Case Report

10.21276/awch.2017.1862

Twin Reversed Arterial Perfusion (TRAP) Sequence with Fetal Intervention: Report of Two Cases

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ABSTRACT

We report 2 cases of TRAP sequence diagnosed antenatally using three-dimensional color Doppler ultrasound. Case 1 was a 44 year old G3A2 mother with monochorionic diamniotic (MCDA) twin pregnancy conceived after in vitro fertilization. Ultrasound imaging showed TRAP sequence at 18 weeks 6 days gestation with early onset intrauterine growth restriction of first twin and second twin being acardiac with flow reversal in umbilical artery and vein. Interstitial laser cord occlusion was performed at 20 weeks 6 days gestation. Serial ultrasound monitoring of growth of first twin done. The first twin delivered at 36 weeks 2 days gestation by cesarean section in view of poor growth. Baby weighed 1.55 kg at birth and had an uneventful neonatal period. Case 2 was 37 yr old primigravida with monochorionic diamniotic twin pregnancy conceived after in vitro fertilization. TRAP sequence diagnosed at 21 weeks 3 days gestation. Interstitial laser cord occlusion was performed at 25 weeks 5 days gestation and baby was delivered by cesarean section at 35 weeks 4 days gestation weighing 2.1 kg at birth. We conclude that early and accurate antenatal diagnosis is essential to improve outcomes in TRAP sequence. Minimally invasive intervention in the form of laser or radiofrequency coagulation of cord is safe and significantly improves the survival and long term neurodevelopmental outcome of pump twin.

Keywords: Acardiac Twin, Assisted Reproductive Technique, in Vitro Fertilization, Laser Coagulation, Pump Twin, Twin Pregnancy

Introduction

The Centers for Disease Control and Prevention (CDC) defines assisted reproductive technique (ART) as any procedure that entails the handling of both eggs and sperm or of embryos for the purpose of establishing a pregnancy. The prevalence of infertility is 10-15% in India. The in vitro fertilization (IVF) market in India has grown at a fast pace, with IVF cycles growing at 18.1% compound annual growth rate consequences being increased incidence of multiple gestation ranging from 21% to 43%. The use of ART has been suggested to be associated with an increased occurrence of monozygotic twinning (MZT) which is approximately 1.5%. MZT is associated with several significant obstetric complications, including preterm labor, intrauterine growth restriction, and fetal death. Twin to twin transfusion syndrome (TTTS) occurs in approximately 9-15% of monochorionic twins and the neurologic sequelae of the surviving twins generate further concerns. Twin reversed arterial perfusion sequence (TRAP) is a rare, unique complication of monochorionic twin pregnancy in which a twin with an absent or a nonfunctioning heart (“acardiac twin”) is perfused by its co-twin (“pump twin”) via placental arterial anastomoses. Estimated incidence of acardiac twins is 2.6 percent of monochorionic twin pregnancies and 1 in 9500 to 11,000 pregnancies. TRAP sequence is also known as acardius or chorioangiopagus parasiticus.

In TRAP sequence, a portion of pump twin’s cardiac output travels through placental arterial-arterial anastomoses to the umbilical artery and eventually to the systemic circulation of the recipient co-twin, thus creating “reversed” circulation in this twin. This is possible because the acardiac twin lacks a functional heart, whose pumping would normally provide forward flow and high systemic pressure. The presence of arterial-arterial anastomoses allows blood to be pumped from the normal twin to the acardiac twin without passing through a capillary bed. The presence of placental vascular anastomoses is common in monochorionic twins and alone is not sufficient for the development of TRAP sequence. TRAP sequence can be diagnosed as early as 11 weeks gestation and is confirmed by assessing the flow pattern in the acardiac twin in a monochorionic pregnancy and continued growth of the acardiac twin on serial ultrasound examinations. The umbilical cord contains 2 vessels in up to 70% of acardiac fetuses. Treatment options are focused on the improvement of the outcome of the pump twin as the acardiac twin is non viable and include antenatal intervention, delivery and expectant management. In this article we report 2 cases of monochorionic pregnancy antenataly diagnosed with TRAP sequence and managed with laser coagulation with successful obstetric and neonatal outcome of the pump twin.
Case Report

Case 1: A 44 year old G3A2 mother with MCDA pregnancy conceived after IVF treatment was diagnosed with TRAP sequence at 18 weeks 6 days gestation using three-dimensional color Doppler ultrasound. Mother was a booked case at an infertility center in Chennai. No history of obstetric and medical risk factors. Ultrasound scans prior to 18 weeks were normal. USG done at 18 weeks suggestive of early onset IUGR of the first twin other one being acardiac fetus with volume of 110ml and flow reversal seen in the umbilical artery and umbilical vein. Mother was under strict supervision with twice weekly ultrasound. Polyhydramnios and increase in the size of the acardiac twin compared to previous scans at 20 weeks gestation. Interstitial laser cord occlusion was performed after consent and under strict aseptic precautions at 20 weeks 6 days gestation. Procedure was free of any immediate complications. Pregnancy was monitored thoroughly. Serial USG done showed large inlet VSD and short long bones with lag of 4 - 5 weeks, fall in growth velocity centiles, but later growth trend remained same. Acardiac twin volume reduced to 30ml and no color flow noted at 34 weeks 3 days gestation. Mother was shifted to our center prior to delivery. Baby was delivered by LSCS at 36 weeks 2 days gestation with birth weight of 1.55kg (<10 centile). Figure 1 shows the placenta with cord that perfused the acardiac twin. Figure 2 shows the acephalic and acardiac second twin. Baby was admitted in NICU in view of prematurity. Baby had an uneventful neonatal period except for jaundice requiring phototherapy for 2 days. Postnatal 2D ECHO showed 3 mm PDA and 6 mm muscular inlet VSD which did not warrant any treatment. Ultrasound abdomen and cranium were normal. Feeds were gradually established and baby discharged 2 weeks after birth. Baby was followed up till 18 months postnatal age. Baby was growing well with normal neurodevelopmental outcomes.

Case 2: 37 yr old primigravida with MCDA pregnancy conceived after IVF treatment was diagnosed with TRAP sequence at 21 weeks 3 days gestation using 3D color Doppler ultrasound. No history of antenatal and medical risk factors. Mother underwent biweekly Doppler ultrasound. Interstitial laser cord occlusion was performed after consent and under strict aseptic precautions at 25 weeks 5 days in view of hydramnios of pump twin and increasing volume in acardiac twin (402ml). Serially repeated doppler USG showed no flow in the acardiac twin and volume reduced to 37ml with high resistance flow in the umbilical artery and increased diastolic flow seen in middle cerebral artery (MCA) and peak systolic velocity (PSV) fell at 1.38 MoM s/o cerebral redistribution. Baby was delivered by LSCS at 35 weeks 4 days gestation with birth weight of 2.1kg. Baby and cranium were normal. Baby is currently under follow up and growth and neurodevelopmental outcome at 20 months postnatal age was normal.

Table 1: Summary of case reports from India.

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Place</th>
<th>Obstetric formula</th>
<th>Age of the mother</th>
<th>Time at diagnosis</th>
<th>Fetal intervention</th>
<th>Neonatal outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Babu et al^12</td>
<td>2015</td>
<td>Chennai</td>
<td>G2A1</td>
<td>23 Yrs</td>
<td>16 wks</td>
<td>RFA at 28 wks</td>
<td>Emergency LSCS at 34 wks 1.76 kg and alive</td>
</tr>
<tr>
<td>Saritha et al^13</td>
<td>2013</td>
<td>Hyderabad</td>
<td>Primi</td>
<td>25 yrs</td>
<td>36 wks</td>
<td>None</td>
<td>LSCS at 36 wks 2.3 kg and alive</td>
</tr>
<tr>
<td>Seema et al^14</td>
<td>2015</td>
<td>Delhi</td>
<td>Primi</td>
<td>27 yrs</td>
<td>29 wks</td>
<td>Cord embolization failed and preterm labor</td>
<td>1.4kgs and alive</td>
</tr>
<tr>
<td>Iqbal et al^15</td>
<td>2014</td>
<td>Kerala</td>
<td>G4P2A2</td>
<td>28 yrs</td>
<td>36 wks</td>
<td>None</td>
<td>LSCS at 36 wks 2.9kgs and alive</td>
</tr>
<tr>
<td>Khanduri et al^16</td>
<td>2015</td>
<td>Lucknow</td>
<td>Primi</td>
<td>22 yrs</td>
<td>31 wks</td>
<td>None</td>
<td>LSCS at 36 wks Wt not known</td>
</tr>
<tr>
<td>Prameela et al^17</td>
<td>2014</td>
<td>Mysore</td>
<td>Primi</td>
<td>21 yrs</td>
<td>24 wks</td>
<td>None</td>
<td>Preterm labor at 24 wks 1.6 kg and died</td>
</tr>
<tr>
<td>Shashidhar et al^18</td>
<td>2012</td>
<td>Kolar</td>
<td>G3A2</td>
<td>28 yrs</td>
<td>Following delivery</td>
<td>None</td>
<td>Gestation not known 2.3kg and died</td>
</tr>
<tr>
<td>Pushpalatha et al^19</td>
<td>2015</td>
<td>Nagar</td>
<td>G2P1L1</td>
<td>26 yrs</td>
<td>16 wks</td>
<td>None</td>
<td>Termination at 19 wks 500 gm</td>
</tr>
</tbody>
</table>

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Discussion

Acardiac twin is classified according to the degree of cephalic and truncal maldevelopment1. Most common anomaly is acardius acephalus in which fetal thoracic organs and head are absent. Our both cases were acardius acephalus. In acardius acormus, only fetal head develops. In acardius amorphus consists of a shapeless mass of tissue with no recognizable human parts and acardius myelacephalus in which head or one or more extremities are partially developed.

The indicators for poor prognosis are as follows10:

1. Ratio of weight of acardiac twin/pump twin >0.70
2. Polyhydramnios
3. Cardiac failure in the pump twin
4. Increase in the relative size of the acardiac twin
5. Hydrops in the pump twin
6. Monoamniotic pregnancy (cord entanglement risk)

Pump twin may develop signs of high output failure like polyhydramnios, cardiomegaly, pericardial and pleural effusions, ascites and tricuspid regurgitation. Development of cardiac failure in the pump twin is related to ratio of the size of acardiac twin to pump twin.

Early attempts were highly invasive and involved hysterotomy and removal of the acardiac twin.

Subsequently, a series of other less invasive intrauterine interventions aiming to arrest the circulation of the acardiac twin were introduced. These included insertion of cord coils, ligation with or without transection of the umbilical cord, endoscopic laser coagulation of placental anastomoses between the pump and acardiac twins, endoscopic laser coagulation and endoscopic or ultrasound-guided monopolar or bipolar diathermy of vessels within the cord supplying the acardiac twin and, more recently, ultrasound-guided ablation of intrafetal vessels by injection of alcohol, monopolar diathermy, laser or radiofrequency.

Laser and bipolar coagulation occlude a portion of the umbilical cord near its placental insertion site. Radiofrequency ablation (RFA) is used to coagulate the abdominal wall at the base of the umbilical cord, rather than direct coagulation of the umbilical cord itself. Intrafetal laser treatment is used for gestational ages less than 16 weeks whereas RFA after 16 weeks because of the higher blood flow rate.

Endoscopic laser surgery uses a rigid 2-mm-diameter fetoscope, housed in a 2.7-mm-diameter cannula introduced transabdominally into the sac of the pump twin and then advanced into the sac of the perfused twin, and neodymium-doped yttrium aluminum garnet laser used to coagulate the umbilical cord artery and vein.

Fig. 1: Placenta with laser coagulated cord that perfused the acardiac twin.

Fig. 2: Acardius acephalus twin.
For intrafetal laser, a transverse section of the lower fetal abdomen of the acardiac twin was obtained with ultrasound, and colour flow Doppler used to visualize the feeding vessels. An 18-gauge needle introduced, and under continuous ultrasound visualization was guided to the fetal abdomen with the tip adjacent to the pelvic vessels. A 400-μm laser fiber then inserted into the needle and advanced to a couple of millimetres beyond the tip of the needle. A YAG or diode laser provides 20 to 40 watts of energy. For RFA, a 14 to 17 gauge needle is inserted into the amniotic cavity of the acardiac twin under ultrasound guidance. Cessation of blood flow from the pump twin to the acardiac twin is confirmed by Doppler ultrasound prior to the end of any procedure. In our cases intra fetal laser coagulation was performed.

In one series of 49 pregnancies affected by TRAP sequence, the perinatal mortality rate for pump twins was 55% without treatment. The principal reasons for this high mortality were development of heart failure and preterm birth and its sequelae. In another series, all five expectantly managed fetuses died in utero before 20 weeks of gestation. Studies generally report survival of 80 to 90 percent of pump twins in pregnancies that underwent in utero coagulation/ablation procedures compared to 45% with expectant management. Maternal complications with intervention are uncommon, and include bleeding, need for laparotomy to complete the procedure, thermal injury, chorioamnionitis leading to maternal sepsis, and disseminated intravascular coagulation. Fetal complications are miscarriage, preterm labor, premature rupture of membrane and amniorrhexis.

TRAP pregnancies are usually delivered between 34-36 weeks of gestation with at least one course of antenatal steroids given between 23-34 weeks.

Various case reports have been published so far across the world. Our report is highlighted by the fact that both are conceived following minimally invasive interventions is good.

Conclusion
Careful monitoring of monochorionic multiple gestation pregnancies is needed for early identification of complications like TRAP sequence which is essential to improve outcomes. Minimally invasive intervention in the form of laser or radiofrequency ablation of cord is safe and significantly increases the survival of pump twin. Intrafetal laser is safe if performed by experienced hands. Pregnancy outcomes following fetal intervention have improved in the past decade due to improved surveillance and monitoring. Long term neurodevelopmental outcomes following minimally invasive interventions is good.

Acknowledgements
Dr S Suresh, Fetal medicine specialist, Mediscan systems, Chennai

Reference
3. Call for Action: Expanding IVF treatment in India 2015.

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Financial or other Competing Interests: None.