Fatal Umbilical Cord Strangulation in the Remaining Co-Twin after Selective Foetocide with Radiofrequency Ablation for Twin-Twin Transfusion Syndrome

Lisanne S.A. Tollenaar^a Patricia C.J. Knijnenburg^b Janina L. Wolf^c
Femke Slaghekke^a Johanna M. Middeldorp^a Monique C. Haak^a
Frans J.C. Klumper^a Dick Oepkes^a Enrico Lopriore^b

^aDivision of Fetal Medicine, Department of Obstetrics, Leiden University Medical Center, Leiden, The Netherlands;

^bDivision of Neonatology, Department of Pediatrics, Leiden University Medical Center, Leiden, The Netherlands;

^cDepartment of Pathology, Leiden University Medical Center, Leiden, The Netherlands

Established Facts

- Monochorionic twin pregnancies can be complicated by twin-twin transfusion syndrome.
- Selective foetocide with radiofrequency ablation might be a suitable therapy option in specific cases of twin-twin transfusion syndrome.

Novel Insights

- Iatrogenic monoamnioticity can be a serious complication after radiofrequency ablation for the twin-twin transfusion syndrome and can lead to cord entanglement resulting in fatal outcome in the co-twin.
- To prevent fatal cord accidents after radiofrequency ablation in twin-twin transfusion syndrome, intensified ultrasound check-ups focussing on the intactness of the inter-twin membrane may be required.

Keywords

Foetal therapy · Foetocide · Monochorionic twins · Monoamniotic twins · Selective termination · Twin oligohydramnion-polyhydramnion sequence · Twin-twin transfusion syndrome · Umbilical cord · Intrauterine death

Abstract

We report a case of a monochorionic diamniotic twin diagnosed with twin-twin transfusion syndrome (TTTS; stage 3) with co-existing severe cerebral damage in the donor twin at 18 + 4 weeks' gestation. After counselling, the parents opted for selective foetocide of the donor twin. For the procedure, radiofrequency ablation (RFA) was used. Serial ultra-

sound examinations at 20 + 1 and 21 + 1 weeks' gestation showed good recovery of the ex-recipient, after which the patient was sent back to the referring hospital. At 29 + 5 weeks' gestation, an unexpected foetal death was diagnosed. On macroscopic placental examination, (iatrogenic) monoamnionicity was detected. In addition, the umbilical cord of the recipient was found to be constricted by the macerated umbilical cord of the ex-donor. This case demonstrates that iatrogenic monoamnionicity can be a serious complication of RFA in monochorionic twins complicated by TTTS, with a subsequent risk for cord entanglement leading to a fatal outcome for the remaining co-twin. Although the actual incidence of iatrogenic monoamnionicity after RFA remains unknown, increased attention to the intactness of the inter-twin membrane even weeks after the RFA may be required.

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Case Report

A 37-year-old woman (G2P1) was referred to our institution at 18 + 4 week's gestation with a monochorionic diamniotic twin pregnancy, complicated with twin-twin transfusion syndrome (TTTS). Ultrasound examination revealed TTTS stage 3, with a stuck twin and a deepest vertical pocket (DVP) of 13 cm in the recipient, and severe growth restriction (estimated foetal weight <2 SD below the mean for gestational age) of the donor with intermittent absent or reversed flows in the umbilical artery. In addition, the ultrasound examination of the brain showed that the donor twin had severe bilateral ventricular dilatation. After counselling, the parents opted for selective foetocide using radiofrequency ablation (RFA) of the donor twin.

For the procedure, a 17-gauge Starburst RFA needle (RITA Medical Systems Inc. Mountain View, CA, USA) was used, applying a maximum power setting of 40 W. Under continuous ultrasound guidance, the needle was directed through the anterior placenta and directly into the abdomen of the donor. A total of 2 thermal cycles were required to cease the hemodynamic circulation in the donor twin. After the RFA, amniotic fluid was drained from to sac of the recipient, resulting in a DVP of 7 cm at the procedure. In total, the procedure took 25 min to complete and there were no maternal or foetal complications in the following 24 h. The next day, the patient was discharged. An ultrasound examination at 20 + 1 showed good recovery of the ex-recipient. At 21 + 1 weeks, a DVP of 2 cm in the amniotic sac of the donor was seen, but there were no signs of complete rupture of the inter-twin membrane. Subsequently, the patient was sent back to the referring hospital. At 29 + 5 weeks' gestation, an unexpected intrauterine foetal death was diagnosed. After delivery, the placenta was shipped to our hospital for further investigation.

Macroscopic examination of the placenta revealed a normal appearance of a recently viable placental share with a central cord insertion for the ex-recipient twin and a macerated placental share with a velamentous insertion of the thin umbilical cord of the ex-donor twin (Fig. 1). The umbilical cord of the ex-recipient was

Fig. 1. Macroscopic examination of the foetal side of the placenta showing the maceration of the placental share of the ex-donor and an intact placental share with central cord insertion for the ex-recipient. At approximately 20 cm from the insertion of the umbilical cord of the ex-recipient, cord constriction is seen. This was caused by the entanglement of the macerated umbilical cord of the ex-donor.

dark red and appeared congested. A circular constriction of the umbilical cord of the ex-recipient, caused by entanglement of the macerated umbilical cord of the ex-donor twin (Fig. 2a), was observed. Figure 2b depicts the appearance of the umbilical cord of the ex-recipient after removing the macerated umbilical cord of the ex-donor.

Microscopic examination of the constricted area of the umbilical cord of the ex-recipient revealed an intact architecture of the vessels walls, lying in Whartons jelly (Fig. 3). In the lumen of both arteries, a thrombus (consisting of fibrin, platelets and lymphocytes) was found.

Discussion

We report a case of cord entanglement resulting in foetal demise in the remaining co-twin after selective foetocide with RFA for TTTS in a monochorionic diamniotic twin. This is the first report showing that RFA might result in iatrogenic monoamnionicity, emphasizing the need for increased awareness of the importance of intensified monitoring throughout the whole course of pregnancy after selective foetocide for monochorionic twins.

As on date, foetoscopic laser coagulation of the vascular anastomoses is the optimal treatment for TTTS. However, selective foetocide might be an alternative therapy option in cases in which laser is technically not feasible or

Fig. 2. **a** The umbilical cord of the ex-recipient showing a tight knot at the base of constriction. **b** The umbilical cord of the ex-recipient after removing the macerated umbilical cord of the ex-donor, showing a thin and pale constricted area.

Fig. 3. Microscopic examination of the constricted area of the umbilical cord of the recipient twin. In both arteries a thrombus is found.

in cases with severe cerebral injury, congenital abnormalities or severe growth restriction in either the donor or recipient twin. Technical options for selective foeticide include bipolar cord occlusion, foetoscopic laser cord coagulation or RFA. RFA has potential benefits over other modalities because the diameter of the shaft is relatively small, making the procedure minimally invasive and applicable in early gestations as well. Consequently, RFA is characterized by high survival rates for the co-twin and low rates of preterm premature rupture of the membranes [1–3]. Aside from the incidence of preterm premature rupture of the membranes, no studies with respect to other procedure-related complications after RFA have been published to date.

As far as our knowledge is concerned, this is the first report that demonstrates that treatment with RFA can re-

sult in iatrogenic monoamnionicity and umbilical cord entanglement with concurrent risk for foetal demise in the remaining co-twin. To date, there are no reports on iatrogenic monoamnionicity after treatment with RFA, but the incidence after treatment with foetoscopic laser coagulation is estimated to be around 20% [4]. Management after the suspicion of iatrogenic monoamnionicity consists of intensified monitoring with ultrasound; hospitalization after viability and a preterm elective Caesarean section at approximately 32 weeks' gestation are considered to prevent cord accidents [5]. Compared to foetoscopic laser surgery, we expect the incidence of iatrogenic monoamnionicity after RFA to be much lower. The rationale behind this is that both procedures are aimed at causing thermal damage on different locations. While RFA is applied only in the internal body of the foe-

tus, laser coagulation is performed at the vascular equator of the placenta, which is often close to, or even in touch with the inter-twin membrane, thereby making it more prone to damage.

Importantly, this case report may warrant increased awareness regarding the frequency and focus of follow-up ultrasound examinations after RFA. In the absence of any maternal or foetal complications at 1 and 2 weeks after the procedure, the patient is often discharged to the referring centre, where the obstetrical care is left to the discretion of the referring gynaecologist. In most cases, further follow-up ultrasound appointments will be in accordance with the care for uncomplicated singleton pregnancies. However, this report illustrates that foetal demise due to cord entanglement after iatrogenic monoamniocity can even occur weeks or months after the RFA is performed. In our patient, the cause of iatrogenic monoamniocity is not entirely clear. Possibly, a prong of the RFA needle was outside the donors' body, thereby causing thermal damage to the inter-twin membrane. An alternative explanation is that when the RFA needle went through the placenta and into the abdomen of the donor, it might have accidentally perforated the inter-twin membrane that was tightly collapsed against the donors' body. Regardless of the cause, iatrogenic monoamniocity was not clearly detected on ultrasound 1 and 2 weeks after the procedure, suggesting that it might have developed or become more distinct in the following weeks. It is therefore advisable to pay increased attention to the intactness of the inter-twin membrane throughout the whole course of pregnancy. In case of iatrogenic monoamniocity on ultrasound, specific attention should be paid to the entanglement of the cords. However, this might be challenging after selective reduction, since the umbilical cord of the

demised foetus will not show Doppler flows anymore. We therefore recommend that echogenic remains of the umbilical cord of the demised foetus be looked at and that increased attention be paid to calibre alterations or flow abnormalities in the umbilical cord of the remaining co-twin during further follow-up examinations. In case of proven iatrogenic monoamniocity, management should consist of intensified foetal monitoring (with hospitalization if viable) and caesarean section at 32 weeks gestation.

In conclusion, this case demonstrates that iatrogenic monoamniocity can be a serious complication of RFA in monochorionic twins complicated by TTTS, with a subsequent risk for cord entanglement leading to a fatal outcome for the remaining co-twin. Although the actual incidence of iatrogenic monoamniocity after RFA remains unknown, increased attention to the intactness of the inter-twin membrane even weeks after the RFA may be required. More studies regarding the incidence and impact of this complication are needed in order to propose adequate ultrasound management guidelines after RFA for complicated monochorionic twins.

Statement of Ethics

The subject involved in this study gave her informed consent. The study protocol has been approved by the institute's committee on human research. No animal experiments were performed in this study.

Disclosure Statement

All authors declare that they have no conflicts of interest to disclose. There was no sponsorship or funding arranged for this case report.

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