Internal MedicineToday
Gonabad University of Medical Sciences
May 2024. Vol 30. Issue 3

Case Report

A Neonate with a Single Umbilical Artery, Bilateral Clubfoot, a Sextuple Nuchal Cord, and a True Knot: A Case Report

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Citation Balgheisi Naseri B, Ashrafi S, Faeal Nazari AS, Jalili Hashemi Z, Narjes Bahri N. [A Neonate with a Single Umbilical Artery, Bilateral Clubfoot, a Sextuple Nuchal Cord, and a True Knot: A Case Report]. Internal Medicine Today. 2024; 30(3): 94-99

:https://doi.org/10.22034/imtj.2024.30.3.94

Received: 12 Jan 2024
Accepted: 22 Mar 2024
Available Online: 30 May 2024

Key words:
Clubfoot
Nuchal Cord
Single Umbilical Artery
True Knot

ABSTRACT

Aims The umbilical cord is a vital conduit between the placenta and the fetus during intrauterine life, and loses its physiological function after birth. Neonates can present with various umbilical cord anomalies. This study aimed to report a rare case of sextuple nuchal cord associated with a true knot, combined with a single umbilical artery and bilateral clubfoot.

Case Presentation A 26-year-old pregnant woman presented with premature membrane rupture and regular contractions. Fetal ultrasonography at 19 weeks of gestation revealed a single umbilical artery and bilateral clubfoot.

Findings Following the appearance of late decelerations unresponsive to supportive measures, the patient underwent an emergency Cesarean section. During the procedure, a sextuple nuchal cord and a true knot were observed. The neonate weighed 3100 g and presented with bilateral clubfoot and a single umbilical artery.

Conclusion This case demonstrates that the presence of multiple nuchal cords combined with a true knot, even in the presence of good Apgar scores, may be associated with abnormal fetal heart rate patterns. Prompt decision-making in such scenarios can prevent adverse outcomes.

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Introduction

The umbilical cord is a vital conduit between the placenta and the fetus during intrauterine life, losing its significance after birth. Numerous anomalies of the umbilical cord exist; if left undiagnosed or improperly managed, they can lead to serious complications [1]. One such anomaly is the single umbilical artery (SUA), which is observed in 0.5% to 2% of pregnancies [2]. Fetuses with SUA are at a higher risk for structural defects, such as cardiovascular and genitourinary anomalies, as well as chromosomal disorders, such as aneuploidies [3]. Consequently, fetal echocardiography and amniocentesis are recommended upon identification of this condition [4]. However, if the fetus appears normal on targeted ultrasound scans in other respects and the pregnancy is otherwise low-risk, an isolated SUA does not increase the risk of fetal aneuploidy [5].

A nuchal cord is another umbilical cord anomaly defined as the presence of cord loops around the fetal neck [6]. The incidence of nuchal cord at birth increases with gestational age, estimated to be between 19% and 24%. Furthermore, a single loop is more common than multiple loops [7]. The effects of nuchal cord on perinatal outcomes remain controversial [8]. Previous research suggests that the presence of multiple tight cord loops during labor is associated with an increased incidence of abnormal fetal heart rate patterns, umbilical artery blood acidemia, meconium passage, and, consequently, an increase in emergency Cesarean deliveries [9, 10]. Nevertheless, recent studies have reported that nuchal cord is not associated with an increased incidence of adverse perinatal outcomes [11-13].

True umbilical knots occur in approximately 0.3%-1.3% of deliveries [14]. These knots, formed by fetal movements, are associated with risk factors, such as polyhydramnios, diabetes, chronic hypertension, monochorionic-diamniotic twinning, long umbilical cord length, and small or male fetuses [15]. True knots can also be associated with an increased risk of fetal intrauterine demise (IUFD) [16, 17].

The following case report describes an emergency Cesarean delivery of a fetus presenting with a SUA, bilateral clubfoot, sextuple nuchal cord, and a true knot, necessitated by recurrent late decelerations.

Study Type

This case report presents a rare and clinically significant case. This manuscript was structured according to the CARE checklist guidelines for case report articles [18]. This study was approved by the Research Council of the Reproductive Health and Population Research Center, Gonabad University of Medical Sciences, Gonabad, Iran, and holds an ethics code from the Regional Committee of Bioethics in Research of Gonabad University of Medical Sciences, Gonabad, Iran.

Patient Presentation

The patient was a 26-year-old Iranian woman with a history of three pregnancies, one delivery, and one spontaneous abortion. She presented to the Midwifery Emergency Department of Chamran Hospital in Ferdows, South Khorasan Province, Iran, complaining of premature rupture of membranes. Her gestational age, based on ultrasound, was 40 weeks and 1 day, while based on the first day of her last menstrual period (LMP), the gestational age was exactly 40 weeks. The parturient reported a past medical history, including a Dilation and Curettage (D&C) procedure. Her previous delivery was a spontaneous vaginal delivery of a male neonate weighing 3100 g, following which the mother experienced uterine atony requiring a blood transfusion.

A targeted anomaly scan performed at 19 weeks of gestation revealed a SUA and bilateral clubfoot (Figure 1).

Dear Madam,

By ultrasonography, a living fetus with normal cardiac activity and normal ectopy, with normalamniotic fluid volume relative to gestational age, was observed.

Measurement	Value	Gestational Age
HC	169 mm	19w, 4d
FL	31 mm	19w, 5d
AC	141 mm	19w, 3d
HL	29 mm	19w, 5d

Given the patient's obesity, a full evaluation was performed. In the review, no anomalies were found in the long bones, skull, CNS, spine, and facial structures of the fetus.

Currently, the nuchal fold (NF) is 4/7 mm, the nuchal translucency (NB) is 5/8 mm, and the anteroposterior (AP) diameter is 3/9 mm. The heart in the four-chamber view is grossly free of pathology. The fetus's stomach, intestines, and kidneys show normal views. The umbilical artery is a solitary vessel.

The lower limbs showed normal clubfoot bilaterally on the image.

The cervical canal was not visualized during the fundal scan (sonography for abdominal circumference was 28 mm via the endocervical approach).

(Normal sonographic findings do not rule out delayed anomalies.)

Figure 1. 19-Week Gestational Ultrasound (Reported Single Umbilical Artery and Bilateral Clubfoot), Clinical Findings (CARE Item 6)

On initial vaginal examination, findings confirmed a cervical dilation of 2 cm, 30% effacement, a fetal head station at −3, cephalic presentation, ruptured membranes, and clear amniotic fluid. The maternal vital signs and non-stress test (NST) were normal. Contractions occurred every one to two minutes at moderate intensity, lasting an average of 45 seconds.

Management Interventions

To enhance uterine contractions and promote cervical ripening, sublingual misoprostol tablets were administered, and nitric oxide gas was administered for analgesia. The initial NST showed variable decelerations, which resolved following supportive measures (Figure 2).

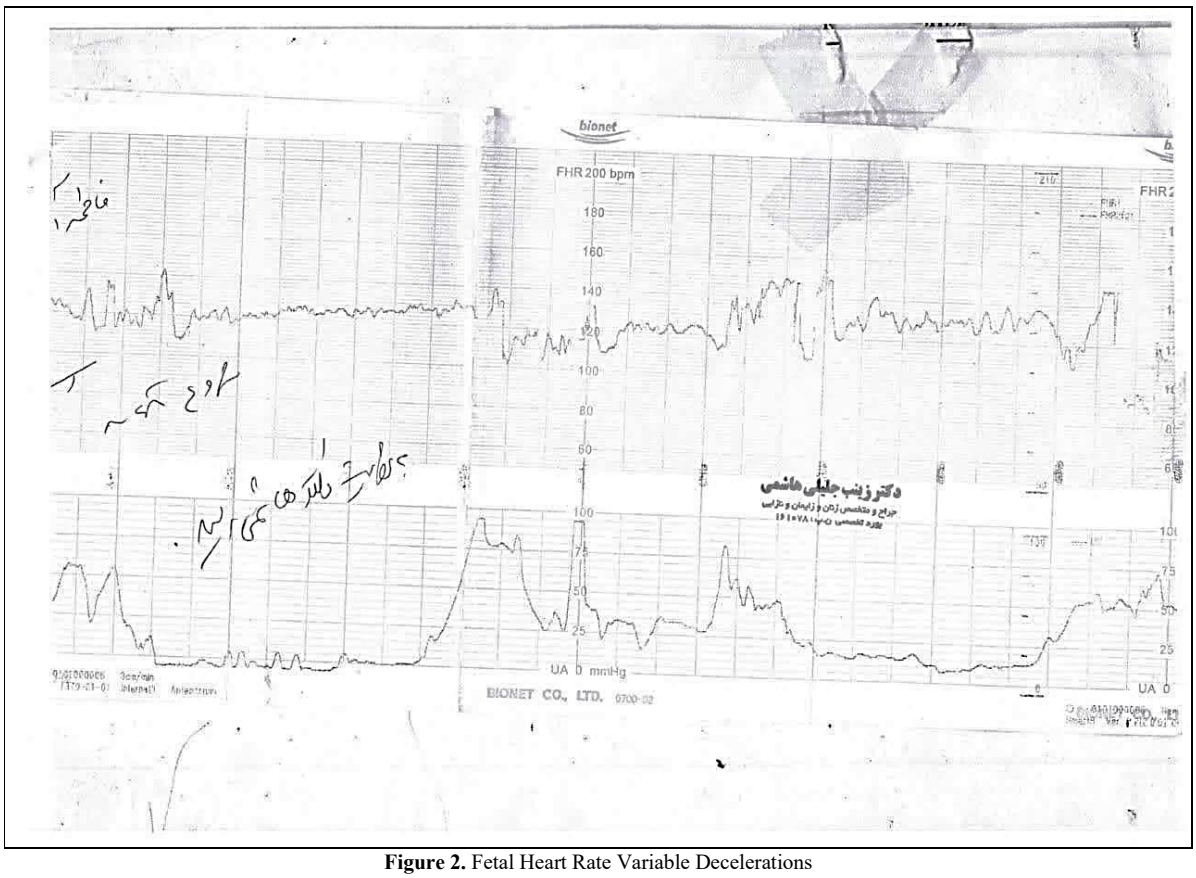

Figure 2. Fetal Heart Rate Variable Decelerations

Subsequently, recurrent late decelerations appeared, which were unresponsive to supportive measures (Figure 3). Ultimately, the mother, at 4 cm dilation and 40% effacement, was prepared for cesarean delivery. Under spinal anesthesia, a cesarean section was performed via a Pfannenstiel skin incision and a Kerr low transverse uterine incision. During the cesarean section, a sextuple nuchal cord and a true knot were observed. The delivery resulted in a live male neonate weighing 3100 g, with an Apgar score of 9 at one minute and 10 at five minutes. Examination of the placenta revealed a SUA, and the neonate exhibited bilateral clubfoot malformation.

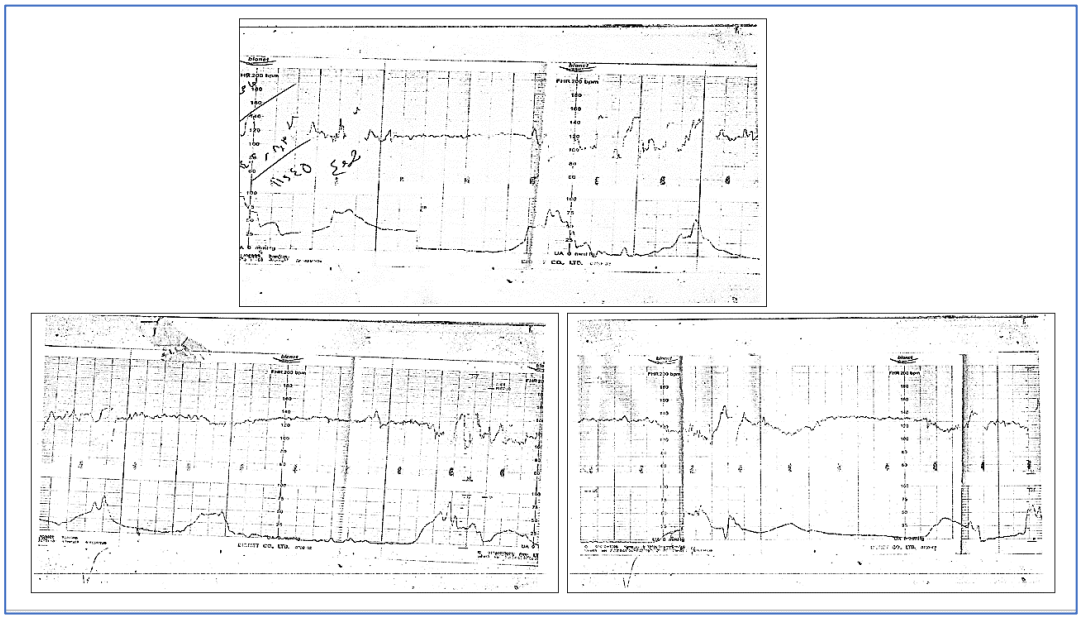

Figure 3. Fetal Heart Rate Late Decelerations

Discussion (CARE Items 11a–11d)

A. Single Umbilical Artery (SUA)

A single umbilical artery (SUA) is one of the most common anomalies of the umbilical cord. The mechanism underlying the development of SUA has not yet been clearly identified. In general, three theories have been proposed: The first theory suggests that only one umbilical artery is present during embryonic development. The second theory proposes that two umbilical arteries initially exist, but one gradually regresses during later stages of development. The third theory postulates that the single umbilical artery represents a persistent primitive allantoic artery that failed to develop further [19].

SUA can be associated with structural defects and chromosomal abnormalities [3]. In this regard, a population-based study utilizing data from 918,933 singleton pregnancies over 16 weeks of gestation in Norway between 1999 and 2014 reported an SUA incidence of 0.46%. Factors, such as fourth or higher parity, smoking, pre-gestational diabetes, epilepsy, chronic hypertension, and assisted reproductive technologies, increased the risk of SUA. Furthermore, a strong association was found between SUA and gastrointestinal atresia, renal agenesis, cardiac anomalies, and central nervous system anomalies. Hernia diaphragmatica, limb reduction defects, and cleft lip/palate showed a weaker association with SUA. Trisomy 13 and 18 also showed a strong association, and the risk for Trisomy 21 was doubled [20]. Another study reported that isolated SUA often occurs with multiple tissue anomalies; for instance, in an autopsy series of 1277 fetuses, 60% of SUA cases were accompanied by other anomalies, including musculoskeletal defects [21].

In a retrospective cohort study examining the association between isolated SUA and adverse pregnancy outcomes/congenital anomalies among 116,501 singleton pregnancies in Denmark, researchers stated that SUA increased the risk of stillbirth and small-for-gestational-age (SGA) neonates but had no effect on the risk of preterm birth or cesarean delivery. Additionally, 3.4% of pregnancies with SUA in this study had undiagnosed congenital anomalies [22]. The results of another retrospective analysis of 1169 singleton pregnancies with prenatal diagnosis of SUA indicated that 84.6% of these fetuses had the isolated type, while 15.4% were accompanied by a structural or chromosomal anomaly. Structural anomalies included cardiovascular (9%), genitourinary (3.5%), gastrointestinal (3%), central nervous system (2.1%), and musculoskeletal (2.9%) [23].

B. Clubfoot

In the reported case, the neonate presented with bilateral clubfoot in addition to SUA. Congenital clubfoot is the most common musculoskeletal malformation of the lower limb extremity, occurring in approximately 1% of newborns. This anomaly, involving the bones, joints, and soft tissues of the distal lower limb, is twice as common in boys as in girls, and half of the cases are bilateral [24]. The etiology of clubfoot is largely unknown, but some studies suggest a higher prevalence in certain chromosomal defects compared to the general population [25]. Recent studies have investigated effective molecular and genetic pathways (including genes involved in bone and connective tissue development); however, a single definitive cause remains unidentified for most cases. Besides genetics, intrauterine factors, such as restricted space and neurological factors, may also play a role [26]. Although isolated clubfoot in most cases does not present a high risk for chromosomal abnormality and is usually not part of a genetic syndrome, it can sometimes be part of a more complex syndrome or be associated with other structural anomalies; for example, reports exist of its association with trisomies (such as Trisomy 18) [27].

C. Nuchal Cord and Umbilical Knot

In the present case, the fetus had a sextuple nuchal cord and a true knot, leading to cesarean delivery due to recurrent late fetal heart rate decelerations. A nuchal cord with multiple loops, similar to the reported case, is a rare phenomenon in obstetrics. The presence of six loops around the neck is highly uncommon and primarily reported in case reports within the literature. In other words, the evidence consists of isolated cases and very few collective studies; thus, six loops are classified as an "extremely rare" event [28].

The incidence rate of a nuchal cord with multiple loops at birth is less than 5%, and this rate decreases as the number of loops increases [29]. One study reported the incidence of a nuchal cord with three or more loops to be 0.5% [30]. For instance, a study evaluating the effects of nuchal cord and the number of loops on delivery outcomes in 42,798 singleton vaginal deliveries found that 8.9% had one loop, 2.4% had two loops, and 0.6% had three loops. Cases with three loops were associated with higher rates of fetal intrauterine death, Apgar scores less than seven at one and five minutes, and instrumental vaginal delivery compared to cases without a nuchal cord [31]. Nevertheless, in another study investigating the association between nuchal cord and electronic fetal health assessment parameters during labor, researchers reported that nuchal cord is associated with Class II NST patterns and instrumental vaginal delivery but does not affect fetal mortality [13]. Conversely, the results of a retrospective study comparing maternal, fetal, and neonatal outcomes in pregnancies with and without a true knot indicated that maternal age, BMI, parity, labor induction, meconium passage, preterm birth, cesarean delivery due to non-reassuring fetal heart rate patterns, and intrauterine death after 37 weeks were significantly higher in the true knot group; however, no significant difference was observed in the five-minute Apgar scores between the two groups [32].

Strengths and Limitations

This case report has significant scientific value due to the simultaneous presentation of several rare anomalies: SUA, bilateral clubfoot, sextuple nuchal cord, and a true knot. Accurate recording of intrapartum findings, documentation of fetal heart rate patterns, and the inclusion of images from

the anomaly scan and NST are key strengths, enabling more precise analysis of the heart rate deceleration mechanisms and clinical decision-making. Furthermore, the combination of these anomalies in one fetus is highly uncommon, and its presentation can improve clinical awareness and rapid decision-making when encountering abnormal heart rate patterns.

The limitations of this report include the absence of long-term follow-up for the neonate to assess the course of clubfoot treatment and its potential outcomes. Moreover, **because case reports are by** nature **limited to** a **single case**, generalization of findings to a wider population is not possible. The lack of fetal genetic testing (given the normal status of other findings) prevented a more precise analysis of the potential link between the constellation of anomalies. Furthermore, the inherent limitations of case reports, such as the absence of a control group and the inability to establish causality, are also present in this study.

Conclusion

This case report demonstrates that the concurrence of anomalies, such as SUA, bilateral clubfoot, a sextuple nuchal cord, and a true knot, can be associated with significant changes in the fetal heart rate pattern, even when other pregnancy findings appear normal. The occurrence of late decelerations resistant to supportive measures in this patient clearly highlights the importance of continuous fetal monitoring and the necessity for prompt decision-making regarding pregnancy termination. Timely diagnosis and appropriate intervention play a crucial role in preventing adverse perinatal outcomes. Given the rarity of such a combination of anomalies, further studies are recommended to investigate the potential association of these findings with short- and long-term outcomes in neonates.

Ethical Considerations

Compliance with ethical guidelines

This study was conducted in accordance with the ethical principles of the Declaration of Helsinki. The patient's data were completely anonymized and rendered unidentifiable. Written informed consent was obtained from the mother prior to publication for the use of clinical information, images, and the dissemination of this case report. The principles of confidentiality, patient privacy, and non-disclosure of any identifying information were fully adhered to. This study was approved by the Regional Committee of Bioethics in Biomedical Research (ethics code: IR.GMU.REC.1403.072).

Acknowledgments

The authors express their gratitude to the management and all midwifery staff at Chamran Hospital in Ferdows for their cooperation in the patient's care and accurate recording of clinical data, and to the neonate's mother for providing informed consent, which facilitated the preparation and publication of this case report. This project was approved by the Research Council of the Reproductive Health and Population Research Center, Gonabad University of Medical Sciences, Gonabad, Iran. Financial support from Gonabad University of Medical Sciences, Gonabad, Iran, for this project is also appreciated (grant number: 1738).

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