

Fetoscopic interventions in complicated monochorionic twin pregnancies

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Selective feticide in monoamniotic twin pregnancies by umbilical cord occlusion and transection

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Abstract

Objective: In monoamniotic twin pregnancies discordant for fetal anomaly, parents may opt for selective feticide. However, the normal co-twin remains at risk of sudden demise from cord entanglement. We report on three cases of successful selective feticide by cord occlusion combined with cord transection.

Methods: We describe technical details and outcome of three monoamniotic twin pregnancies, discordant for fetal anomaly (two cases of anencephaly and one case of congenital heart block), in which cord occlusion was followed by transection of the cord, using contact laser.

Results: The fetoscopic cord occlusion and transection, using laser, was successfully performed at 15, 16 and 19 weeks' gestation, respectively. In one case, amniotic fluid leakage occurred after fetoscopy. The surviving co-twins were born healthy, two out three vaginally, at 36, 38 and 36 weeks' gestation, respectively.

Conclusion: In monoamniotic twins, selective feticide using laser occlusion and transection of the umbilical cord is technically feasible and can lead to near-term vaginal birth of healthy co-twins.

Introduction

Monoamniotic twins occur in approximately 1% of monozygotic twin pregnancies. Monoamniotic twins are at increased risk of fetal anomalies, twin-to-twin transfusion syndrome (TTTS), fetal death and premature delivery. Perinatal mortality rates of 10-70% have been reported. 1;2 Entanglement and knots of the umbilical cords are the major cause of fetal death in monoamniotic twins.³ Cord entanglement is present in over 70% of monoamniotic twins, and is already seen in the first trimester.^{3;4} It is speculated that the occurrence and the fastening of knots occurs more easily when there is more amniotic fluid in relation to the fetal body masses. Several methods have been described that aim at lowering perinatal mortality from cord entanglement: reduction of amniotic fluid by amniodrainage, or administration of sulindac. 5;6 Also, intensive inpatient fetal monitoring with elective preterm delivery has been described.^{6;7} Furthermore, congenital anomalies are found in 15-20% of monozygotic twin pregnancies, and may or may not be concordant.8 In case of congenital anomalies in one fetus of a monochorionic twin pair, the otherwise normal co-twin is at risk for in utero demise due to acute perimortem blood loss into the dying cotwin.9 In addition, obstetric complications, due to the anomaly, for example polyhydramnios in pregnancies complicated by anencephaly, can present a threat to the normal co-twin. In twin pregnancies with one anomalous fetus, selective feticide can be indicated for two reasons, either to prevent the birth of a severely handicapped child, or to protect the normal co-twin against the described complications. Both in monochorionic and monoamniotic twin pregnancies, selective feticide should not be performed using fetal drug injection because of the presence of vascular anastomoses on the placental surface and the risk of acute hypovolaemia in the normal co-twin during demise of the anomalous twin. Therefore, umbilical cord occlusion is the method of choice. However, after cord occlusion in monoamniotic twins, the incidence of cord accidents from entanglement and strangulation is still a serious threat, and reported to be as high as 36%. 10 Therefore, transection of the occluded cord may be life saving for the normal co-twin.

We report our experience with selective feticide using laser energy for occlusion and transection of the umbilical cord, in three cases of monoamniotic twins discordant for congenital anomalies.

Case 1

A 32-year-old woman was referred to our centre, which is the national referral centre for invasive fetal treatment in the Netherlands, at 14+1 weeks' gestation, with a monoamniotic twin pregnancy complicated by one fetus with anencephaly. On ultrasound examination, the presence of anencephaly in one fetus was confirmed, the other fetus showed no signs of structural anomalies. Colour Doppler showed signs of cord entanglement. During counselling, consideration was given to the risk for the normal co-twin on the basis of cord entanglement, the risk of polyhydramnios with anencephaly and other known risks of monochorionic monoamniotic twinning. The couple opted for selective feticide of the anencephalic fetus. The procedure was performed at 15+4 weeks' gestation under general anaesthesia. A prophylactic dose of tocolytics (indomethacin) and antibiotics (amoxycillin/clavulanate) was given. An 18-G needle was entered into the amniotic cavity under ultrasound guidance, followed by introduction of a 0.085-inch soft J-tipped guide wire (Cook®) through the needle using the Seldinger technique. A 10 F teflon cannula, loaded with a dilator, was advanced over the guide wire. When the cannula had entered the amniotic cavity, the quide wire and dilator were removed and the shaft loaded with the fetoscope (2 mm diameter) and laser fibre (400µm) was inserted. This procedure was also sonographically guided. Amniotic fluid was blood stained, as is often the case in the presence of an anencephalic fetus. Amnioexchange with warm Ringers lactate was performed until a clear fluid compartment was reached. The umbilical cord of the abnormal fetus was identified and cord knots were present (Figure 1). Under fetoscopic sight and ultrasound guidance, the umbilical cord of the anencephalic fetus was coagulated by laser energy (Nd:YAG laser, Dornier Medizin Technik, Germering, Germany), close to the abdominal insertion of the umbilical cord, over a distance of 15 mm, using short bursts of 5-10

seconds, with a maximum power of 50 Watts. We aimed at whitening and shrinking the cord over a distance of 15 mm, as can be observed by direct visualisation, using the laser fibre (diameter 0.4 mm) as a reference, and also by sonographic measurement. Vascular occlusion was confirmed with colour Doppler ultrasound. Hereafter, the occluded cord was transected with the same laser fibre, set in the cutting mode (50 Watts). The procedure had an uneventful course. During the procedure, the normal fetus showed no changes in heart rate. Several weeks after the procedure, some leakage of amniotic fluid occurred. This leakage continued during pregnancy and resulted in oligohydramnios. Despite fluid leakage, the pregnancy continued uneventfully until the near term period. At 36+3 weeks' gestation the woman went into labour and gave birth vaginally to a healthy boy weighing 3160 gram and a 5-min Apgar score of 9. The neonatal course was uneventful.

Case 2

A 32-year-old woman, known with insulin-dependent diabetes mellitus, was referred to our hospital at 16+3 weeks' gestation, with a monoamniotic twin pregnancy complicated by one fetus with anencephaly. On ultrasound examination the diagnosis of anencephaly was confirmed. On colour Doppler, signs of cord entanglement were present. The other fetus showed no structural anomalies. After counselling, the couple opted for selective feticide of the fetus with anencephaly. The procedure was performed at 16+4 weeks' gestation under general anaesthesia. A prophylactic dose of tocolytics (indomethacin) and antibiotics (amoxycillin/clavulanate) were given. The placenta covered the anterior uterine wall completely and, although very unusual even in anterior placenta, no placenta-free area could be identified for safe fetoscopic entry. Therefore, a midline laparotomy was performed. The uterus was gently exteriorised and an embryoscope (1 mm diameter) and laser fibre (400µm) were introduced through the posterior uterine wall into the amniotic cavity under ultrasound guidance. Amniotic fluid was clear. The umbilical cord of the abnormal fetus was identified. The presence of multiple complex cord knots was confirmed. Under embryoscopic sight

and ultrasound guidance, the umbilical cord of the anencephalic fetus was coagulated by laser energy, close to the abdominal insertion of the umbilical cord, over a distance of 15 mm, using short bursts of 5-10 seconds, with a maximum power of 30 Watts. Occlusion was confirmed with colour Doppler ultrasound. Hereafter, the occluded cord was transected with the laser fibre (400µm), set in the cutting mode (30 Watts). The procedure had an uneventful course. During the procedure, the normal fetus showed no changes in heart rate. Postoperative course was uneventful. At 37 weeks' gestation the patient developed hypertension and episodes of hypoglycaemia. She was admitted to the referring hospital and ultrasound investigation showed macrosomia of the fetus. Labour was induced at 38 weeks' gestation and a caesarean section was performed because of failure to progress. A healthy girl weighing 4044 gram was born.

Case 3

A 33-year-old woman was referred to our hospital at 18+0 weeks' gestation with a monoamniotic twin pregnancy complicated by one fetus with severe ventricular bradycardia of 47 beats per minute, due to a congenital heart block. Maternal serum tested negative for anti-SSA and anti-SSB antibodies. Diagnosis of congenital heart block was confirmed by fetal echocardiography. Cord entanglement with a complex knot was visible on ultrasound. The other fetus showed no anomalies. After extensive counselling, considering the risk of cardiac failure and death in the fetus with bradycardia and the subsequent risk for the other fetus, the couple opted for selective feticide of the fetus with the heart block. The procedure was performed at 18+6 weeks' gestation under general anesthesia. The placenta was anterior, with a placenta-free window allowing safe percutaneous access. The same procedure as described in case 1 was followed. The amniotic fluid was clear. Once the umbilical cord insertion at the abdominal site of the fetus with the heart block was identified, it became clear that the entangled cords were too close to the abdominal wall of the fetus, making laser coagulation at this site unsafe. At the placental insertion site, the umbilical cords, with insertion close to each other, were free

Chapter 7

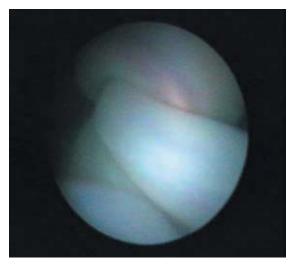


Figure 1 Fetoscopic view of cord entanglement in a monoamniotic twin pregnancy.



Figure 2 Fetoscopic view of the occluded umbilical cord being transected using Nd:YAG laser energy through a 400 µm fibre in the cutting mode.



Figure 3 Fetoscopic view of the umbilical cord at the placental insertion after transection.

of knots over a short distance of circa 5 cm. Using Doppler, the umbilical cord of the fetus with bradycardia could easily be identified, and was subsequently coagulated with laser energy and transected as described above (*Figure 2*). Figure 3 shows the cord insertion at the end of the operation. During the procedure, the normal fetus showed no changes in heart rate. The pregnancy proceeded uneventfully. At 35+6 weeks' gestation, the patient was admitted to the referring hospital with premature rupture of membranes and contractions. A healthy girl was born vaginally, with a birth weight of 3100 gram and a 5-min Apgar score of 9.

Discussion

Selective termination in twin pregnancies is to be considered as a management option in the presence of an anomaly in one fetus, severe enough either to lead to the birth of a severely handicapped child, or when it presents a threat to the normal co-twin e.g. by the development of polyhydramnios with risk of preterm birth. In monochorionic twin pregnancies, the placental anastomoses present a serious threat when one of the twins is critically ill or dying, because of acute perimortem TTTS.⁹ In case of a monoamniotic twin pregnancy, the risk of cord

entanglement and cord knots adds another major risk, even after the death of one fetus.¹⁰

Selective feticide by cord occlusion has been widely accepted as a treatment option for monochorionic diamniotic twin pregnancies in case of discordant congenital anomaly, twin reversed arterial perfusion (TRAP) or severe TTTS with a critically ill fetus.¹¹ In monoamniotic and pseudo monoamniotic twins, additional transection of the umbilical cord has been proposed, to avoid the complications of cord entanglement later in pregnancy.^{3;12-16}

Several techniques have been described to interrupt the blood flow in the umbilical cord. Percutaneous ultrasound-guided injection of thrombogenic material in the umbilical cord of an acardiac twin has been performed in various ways, e.g. trombogenic coils, fibrin superglue, or alcohol embedded suture. 17-22 Unfortunately, this procedure has been associated with death of both twins in approximately half of the cases. 18;19 The first described minimally invasive procedure using two 2-3 mm ports was umbilical cord ligation with a 3-0 Vicryl suture. 23 Endoscopically guided photocoagulation of the cord using Nd:YAG laser through a single port was first described by Hecher *et al.* 24 Ultrasound guided bipolar electro coagulation using a bipolar forceps through a 3 mm port was the next technique used for umbilical cord occlusion. 14 In TRAP sequence, the treatment modality of choice is the so-called intrafetal approach. 25

For transection of the occluded cord, some have proposed the use of scissors, others suggested using a harmonic scalpel via a single 5-mm port technique, or coaxial bipolar electrode (Versapoint®) via a two port technique (diameters 3.8-mm and 2.2mm). 12;13;15 In our three cases, a single port technique with a shaft containing a 2 mm fetoscope or 1 mm embryoscope with a 400 µm laser fibre was used. Under direct and continuous vision, coagulation was performed in the non-contact mode and the cord was transected in the contact mode. Laser occlusion of the umbilical cord has been described to be successful up to 20 weeks' gestation. 26 After 20 weeks, the thickness of the cord and the quantity of Wharton's jelly content limit the effects of laser, and in these cases bipolar coagulation (followed by laser transection) is probably more successful. 27 In monoamniotic twins with cord entanglement

and knots, the optimal identification site of the cord of the affected twin is at the abdominal insertion. Thus, the risk of coagulating the cord of the normal co-twin is small. Another advantage of coagulating the cord close to its abdominal insertion is the lack of mobility of the cord when touching it for the cutting session. In our three cases, the cord was easily transected and no technical complications occurred. In one case, ongoing amniotic fluid leakage occurred, a well-known complication of invasive fetal procedures. Because the risks associated with cord entanglement were ruled out by transecting the cord, near term vaginal birth was made possible.

Conclusion

Laser coagulation and transection of the umbilical cord under direct vision is a minimally invasive single-port-technique that can be safely used for selective feticide in monoamniotic twin pregnancies with discordant congenital anomaly.

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