A rare case of twin reversed arterial perfusion (TRAP) sequence

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ABSTRACT

Twin reversed arterial perfusion (TRAP) sequence is a rare, unique complication of monochorionic twin pregnancy in which one of the twins with an absent or a non-functioning heart (acardiac twin) is perfused by its own co-twin (pump twin) via placental arterial anastomosis. The anomalous twin may appear as a heterogenous mass simulating a teratoma or intrauterine foetal demise and has poorly developed heart, upper body and head. The normal twin faces the excess burden of having to send and receive blood from the acardiac mass as well as its own growing tissue and is at risk of heart failure and preterm birth. Left untreated, in up to 50% of cases, this otherwise normal twin may die in utero or shortly after birth. We report the case of a TRAP sequence in a monochorionic twin pregnancy. Unfortunately, the pump twin died because mother went into premature labour that was complicated by abruptio placenta at 26 weeks of gestation.

Keywords: Twins, monozygotic, placenta, ablation techniques

INTRODUCTION

Twin reversed arterial perfusion (TRAP) sequence is a rare condition seen one in 35,000 of normal pregnancies or one percent of monozygotic twin pregnancies. [1] In this condition, one twin appears normal while the other has multiple lethal anomalies. This situation occurs when the artery to artery anastomosis on a shared placenta causes a perfusion pressure of one twin to overtake that of the other, which then has reversed arterial flow. Blood from the pump or normal twin enters the recipient's iliac vessels so that the lower part of the body is perfused more than upper part. This results in acardiac foetal malformation or acardiac twin. [2] The acardiac twin can present in various ways but the most common is the failure of the foetus to develop either a heart (acardiac) or a head which is incompatible with life. The pump twin is typically normal, however the added circulatory burden can lead to cardiomegaly and heart failure. This may progress to hydrops foetalis with a mortality rate of 50 to 75 % for the pump twin. [3] The risk of chromosomal anomalies in pregnancies with TRAP sequence has been reported in up to 50% of pump twins.

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The diagnosis of a TRAP can be difficult and can be missed. We report an interesting case of a TRAP sequence which was not suspected antenatally that we had encountered in our institution.

CASE REPORT

A 32-year-old Indonesian housewife presented with a complicated second pregnancy. Her first pregnancy was uneventful. In the second pregnancy, antenatal booking in the Maternal Child-Health Clinic was made at 19^{th} week gestation and at this point, ultrasound scan (USS) showed that she had a monochorionic diamniotic twin pregnancy. Twin I was diagnosed to have cystic hygroma with no foetal cardiac activity seen while Twin II was alive at 18^{+2} weeks gestation with no gross abnormality seen.

Her antenatal screen blood pressure and blood investigations were all normal. She was then referred to the Obstetric and Gynaecology clinic and admitted for further investigations and counseling. She was counseled on the risk of miscarriage, intrauterine growth restriction and abnormalities in Twin II baby as well as recommended for closer follow up with serial USSs and blood tests. She was discharged and had two outpatient follow ups. She had another USS which showed polyhydramnios with an amniotic fluid index (AFI) of 30.96cm.

She presented to the labour room in active labour at 25 weeks gestation. Twin I was delivered weighing 340gm and looked severely deformed, described as unidentifiable head, neck and face – like 'a ball of mass'. It had a short right arm which was abnormal and had no left upper limb. The gender could not be ascertained. The Twin II foetal heart sound was difficult to trace and at this point abruptio placenta was suspected. This was confirmed after artificial rupture of membrane of Twin II which revealed blood stained liquor. Twin II baby's delivery was expedited with oxytocin and subsequently the patient delivered a fresh stillbirth female baby weighing 550gm with no gross abnormality. In our case, a twin-to-twin transfusion syndrome was initially suspected and the diagnosis of a TRAP sequence was only made post-natally.

DISCUSSION

In TRAP sequence, the pump twin maintains its normal pattern of foetal circulation. In addition a portion of its cardiac output travels through the placental arterial anastomosis to the umbilical artery and eventually the systemic circulation of the recipient co-twin. This creates a reversed circulation in this twin. The presence of arterial-arterial anastomosis allows blood to be pumped from the normal twin (pump twin) to the acardiac twin without pressure through a capillary bed. In such cases, veno-venous and arteriovenous anastomoses can also occur. [5]

In TRAP sequence, the acardiac twin is classified according to the degree of abnormal development. The most common encountered anomaly is the acardiacacephalous where the foetal thoracic organs and head are absent. Other rare types includes; a) acardiusacormus where only the foetal head develops, b) acardius amorphous that consists of a shapeless mass of tissue with no recognisable human parts and c) acardiusmyelacephalus where the head and one or more the foetal extremities are partially developed. [6] In our case, the Twin I was consistent with the

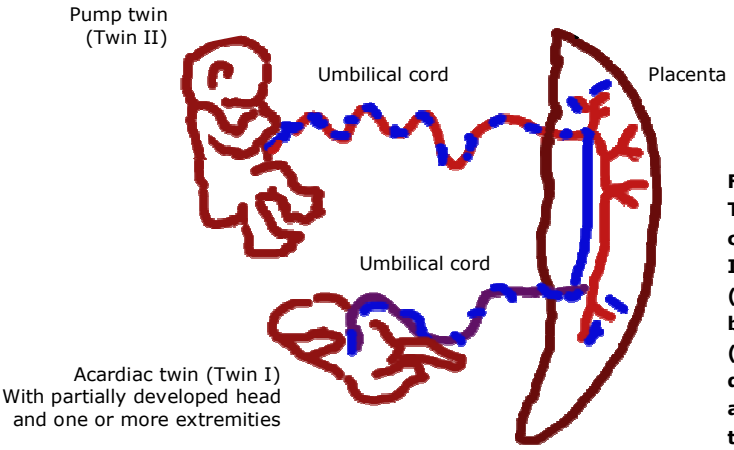

Fig. 1: Diagrammatic illustration of a TRAP sequence in our case: The cardiac output of the pump twin (Twin II) supplies blood to the acardiac twin (Twin I) resulting in a compromised blood flow to the pump twin itself. (Red: oxygenated circulation, Blue: deoxygenated circulation and Purple: arterial circulation of the acardiac twin).

acardiusmyelacephalus type.

TRAP sequence can be diagnosed pre-natally based on characteristic USS findings. However, this can be very difficult as in our case. It should be suspected when one foetus appears anatomically normal and the other lacks apparent cardiac structures and or activity. The diagnosis is confirmed by assessing the flow pattern to the acardiac twin and is supported by observation of continued growth of the acardiac twin on serial USS exam. The umbilical cord has a single umbilical artery in up to 70% of acardiac foetuses. Up to 50% of acardiac twins have cystic hygroma. The differential diagnosis of TRAP sequence is a single intrauterine demise of a twin gestation or an anomalous second twin.

As the acardiac twin is non-viable, treatment for TRAP sequence is focused on improving the outcome for the pump twin. Indicators of poor prognosis include are shown in Table 1. [7]

For continuing pregnancies with TRAP, weekly USS surveillance of the pump twin to look for signs of foetal hydrops and abnormal doppler studies of the umbilical artery, umbilical vein and ductus venosus is recommended. USS surveillance is increased to twice weekly if there is evidence of pre-hydrops (i.e. fluid in one cavity-ascites, pleural effusion). Given the risk of preterm birth, antenatal corticosteroids should be administered to all patients between 24-34 weeks of gestation. [5]

Table 1: Indicators of poor prognosis in a TRAP sequence.

Weight ratio of the acardiac twin: pump twin that is > 0.70
Hydramnios
Cardiac failure in pump twin indicated by abnormal doppler studies including persistent absent or reversed diastolic blood flow in the umbilical artery, pulsatile blood flow in umbilical vein or reversed blood flow in the ductus venosus.
Hydrops in the pump twin
Increase in relative size of the acardiac twin: acardiac : pump twin ratio > 1.0 is considered significant.

For pregnancies with one or more of the poor prognostic criteria, antenatal intervention, delivery and expectant management are the options. Historically interventions in pregnancies with TRAP sequence had been limited to amnio-reduction to reduce hydramnios or relief for the pump twin by administration of sclerosing agents such as alcohol into the umbilical cord of the acardiac twin. [8] For pregnancies with gestational age of between 18 and 27 weeks, current treatment modalities target occlusion of the umbilical cord of the acardiac twin. These options include laser ablation, bipolar cord coagulation and radiofrequency ablation which are performed with local anaesthesia and conscious sedation. [9-11] Foetoscopic cord ligation is an alternative but is less commonly used.

TRAP pregnancies should be delivered at 34 to 36 weeks of gestation depending on the patient's clinical scenario. Indications of compromise in the pump twin would prompt early delivery. Caesarean delivery is indicated for the usual obstetric indications.

In conclusion, TRAP sequence should be strongly suspected if one twin is normal looking and the other is grossly anomalous and lacks apparent cardiac structures or activity. TRAP can be easily differentiated from the more commonly seen Twin-to-Twin Transfusion (TTTS) in monochorionic twins by the presence of polyhydramnios and foetal compromise in the donor twin. In TTTS foetal compromise and polyhydramnios is seen in the recipient twin as there is an imbalance between arteriovenous anastomosis working in each direction, resulting in net flow towards one of the twin (recipient twin).

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