Monoamniotic Twins: A Case Series

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Abstract

Monoamniotic twinning is a rare event associated with a high perinatal morbidity and mortality. In this article, six pairs of monoamniotic twins diagnosed prenatally during 2001-2003 were evaluated. The management protocol included documentation of chorionicity and amnionicity by early ultrasonographic examination, 2-4 weekly serial ultrasound surveillance, antepartum fetal heart rate monitoring initiated at 28 weeks of gestation and a non-interventional approach beyond 32 weeks. In 5 cases, the detection of monoamnionicity was made during the first trimester. Cord entanglement was found in four cases. In only one case (acardiac twin) was there discordance for structural abnormality. In all cases, the pregnancy continued beyond 20 weeks, and with a non-interventional management policy, a live birth rate of 80% beyond 24 weeks was achieved. We believe that, at any given gestation, if prematurity complications are likely to outweigh the risk of an uncertain IUFD, a non-interventional management may be a reasonable option.

Keywords: monoamniotic twin pregnancy, perinatal management

Özet

Monoamniotik İkiz Gebelikler: Altı Olgunun Analizi

Monoamniotik ikiz gebelik nadir görülen ve yüksek perinatal morbidite ve mortaliteyle birlikte bulunan bir durumdur. Bu yazıda 2001 ve 2003 yılları arasında prenatal tanı konulmuş olan 6 monoamniotik ikiz gebelik olgusu sunulmuştur. İzlem protokolümüz erken gebelik ultrasonografisi ile koryonisite belirlenmesi, 2-4 hafta aralıklarla seri ultrason takibi, 28. haftadan başlayarak fetal kalp hızı monitörizasyonu ve acil hallerin dışında 32. gebelik haftasından sonra da gebeliğin sonlandırılmadan izlenmesiydi. Beş olguda monoamniotik ikizlik tanısı ilk trimesterde konuldu. Dört olguda kordonların birbirine dolanmış olduğu görüldü. Bir olgu akardiyak ikiz tanısı aldı. Tüm olgularda gebelik >20 hafta sürdü. Yirmi dört hafta aşıldığında canlı doğum oranı %80 olarak saptandı. Erken doğum komplikasyonlarının tam olarak belirlenemeyen bir *in utero* fetal ölüm riskiyle dengelenmesi halinde monoamniotik ikiz gebeliklerin >32. haftada girişimde bulunmaksızın izlenmesi makul bir seçenek olabilir.

Anahtar sözcükler: monoamniotik ikiz gebelik, perinatal izlem

Introduction

Monoamniotic twins account for approximately 1% of monozygotic twins, and they are at increased risk of preterm delivery and fetal death (1). Many deaths are reportedly caused by cord entanglement, congenital anomalies, and twin-twin transfusion syndrome (2). Recent studies indicate a lower perinatal mortality (10%-12%) than the previously cited risk of 30%-70%. There are different opinions in the literature regarding the proper prenatal monitoring of monoamniotic twins, the risk of fetal death after 32th weeks of gestation and the optimal timing and route of delivery (3-6). The aim of our study was to evaluate the outcome of six monoamniotic twin pregnancies diagnosed prenatally.

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SSK Bakırköy Maternity and Children's Hospital is a referral center for the European side of Istanbul and Thrace with approximately 22 000 deliveries a year. From 2001 to August 2003, 8 monoamniotic twin pregnancies were identified. Two of them were conjoined twins and termina-ted at second trimester. Management protocols employed were those of documentation of chorionicity and amnionicity by early ultrasonographic examination, 2-4 weekly serial ultrasound surveillance, antepartum fetal heart rate monitoring initiated at 28th weeks of gestation and a non-interventional approach beyond 32 weeks, unless an urgent need for delivery occurred. Gestational age at diagnosis, gestational age at delivery, indication for delivery, placental characteristics and the perinatal outcome are shown in Table 1.

Case 1

A 40-year-old woman in her third pregnancy was found to have a monoamniotic twin pregnancy during an ultrasonographic scan at 17th weeks. Genetic counseling was performed for advanced maternal age. However, the parents rejected genetic amniocentesis. A second scan at 23th weeks

Table 1. Outcomes of the presented monoamniotic twins. IUFD=Intrauterine fetal death, C/S=Cesarean section, Non-reass. FHR=Non-reassuring fetal heart rate trace

Case No.	Maternal age	Gestational age at diagnosis (weeks)	Gestational age at delivery (weeks)	Indication for delivery and mode of delivery	Umbilical cord pathology	Perinatal outcome
1	40	17	24	IUFD, vaginal	Cord entanglement true knot	IUFD (both twins)
2	29	11	35	Breech pres., C/S	Cord entanglement	Both twins alive
3	32	9	36	Non-reass. FHR, C/S	Cord entanglement	Both twins alive
4	38	11	32	Repeat C/S, C/S	SUA, velamentous insertion	twin A acardiac twin B IUFD
5	31	7	32	Spontaneous vaginal	Velamentous insertion	Both twins alive
6	25	13	33	Non-reass. FHR, C/S	Cord entanglement	Both twins alive

of gestation showed intrauterine death of one fetus. The next scan at 24 weeks demonstrated intrauterine fetal death (IUFD) of the other one. The delivery was induced with misoprostol. Cord entanglement with a true knot was noted at delivery.

Case 2

A 29-year-old woman in her first pregnancy presented for nuchal translucency screening at 11th weeks of gestation and was diagnosed to have a monoamniotic twin pregnancy. Two-weekly scans were performed. Fetal growth was normal. At 34 weeks of gestation, the patient was hospitalized because of minimal uterine contractions without cervical change. Fetal surveillance was continued as cardiotocographic (CTG) monitoring twice daily. Fetal heart traces of both fetuses were reactive when active labor began at 35+ weeks. A cesarean delivery was performed for breech presentation. The birthweights were 2190 and 2280 g. Cord entanglement was observed. Both babies were discharged well on the second day postpartum.

Case 3

Monoamniotic twinning was diagnosed at 9 weeks in a 32-year-old woman in her second pregnancy. Two-weekly fetal ultrasonographic surveillance was performed. At the 21 weeks scan both fetuses were thought to have mild ventriculomegaly. After genetic counseling, the parents elected the option of amniocentesis. The chromosomal analysis revealed a normal male karyotype. The pregnancy continued to be monitored for adequate fetal growth and to look for evidence of hydrocephaly. No progression of ventriculomegaly was observed in either fetus. Delivery was undertaken at 36 weeks by cesarean section for non-reassuring fetal heart rate pattern. The birthweights were 2080 and 2300 g. Umbilical cord intertwining was found at delivery and the monochorionicity was confirmed. Both babies were discharged and did well.

Case 4

A 38-year-old woman in her second pregnancy presented for nuchal translucency screening at 11+ weeks. In this ultrasound scan, a monoamniotic twinning and the IUFD of one twin of a pair with a marked discordance in crown-rump length (CRL) was diagnosed. Next antenatal visit was at 30+ gestational weeks. We performed an ultrasound scan and the diagnosis of acardiac twin was made. In both twins, single umbilical artery (SUA) was detected. There was a marked polyhydramnios. Management was as an in-patient with twice daily CTG. By the twelfth day after her admission, a non-reassuring CTG with uterine contractions was noted. A repeat cesarean delivery was planned following maternal steroid administration. Unfortunately, the fetus was found to be dead *in utero* before the operation. Autopsy confirmed the diagnosis of acardiac twin. In the other twin, cardiac hypertrophy and SUA was noted (Figure 1).

Case 5

A 31-year-old woman in her first pregnancy was found to have a monoamniotic twin pregnancy by ultrasound scan at 7+ weeks. Fetal surveillance was continued with two-weekly ultrasonography, and CTG monitoring was initiated at 28 weeks of gestation. At 32+ weeks, the patient presented for uterine contractions with a frequency of 2 contractions every ten minutes. Ultrasound scan revealed two live fetuses with vertex presentation and adequate fetal growth. Reassuring fetal heart rate traces were noted at continuous CTG monitoring. The woman delivered vaginally two healthy female fetuses weighing 1630 and 1640 g respectively. Placental examination confirmed monochorionic twinning and velamentous insertion of both umbilical cords (Figure 2). Both babies were discharged on the twelfth day postpartum and did well.

Case 6

A 25-year-old woman, G1P0, presented for nuchal translucency screening at 13 weeks gestation. A monoamniotic twin pregnancy was diagnosed. Delivery was undertaken at 33+ weeks by cesarean section because of non-reassuring CTG. The birthweights were 1790 and 1850 g. Cord entanglement was detected at delivery. The morphological examination of the placenta confirmed the diagnosis of monoamniotic twins. Both babies were discharged without complication.

Discussion

Monoamniotic twinning is a rare event. Approximately 1% of monozygotic twins are monoamniotic (1). Modern ultrasound technology with improvement in resolution has led to an

Figure 1. Case 4, acardiac twin. Note the velamentous insertion of the umbilical cord (arrow)

Figure 2. Case 5, the arrow indicates velamentous insertion of both umbilical cords without intertwining.

increasing number of reports of monoamniotic twins. The criteria for antenatal diagnosis of monoamniotic twins defined by Rodis et al. are absence of dividing membrane, a single placental mass, twins of same sex and enough amniotic fluid to allow free movements of both fetuses (3). The presence of one yolk sac in the first trimester and the demonstration of umbilical cord entanglement were characteristics of monoamniotic twin pregnancy (7).

A high perinatal mortality (30-70%) was reported in the past. However, a recent study suggests a lower perinatal mortality, being about 10% (8,9). The major factor contributing to perinatal morbidity and mortality is umbilical cord accident. Other contributors to the dismal outcome include congenital anomalies, preterm delivery, twin-twin transfusion and intrauterine growth retardation (9). In monoamniotic twins, different antenatal management protocols are employed. There is no consensus regarding optimal fetal monitoring and timing of delivery. However, it is widely agreed that antepartum fetal heart rate trace monitoring should be initiated at viability (10). Rodis et al. reported that of 13 sets of monoamniotic twins, 8 had cord entanglement noted at birth and the only indication for delivery was non-reassuring fetal heart rate patterns. These findings are contradictive with the previous belief that IUFD caused by cord entanglement would be sudden and therefore unlikely to be prevented by even frequent fetal heart rate monitoring (11). Similar findings are presented in our series. In cases 3, 4 and 6 non-reassuring fetal heart rate traces were the indication of delivery, and in two of them, cord entanglement was found. Varying frequencies of fetal heart rate monitoring (twice weekly to daily) were recommended in the literature (1,12). Although monitoring would not always prevent sudden death due to a cord accident, signs of repeated cord compression such as variable decelerations and bradycardia could be looked for (12).

A routine in-patient management of monoamniotic twins from 25 weeks onwards is an expensive intervention. It is reasonable to reserve this management policy for a selected group of patients in whom there is discordant fetal growth, variable decelerations in CTG and abnormal umbilical artery Doppler velocimetry (4). Timing of delivery in monoamniotic twins is also controversial. There is a delicate balance between the risk of prematurity and the risk of intrauterine fetal death. Some authors recommended delivery after fetal lung maturity. Most studies suggest delivery at 32 weeks in order to prevent cord-related deaths (1,4). Recently Roque et al. (13) reported a significant increase in the incidence of perinatal loss beyond 32 weeks among monoamniotic twins, suggesting that delivery after corticosteroid therapy should be strongly considered at this gestational age.

However, others do not agree with the idea that risks to monoamniotic twins necessarily increase as the gestational age advances past early third trimester (6). Tessen et al. (14) suggested that prophylactic preterm delivery offered no advantage. He also claimed that the risk of prematurity outweighed the risk of uncertain fetal death. Cesarean section is the strongly preferred mode of delivery in monoamniotic twins (15). Some authors suggested that labor and vaginal deliveries are not associated with an increased risk of fetal deaths. They concluded that vaginal delivery could be allowed for cases with normal Doppler findings and cephalic presentation of both twins (16).

In our cases, the management methods employed were early diagnosis of chorionicity and amnionicity, 2-4 weekly serial ultrasound surveillance, antepartum fetal heart rate monitoring initiated at 28 weeks of gestation and a non-interventional approach beyond 32 weeks, unless an urgent need for delivery occurs. However, parents were counseled regarding the risks and benefits of waiting and interventional managements respectively. Each woman was evaluated individually after consultation with neonatologists. As a result, vaginal delivery was preferred in case 5. In our small series, umbilical cord entanglement was present in 4

pregnancies, and abnormal cord insertion without entanglement was observed in the remaining two. The incidence of cord entanglement in monoamniotic twins was comparable with previous reports (17,18). Cord accidents may cause sudden death to one or both twins without warning signs. Although a non-reassuring fetal heart rate trace preceded delivery, the fetal death in case 4 could be considered as an example for such an event. In monoamniotic twin pregnancies the incidence of fetal anomaly of one twin is 40-50%. The presence of fetal anomalies was associated with a 42.9% perinatal mortality rate (13). In our series, fetal anomaly incidence was 16%.

With a non-interventional management policy, we have attained a live birth rate of 80% beyond 24 weeks. No neonatal death occurred. We suggest that in prenatally diagnosed monoamniotic twin pregnancies, timing of delivery should be an individualized decision for each unit. Our case series showed that, if prematurity complications are likely to outweigh the risk of an uncertain IUFD at a definite gestation; a non-interventional management may be a reasonable option.

References

1. Beasley E, Megerian G, Gerson A, Roberts N: Monoamniotic twins: case series and proposal for antenatal management. Obstet Gynecol 1999;93:130-134.
2. Dorum A, Nesheim BI: Monochorionic monoamniotic twins-the most precarious of twin pregnancies. Acta Obstet Gynecol Scand 1991;70:381-3.
3. Rodis JF, Vintzileos AM, Campbell WA, Deaton JL, Fumia F, Nochimson DJ: Antenatal diagnosis and management of monoamniotic twins. Am J Obstet Gynecol 1987;157:1255-7.
4. Aisenbrey GA, Catanzarite VA, Hurley TJ, Spiegel JH, Schrimmer DB, Mendoza A: Monoamniotic and pseudomonoamniotic twins: sonographic diagnosis, detection of cord entanglement, and obstetric management. Obstet Gynecol 1995;86:218-22.
5. Pessonnier A, Ko-Kivok-Yun P, Fournie A: Monochorionic, monoamniotic twin pregnancies: diagnostic problems. Outcome risk factors. J Gynecol Obstet Biol Reprod 1994;23:299-302.
6. Carr SR, Aronson MP, Coustan DR: Survival rates of monoamniotic twins do not decrease after 30 weeks' gestation. Am J Obstet Gynecol 1990;163:719-22.
7. Bromley B, Benacerraf B: Using the number of yolk sacs to determine amnionicity in early first trimester monochorionic twins. J Ultrasound Med 1995;14:415-9.
8. Demaria F, Goffinet F, Kayem G, Tatsaris V, Hessabi M, Cabrol D. Monoamniotic twin pregnancies: Antenatal management and perinatal results of 19 consecutive cases. Br J Obstet Gynecol 2004;111:22-26.
9. Allen VM, Windrim R, Barret J, Ohlsson A: Management of monoamniotic twin pregnancies: A case series and review of the literature. Br J Obstet Gyneacol 2001;108:931-36.
10. Blane CE, Dipietro MA, Johnson MZ, White SJ, Louwsma GI, Hamman JE: Sonographic detection monoamniotic twins. J Clin Ultrasound 1987;15:394-6.
11. Rodis J, McIlveen P, Egan J, Borgida A, Turner G, Campbell W: Monochorionic twins: improved perinatal survival with accurate prenatal diagnosis and antenatal fetal surveillance. Am J Obstet Gynecol 1997;177:1046-1049.
12. Lodeira JG, Vintzileos AM, Feinstein SJ, Campbell WA, Nochimson DJ: Fetal biophysical profile in twin gestations. Obstet Gynecol 1986;67:824-7.
13. Roque H, Gillen-Goldstein J, Funai E, Young BK, Lockwood CJ: Perinatal outcomes in monoamniotic gestations. J Matern Fetal Neonatal Med. 2003; Jun;13(6):414-21.
14. Tessen JA, Zlatnik FJ: Monoamniotic twins: a retrospective controlled study. Obstet-Gynecol 1991;77:832-4.
15. Annan B, Hutson RC: Double survival despite cord entwinement in monoamniotic twins. Case report. Br J Obstet Gyneacol 1990;97:950-1.
16. Dubecq F, Dufour PH, Vinatier D, Tibault D, Lefebvre C, Tordjeman N et al: Monoamniotic twin pregnancies. Review of the literarure, and a case report with vaginal delivery. Eur J Obstet Gynecol Reprod Biol 1996;66:183-6.
17. Ashis KS, Langford K, Elliot C, Su LL, Maxwell DJ: Monoamniotic twins: What should be the optimal antenatal management? Twin Res 2003;4:270-4.
18. Su LL: Monoamniotic twins: diagnosis and management. Acta Obstet Gynecol Scand 2002; 81: 995-1000.