

Spontaneous septostomy of monochorionic dichorionic twin pregnancy: a case series

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We present four cases of confirmed spontaneous septostomy in monochorionic diamniotic twin pregnancy and one false positive case in a regional hospital in Hong Kong between 2011 and 2017. Three of the cases of spontaneous septostomy were detected antenatally.

Keywords: Amnion; Chorion; Pregnancy, twin; Umbilical cord

Introduction

Multiple pregnancy is high risk. With the increasing use of artificial reproductive techniques, the number of twin pregnancies is increasing. Ultrasound assessment of chorionicity and amnionicity starting at first trimester is important because different types of twin pregnancies have different risk management. We present five cases of spontaneous septostomy in women with monochorionic diamniotic (MCDA) twin pregnancy. Early detection is important in reducing morbidity and mortality of both twins.

Case presentation

Case 1

In 2011, a 28-year-old, parity 0 woman with spontaneous MCDA twin pregnancy was followed up in our unit. She had impaired glucose tolerance and developed pre-eclampsia since 30 weeks of gestations. She was admitted to our hospital for close monitoring of the blood pressure and for fetal monitoring using ultrasonography and cardiotocography. At 31 weeks of gestation, ultrasonography showed intrauterine growth retardation of the left twin, while the right twin (leading twin) had normal growth. At 32 weeks of gestation, there was swabbing of the twin positions and disappearance of a part of the inter-twin membrane. Steroid prophylaxis was given to prevent preterm delivery. At 33 week of gestation, lower segment caesarean section was performed in view of pre-eclampsia, selective intrauterine growth retardation, and possible spontaneous septostomy. Intra-operatively, twin 2 was delivered after twin 1 without the need of membrane rupture (Figure 1). Twin 1 had a birth weight of 1.89 kg and an Apgar score of 8(1)10(5), whereas twin 2 had a birth weight of 1.31 kg and an Apgar score of 9(1)10(5).



Figure 1. Case 1: intertwin membrane is absent

Case 2

In 2014, a 28-year-old, parity 0 woman with spontaneous MCDA twin pregnancy was followed up in our unit. Ultrasonography showed the right twin having an umbilical cord with one artery (rather than two) and one vein, while the left twin was normal. The intertwin membrane was observed. Regular ultrasonography showed intrauterine growth restriction of the right twin since 34 weeks of gestation, but the patient refused early delivery. At 36 weeks of gestation, elective lower segment caesarean section was performed on request. Intra-operatively, twin 2 was delivered spontaneously without any membrane ruptured after twin 1 was delivered. On gross examination of the placenta, the intertwin membrane was not seen, and both umbilical cords were close at their insertion sites. Twin 1 was a boy with a birth weight of 2.52 kg and an

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Apgar score of 9(1)10(5), whereas twin 2 was a boy with a birth weight of 2.24 kg and an Apgar score of 9(1)10(5). Spontaneous septostomy was diagnosed after delivery.

Case 3

In July 2016, a 31-year-old, parity 0 woman with spontaneous MCDA twin pregnancy (confirmed at 13 weeks of gestation) was followed up in our unit. At 28 weeks of gestation, spontaneous septostomy was suspected, as no definite inter-twin septum was seen and the only remnant of membrane was seen at the right upper quadrant (Figure 2) and left upper quadrant. There was 'cross-over' of the cord at the centre (Figure 2) but no evidence of cord entanglement. The pregnancy was managed as monochorionic monoamniotic twin with regular ultrasonographic monitoring. In view of the risk of preterm delivery, steroid prophylaxis was given, with an aim of early delivery. At 33 weeks of gestation, elective lower segment caesarean section was performed. Twin 1 (left twin) weighed 1.332 kg and twin 2 (right twin) weighed 1.71 kg. The placenta showed twisting of the cords for two rounds.

Case 4

In December 2016, a 32-year-old, parity 0 woman with spontaneous MCDA twin pregnancy (confirmed at

12 weeks of gestation) were followed up at our unit. At 18 weeks of gestation, ultrasonography showed a single umbilical artery for the right twin and normal two umbilical arteries for the left twin. At 32 weeks of gestation, no membrane was observed between two cord insertions, but the intertwin membrane was seen in other parts. There was a switch of position of the twins, with the fetus having a single umbilical artery at the upper left part. Growth of both fetuses was satisfactory with normal liquor and dopplers. There was no evidence of cord entanglement. In view of suspected spontaneous septostomy, prophylactic steroid was given. At 33 weeks and 6 days of gestation, lower segment caesarean section under spinal anaesthesia was performed. Twin 1 (right twin) was a girl with a birth weight of 1.740 kg and twin 2 (left twin) was a girl with a birth weight of 1.825 kg with a good Apgar score. A defect over the intertwin membrane was noted. Both twins were admitted to the special care baby unit for close monitoring. The left twin underwent ultrasonographic assessment of the kidneys for the single umbilical artery.

Case 5

In 2017, a 29-year-old, parity 1 woman with spontaneous MCDA twin pregnancy (confirmed at 13 weeks of gestation) was followed up at our unit. There was discordance of the thickness of nuchal translucency. The patient declined invasive test and opted for non-invasive prenatal testing in a private hospital, with negative results. Ultrasonography showed polyhydramnios in the left twin but no other evidence of twin-twin transfusion syndrome. At 34 weeks of gestation, the only remnant of the intertwin membrane was seen at the upper cavity, with 'normalisation' of the liquor. At 35 weeks of gestation, elective lower segment caesarean section was performed uneventfully. Twin 1 was a boy weighing 2.09 kg and twin 2 was a boy weighing 2.08 kg. As the inter-twin septum was present upon delivery, this case was a false positive.

Discussion

Spontaneous septostomy, or pseudo-amniotic twin in monochorionic diamniotic twin pregnancy has been described, but only one case report of spontaneous septostomy in dichorionic twin was reported¹. The actual incidence is unknown as reported cases are limited. Within 7 years in our hospital, 613 pairs of twin pregnancy were delivered, and 141 (23%) were estimated to be MCDA according to our previous cohort study². Therefore, the incidence of spontaneous septostomy in MCDA twin is estimated to be $4/141=2.8\%$.

Monochorionic twin pregnancies are associated

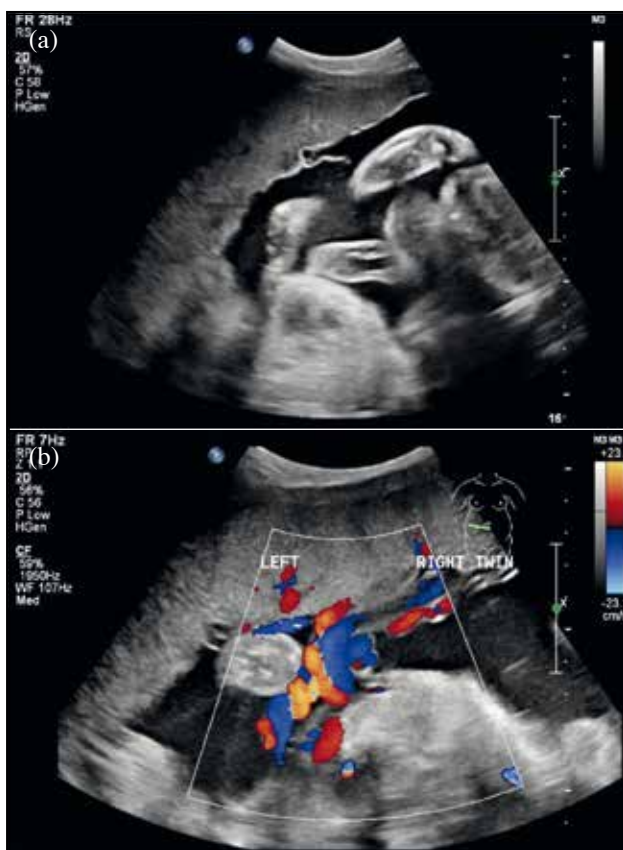


Figure 2. Case 3: ultrasonographs showing (a) remnant of the intertwin membrane and (b) 'cross-over' of the two cords

with higher perinatal risks, compared with dichorionic twins. The vascular anastomosis in monochorionic twins increased the perinatal risks of preterm deliveries, twin-twin transfusion syndrome, selective growth discordancy, and intrauterine death³. The chorionicity should thus best be determined before 14 weeks of gestation⁴. Our patients received ultrasonographic examination at 12 to 13 weeks of gestation. A thin intertwin membrane without lambda sign is indicative of monochorionic diamniotic twin.

According to the Royal College of Obstetricians and Gynaecologists guideline⁴, ‘mapping’ of fetuses, fetal parameters, and liquor volume by measuring single deepest pocket, umbilical dopplers, and middle cerebral artery dopplers should be documented in every scan. In case 4, one of the fetuses had a single umbilical artery, which allowed us to identify the swapping of the position and hence identifying the spontaneous septostomy. In case 2, we missed the clue of a single umbilical artery in one fetus, which is associated with selective intrauterine growth restriction. These cases highlight the importance of detailed mapping of the fetuses in detection of spontaneous septostomy.

Spontaneous septostomy leads to the change of two separated compartments into one single compartment that resembles monochorionic monoamniotic twins⁵. Its main risk is cord entanglement, which occurs in about 60% of spontaneous septostomy and in almost all monochorionic monoamniotic twins⁶. Cord entanglement can lead to fetal demise of both twins⁷. Other risk associated with septostomy is preterm delivery and amniotic band

syndrome⁸. However, there are also cases of septostomy without any complications¹.

Identification of the septostomy depends on clinical suspicion. Features suggestive of spontaneous septostomy include free-floating or folded sheets of amnion⁹ in the gestational sac and body, limbs, or umbilical cord of one twin prolapsed through two chorions into the other sac¹. Careful inspection of the intertwin membrane is recommended through visualising the whole course of the membrane as much as possible at every follow-up examination⁵. The distance between the cord should be documented, as it may be associated with cord entanglement⁹. In our patients, defect of the intertwin membrane was identified when the position of the fetuses switched or when the only remnant of the membrane was seen or when the cords crossed. Nonetheless, case 5 highlighted the pitfall of a false alarm by the absence of part of the inter-twin septum, sudden equalisation of the liquor volume, and suspected cord entanglement. In our series, both the prenatal detection rate and the positive predictive value was 75% (3/4).

Conclusion

Spontaneous septostomy is uncommon but can complicate monochorionic diamniotic twin pregnancy with adverse perinatal outcomes. Prenatal detection with high clinical suspicions and detailed mapping is important during serial antenatal scans. Early detection and management improve outcome.

Declaration

The authors have no conflict of interest to disclose.

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