

# Successful management of discordant gastroschisis in monochorionic diamniotic twin: a case report

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## Summary

**Background:** Monochorionic diamniotic (MCDA) twin pregnancy with gastroschisis carries a poor prognosis. Live birth and well development of both twins are extremely rare. **Case:** The authors report a rare case of discordant gastroschisis in MCDA twin. Both twins were followed up nine months after intrapartum fetal operation, and both are in good health until now. **Conclusion:** This report expands successful management of discordant gastroschisis in MCDA twins. Early diagnosis, intensive prenatal care, and multidisciplinary consultation are recommended in management of discordant gastroschisis in MCDA twin.

**Key words:** Monochorionic diamniotic twin; Gastroschisis; Intrapartum fetal operation.

## Introduction

Gastroschisis is a congenital anterior abdominal wall defect with the uncovered abdominal contents protruding through the defect. The cause is not yet clear. The incidence reported is between four and five per 10,000 live births [1]. Prenatal diagnosis of gastroschisis relies on sonography. Early closure can prevent heat and water loss, infection, and bowel edema. Immediate primary fascial closure should be done when possible. Gastroschisis delivery and immediate repair are important in the operating room. Monochorionic diamniotic (MCDA) twin pregnancy with gastroschisis is rare. This report expands successful management of discordant gastroschisis in MCDA twins.

## Case Report

A 24-year-old woman (gravida 1, para 0) conceived MCDA twins naturally. At 15 weeks of gestation, twin B was diagnosed with anterior abdominal wall defects with  $2.0 \times 2.4 \times 2.0$  cm bowel herniated by ultrasonography while twin A was normal. Second trimester Down's syndrome screening for both twins was normal. The maternal history was unremarkable for smoking, drug or alcohol abuse. No significant family or obstetric history was elicited. Close follow-up ultrasound evaluation every two weeks was maintained with multidisciplinary consultation. At 20 weeks ultrasound, evaluation still showed anterior abdominal wall defects of twin B other than twin A (Figure 1). At 32 weeks, ultrasound showed a  $7.1 \times 4.3 \times 5.0$  cm bowel-like echo floating in the amniotic cavity, while at 35 weeks it increased to  $8.9 \times 5.6 \times 6.5$  cm. Electronic fetal heart rate monitoring performed biweekly from 32 weeks gestation. Twins were delivered by emergency cesarean section at 35 weeks and five days of gestation due to suspected fetal distress of the twin with gastroschisis. They were female, twin A weighted 1,950 g with Apgar scores of 10 at one minute and 10 at five minutes; twin B (Figure 2) weighted 1,890 g who had a three-cm abdominal wall defect to the

right of the umbilicus with the uncovered abdominal contents (most of the small intestine, part of the colon and stomach, ileocecal, and appendix) protruding through the defect. Twin B was given endotracheal intubation, nasogastric tube, placement of peripheral intravenous line for anesthetic and paralysis, and immediate surgical repair in the operation room. Temperature, electrocardiogram, pulse, blood pressure, and oxygen saturation were continuously monitored. After intrapartum fetal operation, twin B was transferred to neonatal intensive-care unit (NICU). She stayed in NICU for three weeks before discharged, and the abdominal wall healed well at 16 days (Figure 3). Karyotype test of both newborns' cord blood was normal (46XX). Monochorionic diamniotic placenta was identified with pathological examination. Both twins were followed up by nine months and are in good health until now.

## Discussion

The etiology of gastroschisis remains elusive. Discordant gastroschisis in MCDA twin pairs is extremely rare. Reports from the literature only showed one pair of female and two pairs of male discordant gastroschisis monozygous twins [2, 3], but the placenta was not available for examination, and the management and follow-up were unclear. As monozygotic twins are essentially genetically identical (with the exception of post-zygotic somatic mutations), this case may suggest environmental factors that play a greater role than genetic factors in the development of gastroschisis as reported before [2].

Progressive evolution of the treatment modalities has led to increased survival in patients with gastroschisis. This is due to improvement in the prenatal diagnosis and post-operative treatment for respiratory compromise, treatment of sepsis, and the use of parenteral nutrition until enteral feeds. This case also showed that immediate surgical repair after delivery is very important.

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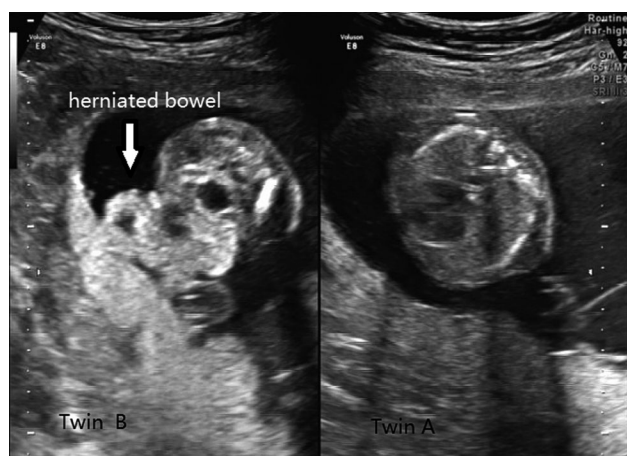


Figure 1. — Twenty weeks' ultrasound shows anterior abdominal wall defects of twin B other than twin A.



Figure 2. — Twin B weighted 1,890 g who had a three-cm abdominal wall defect to the right of the umbilicus with the uncovered abdominal contents.



Figure 3. — The abdominal wall of twin B healed well at 16 days after intrapartum fetal operation.

The mean gestational age at spontaneous delivery in gastroschisis is approximately 36 to 37 weeks [4]. The risk of in utero fetal death is higher in fetuses with gastroschisis. The recommended mode and timing of delivery is still debated, because labor may be deleterious to bowel loops (by compression and twisting) and ruptured membranes may contribute to neonatal infectious complications. A major controversy in the perinatal management of these conditions is whether cesarean delivery leads to an improved neonatal outcome. However, most authors have found no significant benefit to cesarean section. A systematic review of 27 peer-reviewed observational studies similarly found insufficient evidence to support induction of labor in gastroschisis. Additionally, there was no significant relationship between mode of delivery and time until enteral feeding or length of hospital stay [5]. A randomized controlled trial (RCT) also showed the same results [6]. A retrospective study showed that immediate vs traditional surgery resulted in neonates that had less edematous bowel with less fibrous peel, were more likely to be closed primarily, spent less time on a ventilator, seemed to be fed sooner, and were discharged home earlier [7].

Early diagnosis, intensive prenatal care, and multidisciplinary consultation are recommended in management of this case. The present team included obstetricians, sonographer pediatricians, anesthesiologists, pediatric surgeons, and nurses. As survival rates following live birth of an infant with gastroschisis continue to improve, better evaluation would seem to be required, both to long-term nutritional and neuro-developmental outcomes.

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