Ileal blowout due to ileal atresia in a donor with twin-to-twin transfusion after fetoscopic laser surgery

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Twin-to-twin transfusion syndrome (TTTS) occurs in 10% of monochorionic twin pregnancies, and can lead to a variety of complications due to imbalanced blood flow across placental vascular communications. In this report, we describe a case of intestinal injury in TTTS after fetoscopic laser ablation of the communicating vessels. Intestinal ischemic diseases have been reported in the recipient fetus after fetoscopic laser treatment in TTTS. In this report, we describe a unique case of ileal atresia in the donor twin. *Ann Pediatr Surg* 14:42–43 © 2018 Annals of Pediatric Surgery.

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Background

Twin-to-twin transfusion syndrome (TTTS) is a severe complication in monochorionic twin pregnancies. TTTS is caused by an imbalanced blood flow across transplacental vascular anastomoses, resulting in a situation with a 'donor' fetus and a 'recipient' fetus, with the donor having a hypovolemic condition and the recipient having a hypervolemic condition. Four types of vascular anastomoses have been described: arteriovenous (AV), venoarterial (VA), arterioarterial (AA), and venovenous (VV) anastomoses. Vascular anastomoses are seen in almost all monochorionic twins, but only 10% actually develop TTTS [1].

Diagnosis of TTTS is based on the prenatal detection on ultrasound of a monochorionic placenta with twin oligohydramnios or polyhydramnios sequences.

Other signs may include fetal Doppler flow abnormalities. Signs to look for are reversed end-diastolic flow velocity in the umbilical artery, reversed flow in the venous duct, or pulsatile flow in the umbilical vein. These are indications for more advanced stages of TTTS.

Untreated TTTS has high fetal mortality estimated at approximately 85%. Several interventions have been developed to reduce this rate, including laser coagulation, serial amniotic reduction, and selective umbilical cord coagulation. Laser coagulation is the preferred treatment for TTTS nowadays. During this fetoscopic procedure, the anastomoses between donor and recipient are inspected, and then sealed off by means of intrauterine laser coagulation.

The fetal survival rate after laser coagulation is 75%, with a 54% probability of both fetuses surviving, a 27% probability of one fetus surviving, and a 19% probability of none of the fetuses surviving. These figures are based on the majority of TTTS laser surgeries performed worldwide [2]. Notwithstanding improved fetal survival rates after laser surgery in TTTS, however, this treatment is also associated with other potential complications. The incidence of neurological disorders in surviving fetuses is 4.5–11% [3]. Elevated risk of intestinal complications has been reported, including necrotizing enterocolitis and intestinal atresia.

This case report describes the occurrence of intestinal atresia as a complication after fetoscopic laser ablation of transplacental vessels to treat TTTS.

Case

A monochorionic twin pregnancy with TTTS (Quintero stage 2) was successfully treated with fetoscopic laser surgery at 18 weeks of gestation. Both twins survived the procedure; however, severe intrauterine growth retardation complicated the growth and development of the ex-donor twin. At 30 weeks and 5 days of gestation, an emergency cesarean section was carried out because of an abnormal Doppler ultrasound and decelerations on cardiotocography in the ex-donor twin. Apgar scores of the ex-donor twin were 8, 9, and 10 after 1, 5, and 10 min, respectively. Birth weight was 625 g (< 2 SD). Because of prematurity and severe growth restriction, he was admitted to the Neonatal Intensive Care Unit. Placenta injection with color dye was performed after delivery, and no residual anastomoses were detected. On day 3 of life, increasing abdominal distension and free air on abdominal radiography were detected, and he was transferred to our pediatric surgical center.

A laparotomy was performed. An atretic segment of the mid-ileum was found to be complicated by a blowout proximal to the obstruction. Resection of 6 cm of the atretic ileal segment including the perforated part and a split ileostomy were performed.

The postoperative course was complicated by stoma retraction requiring revision. The patient had an uncomplicated recovery.

Discussion

In this case, ileal atresia developed after fetoscopic laser coagulation for TTTS. The link between TTTS laser coagulation and intestinal atresia has been described before. Five case reports have described intestinal atresia

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occurring in surviving fetuses after intrauterine laser surgery [4–7]. In five of these cases, intestinal atresia was found in the 'recipient' fetus, after the donor fetus had died in utero. Only one case report has described the postnatal finding of multiple jejunal atresias in a 'donor' fetus after intrauterine laser coagulation at 16 weeks of pregnancy. The atretic segments were removed laparoscopically, and an end-to-end anastomosis was made. The postoperative course in this patient was uneventful.

This case report is, to our knowledge, the first to describe a donor twin with postnatal ileal atresia after intrauterine laser coagulation at 18 weeks of pregnancy, complicated by postnatal bowel perforation. In the postoperative course, a relaparotomy had to be performed for revision of the ileostomy.

The occurrence of intestinal atresia after laser surgery in TTTS has been described sporadically. A theory of intestinal atresia is an intrauterine vascular disruption inducing necrosis of the fetal bowel. As the fetal bowel is sterile, this necrotic segment is resorbed, leaving blind proximal and distal bowel ends. It may be that fetoscopic laser coagulation changes fetal blood flow causing intestinal ischemia by obstruction, which would lead to sterile necrosis of, in this case, the ileum. Another theory is that mesenteric hypoperfusion may be secondary to emboli released into the fetal circulation. These emboli may obstruct the mesenteric vessels causing ischemia and necrosis. However, in the present report, placenta injection with color dye showed no evidence of residual anastomoses, eliminating the likelihood of emboli as an etiologic mechanism. Finally, the occurrence of ileal atresia may also be secondary to the hypovolemia in the donor as part of TTTS before the laser coagulation therapy.

In conclusion, this is the first report of ileal atresia in a donor fetus following laser coagulation for TTTS, which may have been due to mesenteric ischemia. This case report illustrates that close monitoring of neonates is warranted in view of this serious complication.

Conflicts of interest

There are no conflicts of interest.

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