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Endoscopic laser coagulation of umbilical cord vessels in twin reversed arterial perfusion sequence

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ABSTRACT

In monozygotic twin pregnancies with reversed arterial perfusion (TRAP) sequence, the donor twin is at high risk of perinatal death. This paper describes the use of endoscopic surgery in the management of this condition. In four cases of TRAP sequence presenting at 17, 20, 26 and 28 weeks' gestation, respectively, an endoscope was introduced into the uterus under local anesthesia and a Nd–YAG laser was used to coagulate the umbilical cord vessels of the acardiac twin. Laser coagulation was successful in arresting blood flow to the acardiac fetus in the cases treated at 17 and 20 weeks, and healthy infants were delivered at term. In the pregnancies treated at 26 and 28 weeks, the umbilical cords were very edematous and laser coagulation failed to arrest blood flow; healthy infants were delivered after spontaneous labor at 29 weeks. These findings suggest that, during mid-gestation, endoscopic laser coagulation of the umbilical cord vessels of the acardiac twin is an effective method of treating TRAP sequence. In later pregnancy, alternative methods of treatment are needed.

INTRODUCTION

The most extreme manifestation of twin–twin transfusion syndrome is acardiac twinning (acardius chorioangiopagus parasiticus) which occurs in approximately 1% of monozygotic twin pregnancies. This twin disorder has been named twin reversed arterial perfusion (TRAP) sequence because the underlying mechanism is thought to be disruption of normal vascular perfusion and development of the perfused twin due to an umbilical arterial-to-arterial anastomosis with the donor or pump-twin[2]. At least 50% of donor twins die due to congestive heart failure or severe preterm delivery, the consequence of polyhydramnios[2,3]. All perfused twins die due to the associated multiple malformations.

This paper describes the application of endoscopically guided Nd–YAG laser[4,5] to coagulate the umbilical cord vessels supplying the perfused fetus in twin pregnancies affected by TRAP sequence.

MATERIALS AND METHODS

Nd–YAG laser coagulation of the umbilical cord vessels supplying the perfused fetus was attempted in four twin pregnancies affected by TRAP sequence. The condition was diagnosed by routine ultrasound examination and the patients were referred to our center at 17, 20, 26 and 28 weeks' gestation. In cases 1, 3 and 4 there was polyhydramnios. Ultrasound examination demonstrated that in all cases the pregnancies were monochorionic and diamniotic and the placentae were anterior. The perfused fetuses were acardiac, anencephalic and severely hydropic. The estimated fetal weight, calculated from the measurements of femur length and abdominal circumference, of the acardiac twin expressed as a percentage of that of the donor was 139–174%. The donor fetuses were appropriately grown and they did not have any obvious defect; however, in the three cases with polyhydramnios the fetal bladder appeared distended and the heart was dilated. The patients were counselled about the nature, prognosis and possible therapeutic options for the TRAP sequence, and gave written consent for endoscopic laser treatment. Approval for this therapeutic intervention was obtained from the hospital ethics committee.

Ultrasound examination was first performed to localize the placenta, the inter-twin amniotic membrane and the placental insertion of the umbilical cords. The appropriate site of entry on the maternal abdomen was chosen to avoid injury to the placenta or donor fetus and to allow access to the umbilical cord of the perfused fetus.

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Local anesthesia (lignocaine 2% infiltrated down to the myometrium) was then given. Under continuous ultrasound visualization, a rigid 2-mm diameter fetoscope (field of vision 75°) housed in a 2.7-mm diameter cannula (KeyMed, Southend, UK)[5] was introduced transabdominally into the amniotic cavity of the donor twin. A combination of ultrasonography and direct vision was used to identify the umbilical cord at the placental or fetal insertion. The fetoscope was pushed into the sac of the perfused twin and Hartmanns solution (50–100 ml) was infused to make visualization easier. A 400-μm diameter Nd–YAG laser fiber (MBB, Munich, Germany) was then passed down the side-arm of the cannula to 1 cm beyond the tip of the fetoscope. The umbilical cord artery and vein were then targeted using a pilot light and the Nd–YAG laser was administered by 3-s shots using an output of 30–50 watts at a distance of 1 cm. The total procedure took 10–30 min to complete and the patients were allowed home after a few hours.

RESULTS

In the two patients that were treated at 17 and 20 weeks' gestation, a total of 375 and 1000 joules were used, respectively, to achieve coagulation of the umbilical cord artery and vein. Color flow Doppler ultrasound, performed immediately after the procedure, demonstrated the lack of blood flow to the acardiac fetus. Subsequently, serial ultrasound scans demonstrated normal growth and biophysical profile in the surviving fetuses that were born after spontaneous labor at 39 and 38 weeks, weighing 2950 g (female) and 3000 g (male), respectively. Both babies are healthy and now at the age of 6 and 9 months, respectively, are developing normally. However, the first baby was noted to have a cardiac murmur in the neonatal period. Echocardiography revealed minimal right atrial and ventricular dilatation, and slight thickening and stenosis of the pulmonary valve. The pressure gradient across the valve was 10–15 mmHg and this was thought to be of no clinical significance.

In the pregnancies treated at 26 and 28 weeks, the umbilical cords were very edematous and laser coagulation failed to arrest blood flow despite the administration of 10 000 and 15 000 joules. In the first case, cordocentesis was performed and 20 ml maternal blood was injected into the Wharton's jelly with the aim of producing cord tamponade, but this was also unsuccessful in arresting blood flow. Subsequently, fetal blood sampling was carried out demonstrating severe fetal anemia that was treated by intrauterine fetal blood transfusion[6]; the fetal hemoglobin concentration increased from 4.7 g/dl to 9.1 g/dl. Another transfusion was given at 27 weeks when the respective pre- and post-transfusion fetal hemoglobin concentrations were 8.0 g/dl and 12.2 g/dl. At 29 weeks after spontaneous rupture of the membranes, an emergency Cesarean section was performed because of transverse lie. A male infant weighing 964 g was delivered together with the acardiac twin. He had an uneventful neonatal course and now at 8 months of age he is growing and developing normally.

The patient that was treated at 28 weeks had spontaneous labor at 29 weeks and delivered vaginally a healthy male weighing 1900 g. In the neonatal period, he had respiratory distress syndrome that was treated successfully. In addition, he had a cardiac murmur and echocardiography demonstrated a small ventricular septal defect. Now he is 8 months of age and is developing normally.

DISCUSSION

This study demonstrates the feasibility of a minimally invasive endoscopic technique in the disruption of TRAP sequence. In the two cases treated at 17–20 weeks' gestation, the pregnancies progressed uneventfully and resulted in the birth of healthy infants at term. However, by the end of the second trimester, laser coagulation of the umbilical cord vessels is impossible, presumably because of the protection provided by the thick edematous Wharton's jelly.

An alternative method of managing twin pregnancies complicated by the TRAP sequence is prevention of polyhydramnios-related preterm delivery by serial amniodrainage or the administration of indomethacin to the mother[7,8]. However, such therapy does not reduce the risk of congestive heart failure and consequent intrauterine or neonatal death of the donor twin. There is one case report of successful reversal of heart failure in the donor fetus by administration of digoxin to the mother[9].

Recent attempts at treatment have concentrated on surgical removal of the acardiac twin or occlusion of its umbilical cord. There are seven reported cases of selective removal of the acardiac fetus at hysterotomy which was performed at 19–26 (mean 22) weeks' gestation[10–12]. Postoperatively, the mothers remained in hospital for 5–34 days and received rigorous tocolytic therapy with indomethacin, magnesium sulfate and β-mimetics; two mothers developed mild pulmonary edema. In one case there was placental abruption and fetal death within 2 h of the procedure. In the other six cases, the healthy donor twins were delivered by Cesarean section at 27–37 (mean 33) weeks' gestation and survived; in five cases emergency delivery was undertaken because of placental abruption in two, premature labor in two and preterm prelabor rupture of membranes in one.

There are five previous reports of attempts to arrest blood flow in the umbilical cord vessels of the acardiac twin. Porreco and colleagues used an ultrasound-guided technique to introduce a thrombogenic coil in the single umbilical cord artery of the acardiac twin at 24 weeks' gestation[13]. There was immediate cessation of blood flow in the recipient cord and, after an uneventful pregnancy, the normal twin was delivered at 39 weeks' gestation. However, in two other cases the injection of thrombogenic coils or fibrin into the umbilical cord artery of the acardiac twin was associated with the death of both twins[14,15]. McCurdy and colleagues reported a pregnancy

complicated by heart failure in the donor and polyhydramnios at 19 weeks' gestation[16]. Amniodrainage was performed and maternal digoxin therapy was given but the hydramnios recurred and the heart failure persisted. Under general anesthesia, endoscopic ligation of the cord of the acardiac twin was performed but both fetuses died within 24 h of the procedure. More recently, Quintero and colleagues reported the successful ligation of the umbilical cord of an acardiac twin by fetoscopy at 19 weeks' gestation[17]. Under general anesthesia two 2-mm cannulas were introduced into the uterine cavity, one for the endoscope and the second for introduction of the suture; in addition, an 18-gauge needle was used for infusion of Ringer's lactate solution into the sac of the acardiac twin. The suture was manipulated round the cord and both ends were brought to the outside. A tight extracorporeal knot was tied and this was pushed onto the cord. A second knot was tied around the cord approximately 3 cm from the first. After tocolytic therapy and 36 h of hospitalization, the patient was sent home. Premature rupture of membranes occurred 2 weeks later but resolved within a week and a healthy baby was delivered at 36 weeks' gestation.

Moore and colleagues reported that the outcome of the donor twin depends on the weight of the acardiac fetus; when the percentage weight compared to the donor was > 70%, 50–70% and < 50%, the respective risk for congestive heart failure in the donor was 100%, 70% and 8%[3]. However, the value of these data in the antenatal management of affected pregnancies is limited by our current inability to estimate the weight of acardiac anencephalic fetuses with varying degrees of hydrops. Since in TRAP sequence perinatal mortality for the donor twin is at least 50%[2,3] and the condition can be diagnosed easily by routine ultrasound examination in early pregnancy, it could be argued that in all cases prophylactic treatment should be considered at mid-gestation. Fetoscopic laser coagulation of the umbilical cord vessels of the acardiac twin is effective and appears to be the least invasive of the currently available techniques. In patients presenting in later pregnancy with polyhydramnios and evidence of heart failure in the donor, alternative methods should be considered.

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