

Cord Entanglement in Monoamniotic Twin Pregnancy: A Case Report and Review of the Literature

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Monoamniotic twin gestations have been associated with increased perinatal mortality rates of up to 30-50%. Umbilical cord entanglements, twin-to-twin transfusion syndrome, congenital malformations, intrauterine growth restriction, and prematurity are responsible for the high perinatal morbidity and mortality. Cord entanglement can potentially compromise the cord blood flow and cause fetal demise. There is still no consensus on the antenatal management and timing of delivery of these rarely encountered cases. Here, we present a case of monoamniotic twin pregnancy with cord entanglement diagnosed in labour in a 21-year-old primigravid woman and review the obstetric literature about the management of monoamniotic twin pregnancies.

Key Words: Monoamniotic twin pregnancies, Fetal cord entanglement

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Introduction

Monoamniotic twinning is diagnosed when a twin pregnancy is seen in a single amniotic sac with a single placenta. It is a rare event, which occurs in about 1% of monozygotic twins.¹ Prevalence of perinatal death is increased in monoamniotic twins.² It has been suggested that the cord entanglement is the one of the main causes of fetal death.³ Umbilical cord entanglement is found in all monoamniotic twins when it is systematically evaluated by ultrasound and color Doppler.⁴ There is still controversy regarding the antenatal management of these rarely encountered cases. Here we present a case of monoamniotic twin pregnancy with cord entanglement diagnosed in labour and review the obstetric literature about cord entanglement and management of monoamniotic twin pregnancies.

Case Report

A 21-year-old primigravid woman was admitted to the obstetric unit with the diagnosis of spontaneous twin gestation in labour. Her last menstrual period was unknown. She had been diagnosed to have a twin pregnancy at 12 weeks of gestation.

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She received irregular antenatal care. When she was admitted for labour, ultrasonographic examination revealed a monoamniotic twin pregnancy at 35-37 weeks of gestation with vertex-breech presentation. The monochorionic placenta was lying anteriorly and the umbilical cords inserted close to one another, with multiple connections of the vessels of both twins. Amniotic fluid measurement, umbilical artery Doppler examinations and fetal heart rate patterns of both twins were in normal range. Cesarean section was determined to be the optimal route of delivery due to monoamniotic twinning. During the operation, it was noted that the cords were intertwined and formed a loose knot (Figure 1) at 15 cm distance from the placental insertion (Figure 2). Two female babies weighing 2540 and 2420 g were delivered with normal Apgar scores. The babies were physically normal and had an uneventful neonatal period.



Figure 1: Loose knot formed by umbilical cords.



Figure 2: Entangled umbilical cords and monochorionic placenta.

Discussion

Monoamniotic twinning occurs with an estimated prevalence of between 1 in 5000 and 1 in 25 000 pregnancies.¹ These twins share a single placenta with two umbilical cord insertions which are frequently close together as well as a single amniotic sac and are thought to arise from a late cleavage of the inner cell mass. Because of the improved antenatal care, mortality rates in monoamniotic twins has declined from 30-70%⁵ to 10-20%.⁶ The high perinatal mortality rate is mainly due to entanglement and knotting of the umbilical cords and another complications in monochorionic twin pregnancies, such as conjoined twins, preterm delivery, low birth weight and twin-to-twin transfusion syndrome (TTTS). Studies which excluded pregnancy losses from conjoined twins, TTTS, discordant anomaly and spontaneous miscarriage before 20 weeks, reported perinatal loss rates as low as 5-10%.^{1,7} The largest literature review of 133 monoamniotic twin pregnancies reported the prevalence of cord entanglement as 95%; thirty of 93 pregnancies were diagnosed on ultrasound and, 58 at delivery.⁸ The antepartum mortality rate is related to the site of entanglement, higher when close to the fetus than to the placenta.⁹

Cord entanglement can be diagnosed by using Doppler ultrasound. It was noted as early as 12 weeks of gestation in some monoamniotic pregnancies.⁴ In a prospective observational study by Dias et al., cord entanglement was diagnosed by B-mode and color Doppler ultrasound at 11-16 weeks' gestation.⁴ Kuwata et al. demonstrated structural and morphological features of cord entanglement by three-dimensional color Doppler in their case report and stated that it was possible to display cord entanglement's accurate proximity to the fetus and placenta by using three-dimensional color Doppler.¹⁰

Structural anomalies occur more frequently, affecting up to

20% of monoamniotic twin pregnancies compared with 6% of diamniotic twins.^{2,11,12} Thus, monoamniotic twins deserve detailed sonographic follow up by experienced sonographers.

There is still no consensus about the optimal antenatal management and timing of delivery in monoamniotic twin pregnancies. Monoamniotic twins are at increased risk of unexpected fetal demise. Fifteen percent of monoamniotic twin pregnancies were complicated by in-utero fetal death after 20 weeks.¹³ After 32 weeks of gestation, incidence of fetal demise was 4% in monoamniotic twin pregnancies, while it was 1.2% in diamniotic twins.¹² The risk of in-utero fetal death is largely attributed to umbilical cord compression.⁷ Fetal movements may increase tightening of an already entangled umbilical cord which could lead to occlusion, fetal distress and death.⁷ However, as almost all monoamniotic pregnancies are complicated by cord entanglement, other mechanisms have also been suggested. Acute exsanguination across the large caliber anastomoses probably triggered by cord compression might be an important cofactor.^{1,2}

Although fetal death is usually unexpected, it has been suggested that elective preterm birth and careful surveillance of monoamniotic twins may improve survival rates.^{6,14} Recommended timing of delivery is between 32 and 35 weeks of gestation.⁸ Lewi stated that delivery at 32 weeks carried a smaller risk of death than remaining in utero, as at this week, the risk of mortality of a moderate preterm birth appears to be four times lower than that of remaining undelivered.¹⁵ According to available literature, about 1 in 25 monoamniotic pregnancies is complicated by fetal demise after 32 weeks.¹³

Intensive fetal surveillance in monoamniotic twin pregnancies should be started when viability is achieved.^{3,16,17} The optimal method of fetal surveillance remains unclear. In a prospective case series, Pasquini et al. reported that the only reliable method for fetal monitorization was Doppler ultrasonography and cardiotocography had high false negative rates for fetal distress.⁷ Antenatal corticosteroids for fetal lung maturation is recommended because urgent delivery may be necessary.^{7,17} The hospitalization requirement is also controversial and should be individualized based on antenatal findings, although in-patient management has been reported to improve the survival rates.^{1,2,18} Most authors would offer cesarean birth, because the risk of vaginal delivery related primarily to cord entanglement and cord compression during labour, especially after delivery of first infant is considerably high. Pasquini et al. suggested that the use of sulindac, a non-steroidal anti-inflammatory drug of the arylalkanoic acid class, to create oligohydramnios, could reduce the cord accidents and improve the perinatal surveillance.⁷ On the other hand, due to potential side effects and no definite proof of benefit, its use has not gained widespread popularity.

In the case presented here, the entangled cord formed a loose knot far from the fetus. Therefore we suggested that it did not compromise the fetal circulation and pregnancy advanced to the third trimester without any complications.

In conclusion, monoamniotic twin pregnancies are associated with a high perinatal mortality. Umbilical cord entanglement in these pregnancies is a biologically plausible phenomenon that can be demonstrated antenatally in all monoamniotic twins. Although fetal demise can not be completely preventable, intensive fetal surveillance when viability is achieved can lead to improved outcome. On the other hand, parents should be warned that sudden fetal death can occur in spite of close surveillance. In view of the rarity of this event, large prospective studies are needed to assess the optimal antenatal management and timing of delivery.

Monoamniyotik İkiz Gebelikte Kordon Dolanması: Vaka Sunumu ve Literatürün Gözden Geçirilmesi

Monoamniyotik ikiz gebelikler %30-50'lere varan artmış perinatal mortalite oranları ile birliktedir. Bu tür gebelikler umbilikal kordon dolanması, ikizden ikize transfüzyon sendromu, konjenital malformasyonlar, intrauterin gelişme geriliği ve prematürite yüksek perinatal mortalite ve morbiditeden sorumludur. Bu durumlardan özellikle kordon dolanması, kan akımını bozabilme ve fetal kayıba neden olabileme potansiyeli taşıması nedeniyle önemlidir. Literatürde, bu nadir rastlanan vakaların antenatal takibi ve doğumun zamanlanması konusunda halen fikir birliği yoktur. Bu vaka sunumunda, 21 yaşında primigravid bir hastada, doğum eylemi sırasında tanısı konulmuş, kordon dolanması ile birlikte olan bir monoamniyotik ikiz gebelik vakası takdim edildi ve monoamniyotik ikiz gebeliklerin takibi ve tedavisi konusunda obstetrik literatür incelendi.

Anahtar Kelimeler: Monoamniyotik ikiz gebelikler, Fetal kordon dolanması

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