nuchal entanglement (10). The relevance of this finding in terms of fetal demise is not yet known.

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