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Case Report
Acardiac Twin

Twin Reversed Arterial Perfusion Syndrome (TRAP or Acardiac Twin) – A Case Report

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Acardiac Twin known as TRAP (Twin Reversed Arterial Perfusion) syndrome is a rare and serious complication of mono-zygotic multiple gestation. This results in the existence of an Acardiac twin which fails to develop the head, upper limbs, and heart and is dependent upon a normal twin to provide circulation using vascular anastomosis. The pump twin although structurally normal will have high morbidity and mortality. This case report presents an antenatally diagnosed mono-chorionic twin pregnancy with delivery of a normal twin and Acardiac amorphous co-twin.

Keywords: Cardiac Twin, Fetal Anomaly, Twin Reversed Arterial Perfusion Syndrome

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None	Nil	Written Informed Consent was obtained from the patient for publication of this case report with clinical details and accompanying images.	15.21	

Introduction

Multiple gestation accounts for 10% of perinatal morbidity and mortality. Foetuses in multiple pregnancies suffer a variety of complications, some of which are unique to multiple gestations such as TTTS (twin-to-twin transfusion syndrome) and TRAP (twin reversed arterial perfusion syndrome). Zygosity is important in determining the complications as monozygotic have unique complications than dizygotic pregnancy. The incidence of TRAP is 1 in 35000 twin pregnancies [1] and 1% of mono chorionic twin pregnancies.

TRAP sequence occurs due to abnormal placental vascular anastomosis (arteroaterial) [2] which results in the consequent flow of deoxygenated blood from the pump twin to the recipient twin through the umbilical artery (reversal of blood flow in Acardiac twin). This causes a spectrum of anomalies as a consequence of severe hypoxemia with 100% mortality of Acardiac twins. The pump twin suffers congestive cardiac failure and hydropic changes due to pumping blood to the Acardiac twin. Mortality for pump twins is 50-70% [3]. However, early identification, follow-up and treatment improve the survival rate of the pump twin. We report a case of TRAP sequence, which created a considerable management dilemma in saving the pump twin.

Case Report

A 33-year-old G5P4L4 with 2 previous caesarean sections with an anomalous scan at 18 weeks suggestive of twin gestation with single live foetus and another foetus with absent heart pulsations (? vanishing twin syndrome). A growth scan was done at 28 weeks suggestive of twin gestation with a single live foetus with normal Doppler study and a Second twin as an Acardiac twin. Pregnancy was followed with sequential Doppler Ultrasonography.

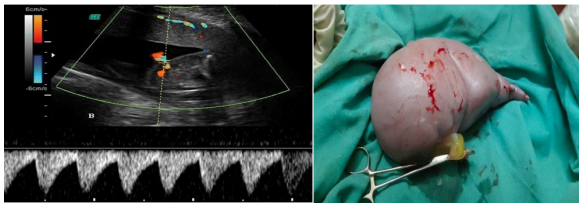

Fig 1: Doppler showing the reverse arterial flow

After coverage of 2 doses of steroids, the pregnancy was terminated by Elective caesarean section

At 36 weeks and delivered alive female weighing 1.91 kg with Apgar scores of 7 and 8 and no gross congenital malformations. The second twin was dysmorphic with no distinct head, upper limbs, spine, and lower limbs with short umbilical cord. Histopathology of Placenta confirmed monochorionicity. The first twin was admitted to the NICU for prematurity and discharged.

Fig 2: Normal twin and Acardiac twin

Discussion

Acardiac twining is one of the rare congenital anomalies characterized by the formation of a malformed foetus which is nonviable and compromises the well-being of the pump twin by following mechanisms: [4]

1. Increased blood flow can lead to congestive cardiac failure
2. Flow of de-oxygenated blood from the Acardiac twin to the pump twin can cause hypoxia and growth restriction.
3. Preterm delivery

According to Malone's classification [5], the acardiac twin in the TRAP sequence is described as hemicardius (imperfectly formed heart) and holoacardius (absence of heart).

According to Joseph C classification6, an acardiac fetus is categorised into

1. Acardius-acephalus - no cephalic development, most common variety
2. Acradius-anceps - some cranial structures and neural tissue development seen and the most advanced type of acardius [7]
3. Acardius-acormus-cephalic structures are present with absent truncal development
4. Acardius-amorphous - least developed with no distinguishable cephalic or truncal structure.

Acardius myelacephalus refers to a partially developed head with identifiable upper limbs with some nervous tissue [8]. Sonographic documentation of reverse flow in the umbilical arteries is a pathognomonic feature of the TRAP sequence [9].

Interventions in the past were highly invasive. Subsequently, less invasive techniques were introduced such as 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 causes occlusion of the umbilical cord at 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. Endoscopic laser surgery uses a rigid 2-mm-diameter fetoscope and introduces neodymium-doped yttrium aluminium garnet laser to coagulate the umbilical cord artery and vein.

Conclusion

Careful monitoring of monochorionic twin gestation pregnancies is required for early identification of complications like TRAP sequence. TRAP sequence requires continuous monitoring with Doppler USG. Minimally invasive interventions such as Laser coagulation and radiofrequency ablation of the cord are relatively safe. Long-term neurodevelopmental outcomes following minimally invasive interventions are good.

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