Severe polyhydramnios in twin reversed arterial perfusion sequence: successful management with intrafetal alcohol ablation of acardiac twin and amniodrainage

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ABSTRACT
We present two cases of twin reversed arterial perfusion (TRAP) sequence complicated by severe polyhydramnios during the second trimester. Both cases were successfully managed at 27 and 26 weeks with ultrasound-guided intrafetal alcohol ablation of the acardiac twin and amniodrainage. The pump twins were delivered at 35 and 37 weeks, respectively, and both had uneventful postnatal courses. These cases suggest that, in the setting of severe polyhydramnios, prenatal intervention in TRAP sequence could certainly improve the otherwise poor prognosis of the pump twin. Targeting the main intra-abdominal vessel of the acardiac twin rather than its umbilical cord seems to be a good alternative in the prenatal treatment of these cases. The option of percutaneous intrafetal alcohol injection is widely available and less invasive, simpler and easier to perform than recently advocated endoscopic techniques.

INTRODUCTION
Fetal acardia is the most severe malformation seen in humans, all organs being invariably affected with varying degrees of severity7. Although in recent years much knowledge has been gained in understanding its natural history and underlying pathophysiology, the primary cause of this developmental anomaly has not yet been fully elucidated. A single artery-to-artery anastomosis allowing reversal of blood from the pump to the acardiac twin, a condition known as the twin reversed arterial perfusion (TRAP) sequence2, is a constant feature of this condition, suggesting primary cardiac agenesis or, more probably, cardiac dysmorphogenesis in the etiology of these cases3.4. In a significant number of cases the size of the acardiac is smaller than the pump twin. In such circumstances, the pump twin usually shows no evidence of cardiac insufficiency and has a favorable perinatal outcome without the need for any antenatal intervention. In others, the growing parasitic mass may divert a considerable amount of blood from the pump twin’s circulation, eventually leading to cardiac failure, polyhydramnios and preterm delivery2,5,6.

We describe two cases of TRAP sequence complicated with severe polyhydramnios in the second trimester, a complication which is almost invariably associated with a dismal perinatal outcome2,5,6. A rescue procedure involving ultrasound-guided intrafetal alcohol ablation of the acardiac twin and amniodrainage was instrumental in improving the otherwise poor outcome in these cases.

CASE REPORTS
Case 1
A 29-year-old primigravida was referred at 27 weeks of gestation because of a twin pregnancy complicated with TRAP sequence and severe polyhydramnios. A monochorionic-diamniotic twin pregnancy was diagnosed at 13 weeks, at which time one twin appeared structurally normal whereas the other was grossly abnormal and had no apparent cardiac activity. Follow-up scans at the referring institution revealed increasing size of the suspected dead twin and increased amniotic fluid volume in the second trimester leading to tense polyhydramnios at 27 weeks. At referral the pump twin showed growth restriction, a distended bladder and an edematous umbilical cord but no structural anomalies. The acardiac-acephalus twin was twice as large as the pump twin and had rudimentary arms and legs, massive subcutaneous edema and large cystic hygromas. Color Doppler ultrasound confirmed reverse blood flow to the acardiac, confirming the diagnosis of TRAP sequence. The poor prognosis of TRAP sequence when severe

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polyhydramnios develops was explained to the parents and available therapeutic options were discussed at length. They gave informed consent for ablation of the acardiac twin with percutaneous intrafetal alcohol injection and amniodrainage.

The ablation procedure was performed as previously described. Briefly, the intra-abdominal segment of the single umbilical artery was identified with color flow imaging (Figure 1) and a 20-gauge spinal needle was passed under continuous ultrasound guidance. Two milliliters of absolute alcohol was injected into the arterial vessel with almost immediate cessation of blood flow to the acardiac. The procedure was completed with amniodrainage of 2000 mL using the technique described by Elliott and colleagues. Subsequent follow-up scans showed progressive shrinkage of the acardiac twin, appropriate growth intervals of the pump twin and normal amniotic fluid volume. However, from 33 weeks onwards the pump twin showed moderate oligohydramnios and increased downstream resistance in the umbilical artery. At 35 weeks an amniocentesis for lung maturity studies revealed an L/S ratio > 2 and meconium-stained amniotic fluid. In view of breech presentation of the pump twin, a Cesarean section was performed delivering a female infant weighing 1925 g and with Apgar scores of 8 and 9 at 1 and 5 min, respectively. The mummified acardiac twin weighed 250 g, measured 24 cm and had a two-vessel umbilical cord 14 cm in length (Figure 2). The pump twin had an uneventful postnatal course and was discharged in good condition. The monochorionic-diamniotic placenta weighed 310 g. Direct artery-to-artery and vein-to-vein anastomoses were noted connecting the two umbilical cords.

Case 2

A 37-year-old primigravida was referred at 26 weeks of gestation because of a twin pregnancy complicated with TRAP sequence and severe polyhydramnios. Routine ultrasound at 18 weeks had revealed a monochorionic-diamniotic twin pregnancy with moderate polyhydramnios. One twin was structurally normal but the other was grossly abnormal and had no cardiac activity, which initially led to the suspicion of a single intrauterine demise of an abnormal twin. The diagnosis of TRAP sequence was confirmed at 24 weeks, when a follow-up scan demonstrated reverse blood flow to, and increasing size of, the suspected dead twin. At referral the pump twin was appropriately grown but exhibited several features of cardiac insufficiency including mild pericardial effusion, myocardial hypertrophy, umbilical vein pulsation, tricuspid regurgitation, a distended bladder, edema of the umbilical cord and tense polyhydramnios. The acardiac twin was much larger than the pump twin and had rudimentary spine and legs, massive subcutaneous edema and large cystic hygromas. After counseling, the parents gave informed consent for ablation of the acardiac with percutaneous intrafetal alcohol injection and amniodrainage.

Occlusion of blood flow was successfully achieved at the second attempt because dislodging of the needle during the first puncture prevented intravascular injection. The procedure was completed with amniodrainage of 3100 mL. Follow-up scans demonstrated absence of blood flow and progressive shrinkage of the acardiac twin but persistence of moderate polyhydramnios and myocardial hypertrophy with mild pericardial effusion in the pump twin. After a short course of indomethacin and digoxin for transplacental treatment, the polyhydramnios resolved and improving cardiac function with adequate growth intervals of the pump twin were demonstrated. At 37 weeks 5 days, an elective Cesarean section was performed delivering a healthy female infant weighing 2930 g and with Apgar scores of 8 and 9 at 1 and 5 min, respectively. The mummified acardiac-acephalus twin weighed 200 g, measured 16 cm and had a two-vessel umbilical cord that was 15 cm in length. The pump twin had an uneventful postnatal course and was discharged in good condition. The monochorionic-diamniotic placenta weighed 500 g and single artery-to-artery and vein-to-vein anastomoses between the cord insertions were demonstrated on the fetal surface of the placenta.

DISCUSSION

The main pathophysiologic event leading to serious complications in TRAP sequence is the vascular steal...
phenomenon. Consequently, all prenatal procedures designed to treat this condition have been focused on the occlusion of the circulation to the acardiac twin. Hysterotomy with selective delivery of the acardiac was initially attempted by several groups8,9,10. However, this approach has been largely abandoned not only because of its invasiveness but also because less invasive techniques are now available. These include ultrasound-guided intrafunicular injection of steel coils, quick-setting gels and soaked-suture material11,12, umbilical cord ligation with the use of endoscopy13–15 or cord grasping devices16 and endoscopic laser coagulation of the umbilical cord17–19. In view of the technical difficulties and substantial risks associated with intrafunicular occlusion, which per se could trigger the death of the pump twin, a novel approach targeting the main intrafetal rather than the cord vessels has been advocated7. This technique has been used to inject absolute alcohol as a vascular sclerosant7 and, recently, to deliver thermocoagulation energy through a wire device20, with technical success in arresting the circulation to the acardiac and survival of the pump twin in all reported cases.

Arias and colleagues recently reviewed 22 cases in which antenatal invasive treatment for TRAP sequence was attempted19. They concluded that the best options were endoscopic laser coagulation of the umbilical cord before 24 weeks and endoscopic-assisted umbilical cord ligation thereafter19. Although we agree that the preliminary experience with intrauterine endoscopic techniques is encouraging, this approach still requires specialized equipment and highly skilled operators, which are only available in a few centers around the world. Our report suggests that good results can also be obtained with a less sophisticated technique such as intrafetal intravascular alcohol injection, even in the presence of severe polyhydramnios and at advanced gestational ages. The main advantages of this technique over endoscopic-guided procedures include shorter intra-operative time, since the procedure is usually completed in a few minutes, and less invasiveness, since it only requires the introduction of a spinal needle. In addition, this technique can be performed by any obstetrician with adequate training in fetal blood sampling, the main challenge being the correct identification of the targeted vessel, which makes color Doppler imaging necessary to perform the procedure. Moreover, since the procedure requires only a minimum of material such as absolute alcohol and an amniocentesis needle, this technique can be easily implemented in any tertiary referral center. According to our experience, intrafetal alcohol ablation could provide an inexpensive, rapid and straightforward solution to a vexing problem that is associated with a high perinatal mortality rate if left untreated. We suggest that the simplicity of this technique makes alcohol ablation one of the treatments of choice for ablation of acardiac twins, especially in the majority of those referral centers where endoscopic techniques are not available.

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ADDENDUM

After the submission of this manuscript, an article advising against the use of intravascular alcohol injection for umbilical vessel occlusion was published (Denbow ML, Overton TG, Duncan KR, Cox PM, Fisk NM. High failure rate of umbilical vessel occlusion by ultrasound-guided injection of absolute alcohol or embrucrilate gel: Prenat Diagn 1999; 19: 527–532). Only one of the 12 procedures was carried out in a pregnancy complicated with the TRAP sequence. In this particular case the abnormal twin had a functioning rudimentary heart and was first given intracardiac potassium chloride (KCl) to achieve bradycardia. The procedure was complicated by the death of the pump twin. Intracardiac administration of KCl in one monoclonal twin has been shown by several authors to be associated with the death of the cotwin. Failure of alcohol ablation could therefore be the result of direct passage of KCl to the pump twin’s circulation and can not be solely attributed to the alcohol injection.

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