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BRIEF COMMUNICATION

Acardiac Twin: A Report of Two Cases

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Abstract Multiple gestations, especially monochorionic twins are associated with unique complications such as twin to twin transfusion syndrome (TTTS) and twin reversed arterial perfusion (TRAP) sequence due to preferential blood flow within the vascular communications between the two fetuses. TRAP sequence is a rare complication of monochorionic twins. The authors describe two cases of acardiac twins, one diagnosed at 17 weeks and the other at 24 weeks. While the first one was lost to follow-up, the second pregnancy was managed expectantly with close antepartum surveillance and had a term vaginal delivery of a healthy baby, along with the acardiac twin which was non-viable. The pump twin continues to be developmentally normal.

Keywords Monochorionic twins · Acardiac twin · TRAP sequence · Hydrops in twins

Case Reports

Case 1

A 21-year-old primigravida was referred to authors' hospital at 17-week gestation with an ultrasound showing twin gestation with anomaly in one twin. This was a spontaneous conception after 2 year of married life. There was no history of twinning in the family and she was not found to have any antenatal risk factors. There were no scans done in the first trimester. She had her first ultrasound (USG) at 17 weeks and was told to have twin pregnancy with one anomalous twin. She came to the authors for a second opinion.

Scan done in authors' institution confirmed multiple gestations. There was a single placenta, located posteriorly, with a thin dividing membrane between the two gestational sacs. Twin A had grossly abnormal anatomy with absence of the heart in the thorax, along with lack of head and upper limbs (Fig. 1). The spine ended abruptly in the cervical region. Umbilical artery Doppler study showed reversal of flow. Twin B was structurally normal with fetal biometry corresponding to the gestational age and had no evidence of hydrops or polyhydramnios (Fig. 2). The options for further management were discussed with her, including expectant management with close monitoring of pump twin on ultrasonography (USG) and delivery once the normal twin developed hydrops; cord ligation or laser coagulation of umbilical cord of acardiac twin under fetoscopic guidance and termination of pregnancy. She was referred to the fetal medicine centre for further evaluation and management. Unfortunately, she was lost to follow-up.

Case 2

A 30-year-old gravida 3 was referred to authors' institution at 24-week gestation with an USG showing twin gestation with abnormality in one of the twins. Married for 11 year, she had two children aged 10 and 5 years, both were delivered vaginally. This was a spontaneous conception and thus far, she did not have any complications in this pregnancy. She had her first scan at 16-week gestation and was told to have single, live fetus, corresponding to the gestational age.

A repeat scan done at 24 weeks, however, revealed twin pregnancy, with twin A corresponding in growth to 24–25 weeks with no gross abnormalities. In twin B, only the thorax and abdomen were visualized. Heart, cranium and

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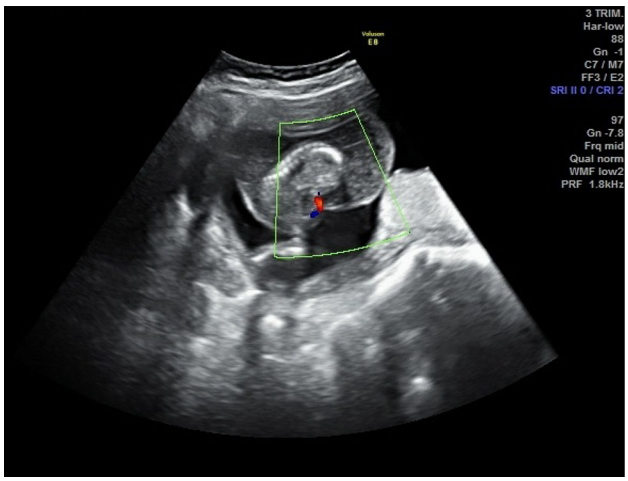

Fig. 1 (Case 1) Twin A: An amorphous mass with absent head, heart and limbs suggestive of acardiac twin. Color doppler study confirms the absence of heart in the thorax

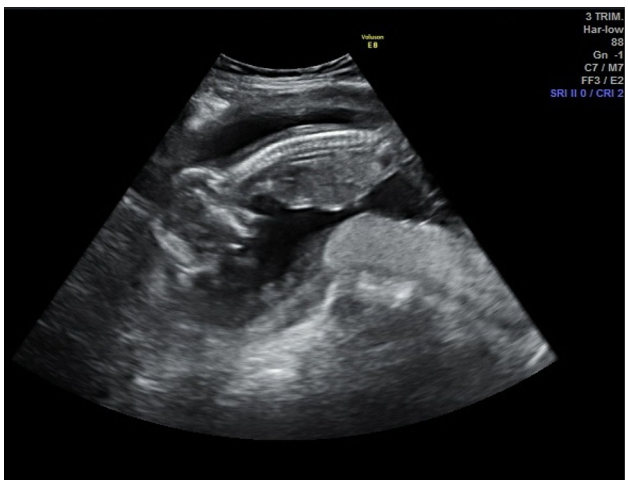

Fig. 2 (Case 1) Twin B: Monochorionic counterpart of the acardiac twin, showing normal anatomy and biometry. A single placenta is noted posteriorly

all limbs were absent and no fetal movements were noted. There was a single placenta, located posteriorly with thin dividing membrane between the amniotic sacs.

Scan done at 25 weeks confirmed the above findings and diagnosis of twin reversed arterial perfusion (TRAP) sequence with acardiac twin was made. The pump twin showed no evidence of hydrops or polyhydramnios at initial evaluation. The patient was counseled regarding poor prognosis of the abnormal twin and was advised close monitoring of the pregnancy. She was followed-up fortnightly with serial scans to look for evidence of hydrops in the pump twin. Interval growth, amniotic fluid index and umbilical artery Doppler were also assessed at each visit. Serial scans showed consistent interval growth, normal umbilical artery Doppler and no evidence of hydrops in the pump twin. The size of the acardiac twin was constant throughout the pregnancy.

Labour was induced at 37-week gestation and she had normal vaginal delivery of both fetuses. A healthy baby boy weighing 2.5 kg was born (Fig. 3) along with expulsion of the acardiac mass weighing 330 g (Fig. 4). The placenta weighed 490 g and was found to be monochorionic with presence of vascular anastomoses between the acardiac and normal twin on gross examination (Fig. 4). The pump twin is now 2-year-old and is developmentally normal.

Discussion

TRAP sequence is a rare complication of monochorionic twin gestation, affecting approximately 1% of such cases [1]. Due to lack of a beating heart, the acardiac twin is dependant on pump twin for its circulation. This is assisted by the presence of large arterial anastomoses within monochorionic placenta. Under normal circumstances, the umbilical arteries carry the deoxygenated blood away from the fetus to the placenta in order to receive oxygen and nutrients from the maternal circulation. In an acardiac twin, however, the deoxygenated blood in the umbilical artery flows towards the fetus, preferentially perfusing lower part of the body. Hence, the entity is known as "twin reversed arterial perfusion". Chronic hypoxemia leads to maldevelopment of upper part (cephalic and truncal components) of the acardiac twin, making it non-viable.

The normal or pump twin has not only to take care of its own perfusion, but also that of its acardiac counterpart. This leads to hemodynamic stress and, ultimately, cardiac failure and death. The risk of cardiac failure in the pump twin depends upon the relative size of the acardiac twin in

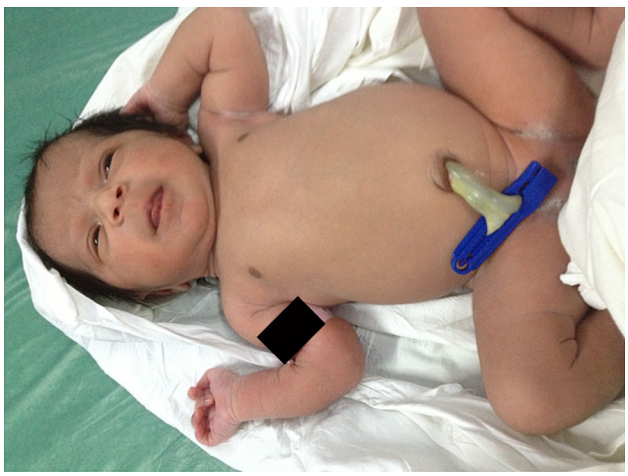

Fig. 3 (Case 2) Pump twin at delivery: healthy male baby weighing 2.5 kg

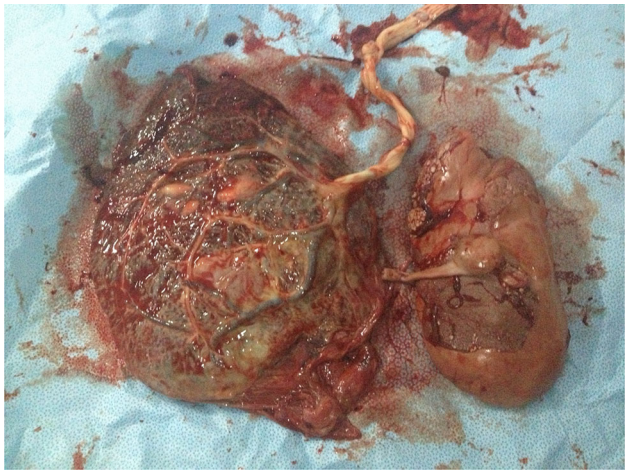

Fig. 4 (Case 2) Monochorionic placenta with acardiac twin and umbilical cord of the pump twin

relation to its pump. If the ratio of size of the acardiac to its pump twin is >0.70, then risk is increased to 30% from 10% when the ratio is less. Left untreated, there is 50–55% mortality of pump twin [2].

The management options in such cases include: delivery, expectant management and intrauterine interventions. Expectant management includes close monitoring with serial scans. This is indicated when the size of acardiac twin is <70% size of pump twin, with no evidence of cardiac failure or hydrops in the latter. Serial ultrasounds are done to look for hydrops and/or polyhydramnios in the pump twin. On Doppler evaluation, absent/reversed flow in umbilical artery and abnormal ductus venosus waveform are suggestive of cardiac failure in the pump twin. Antenatal steroids is administered between 24 and 34 weeks because of increased risk of preterm labour.

Intrauterine interventions are indicated in fetuses with one or more poor prognostic features. These include: size of acardiac twin >70% of pump twin; polyhydramnios (defined as maximum vertical pocket as ≥8 cm); development of cardiac failure or hydrops in pump twin and increase in relative size of cardiac twin [2–4].

Aim of in utero intervention is to prevent morbidity and mortality in the pump twin due to evolving cardiac failure by causing dissociation of circulations of both twins. The current treatment modalities include ligation of the umbilical cord of the acardiac twin; fetoscopic laser coagulation or radiofrequency ablation or bipolar diathermy of vessels in the umbilical cord supplying the acardiac twin and endoscopic laser coagulation of placental anastomoses between the acardiac and the pump twin [5–9]. USG guided ablation of intrafetal vessels by using monopolar diathermy, laser or radiofrequency has become popular in the last two decades [10]. The perinatal mortality with expectant management is 55% in comparison to only 20% with intrauterine interventions [11].

With the widespread use of first trimester scan for aneuploidy screening, more cases of TRAP are being picked up at 11–13 weeks. Lewi et al. found that following early diagnosis, if intervention is delayed until 16–18 weeks, it not only leads to spontaneous cessation of circulation in the acardiac twin in 60% cases, but also to death or brain damage in 60% of the pump twin [12]. Hence early intervention is being advocated now.

The technique of choice in early intervention is intrafetal laser coagulation or radiofrequency ablation rather than fetoscopic based procedures since they are less invasive (as they can be done via 17–18 G needle inserted in utero, without requiring a fetoscope) and hence, less likely to cause miscarriage. The optimal timing of such intervention being proposed is 12–14 weeks [13]. However, further studies will be required to compare the survival of pump twin in early (12–14 weeks) vs. delayed intervention (16–18 weeks).

Conclusions

TRAP sequence is a rare complication of monochorionic twins. A high index of suspicion is required for accurate diagnosis and optimal subsequent management of the normal twin. Such cases need to be referred to and evaluated by fetal medicine specialists, especially those with expertise in fetoscopic interventions. Close monitoring of the pump twin and delivery at the right time is of paramount importance in fetuses with TRAP sequence being managed expectantly. In utero therapy is indicated in fetuses with presence of one or more poor prognostic features on USG. These include cord ligation of the acardiac twin, fetoscopic laser coagulation, diathermy or radio-frequency ablation of the cord and intrafetal laser ablation or radiofrequency of vessels in the acardiac twin. The overall survival rate with in utero interventions is 80% as compared to 45% with expectant management. Intrafetal laser coagulation of feeding vessels in the acardiac twin is the preferred modality especially in the early intervention at 12–14 weeks.

Compliance with Ethical Standards

Conflict of Interest None.

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