CASE REPORT

DIAGNOSIS AND MANAGEMENT OF TWIN REVERSED ARTERIAL PERFUSION SEQUENCE AT THE KORLE BU TEACHING HOSPITAL, GHANA

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Abstract

Twin Reversed Arterial Perfusion (TRAP) sequence is a rare complication of twin gestation unique to monochorionic twins, reported to occur in about 1% of all monochorionic pregnancies. In this condition, an acardiac twin, with an absent or non-functioning heart, is perfused by its co-twin (referred to as the "pump" twin) via placental anastomoses. It is associated with a high mortality for the pump twin without intervention. We report two cases of TRAP sequence diagnosed prenatally by ultrasound at the Korle Bu Teaching Hospital. We describe how this rare anomaly can be diagnosed and managed successfully even in a low resource setting to yield good perinatal outcomes.

Key Words: TRAP sequence, monochorionic, twin gestation, acardiac twin, pump twin, congenital anomalies

Introduction

Twin Reversed Arterial Perfusion (TRAP) sequence is a rare complication of monochorionic twin pregnancies. It is also known as acardiac twinning due to the fact that one twin (the "recipient") lacks a functioning cardiac system and so is perfused by the normally developing "pump" (donor) twin with deoxygenated blood through anomalous vascular connections in the placenta. Without treatment, mortality in the "pump" twin may be as much as 50-75%.[1]

Based on data published in 1953, the incidence of TRAP sequence is about 1 % of monochorionic twin pregnancies and 1 in 35,000 pregnancies overall.[2] However, a recent study by van Gemert *et al* indicates that this condition may not be as rare as was once thought, occurring in about 1 in 9,500 to 1 in 11,000 pregnancies.[3] The rate of recurrence is estimated at 1 in 10,000.[4]

This case series describes two such rare occurrences diagnosed in Accra, Ghana. The challenges and pitfalls in the diagnosis and management of these patients in low-resource settings will also be discussed in comparison with recommended guidelines. This case series hopes to create awareness about this seemingly rare conditions and highlight the need for increased vigilance in prenatal diagnosis.

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Case report

Case 1

The patient was a 32-year-old primigravida, who presented at 40 weeks gestation for a routine ultrasound evaluation of foetal wellbeing. She was a regular antenatal care attendant and had had six uneventful visits during the index pregnancy as at the time of presentation. Her gestational age had been estimated from an ultrasound scan done in the 10th week which had reported a singleton pregnancy. She however had not had any other ultrasound evaluation of the foetus until she presented.

The findings of the ultrasound done at 40 weeks documented a live foetus in cephalic presentation with an increased amniotic fluid volume. The estimated foetal weight was 2.4 kg with no gross anomaly seen. Umbilical artery Doppler was also normal.

In addition to this, a well circumscribed heterogeneous mass was found adjacent to the placenta. It demonstrated low velocity non-pulsatile flow seen around the central echogenic foci.

The initial differential diagnoses based on a history of a singleton pregnancy included a placental chorioangioma or teratoma. Based on the ultrasound features described, an impression of TRAP sequence was still considered. The patient was counselled and she opted for a Caesarean section. A live male infant was delivered weighing 2.5 kg. Apgar scores were 8 and 9 at 1 and 5 mins respectively.

An amorphous mass weighing 1.5kg, with vestigial limbs was removed following the delivery of first baby. It had no other distinct foetal features apart from hair on the presumed cranial region. There was a two-vessel umbilical cord that had a velamentous cord insertion onto the monochorionic placenta. There was no dividing

membrane. Based on these unique findings, a diagnosis of TRAP sequence was confirmed. **(**Figures 1-2)

The new-born was evaluated by the paediatric team and found to be grossly normal. Further investigations of the pump twin were declined by the parents. An autopsy of the acardiac twin was also declined.

Both mother and pump baby were discharged on the third post-operative day having had an uneventful post-operative period. They continued to be well and healthy at both postnatal visits at 2 and 6 weeks respectively.

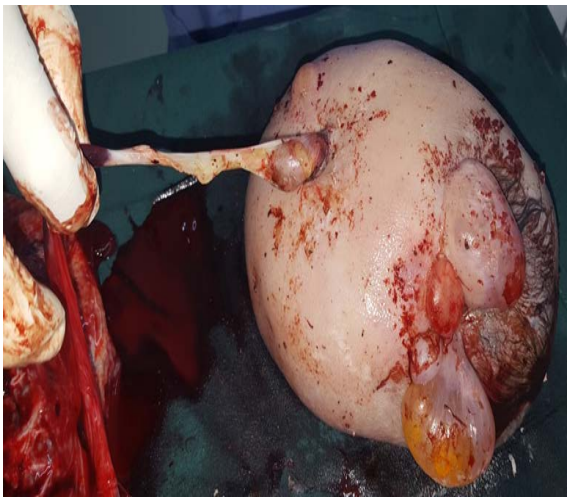

Fig 1 Anterior view of amorphous acardiac twin showing its two-vessel cord

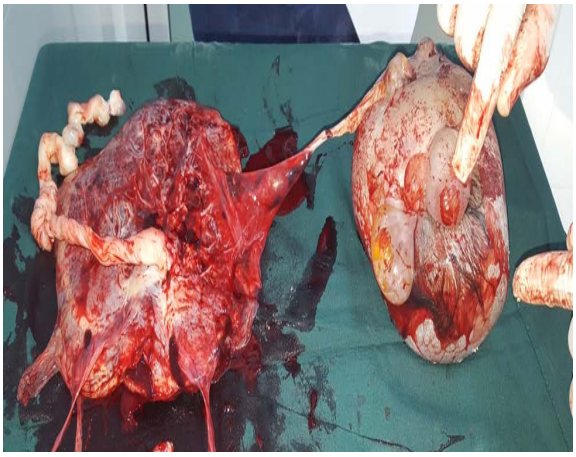

Fig 2 Acardiac twin with monochorionic placenta

Case 2

The baby demonstrated normal developmental milestones during the 6-month follow-up period.

Case 2

The second patient was a 33-year-old primiparous who was referred to our hospital at 28 weeks with a provisional diagnosis of twin gestation with intrauterine foetal demise of one twin made during routine antenatal care. Her antenatal period had been uneventful before this finding.

During the repeat ultrasound scan at our facility, a diamniotic twin gestation was found with a single anterior placenta associated with a very thin dividing membrane (2mm thick), suggestive of monochorionicity. Twin A was a live foetus in transverse lie with an estimated foetal weight of 1.26kg. No gross foetal abnormalities were seen. An umbilical artery Doppler showed a normal waveform. There was however, marked polyhydramnios and funnelling of the cervix.

Twin B presented as an acephalic mass with poorly formed thorax and abdomen. It also had rudimentary asymmetrical limbs with unilateral talipes. There was active movement of the lower limbs despite the absence of cardiac activity in the thorax. Blood flow was demonstrated in the umbilical vessels. **(Figures 3-4)**

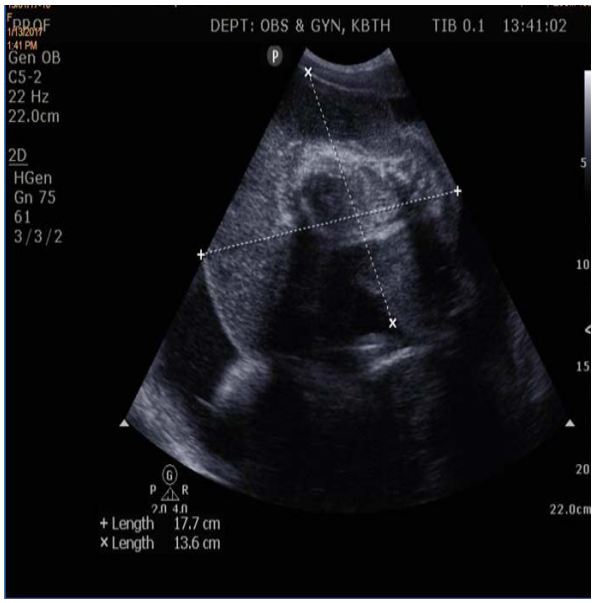

Fig 3 Ultrasound image of acardiac twin showing its dimensions

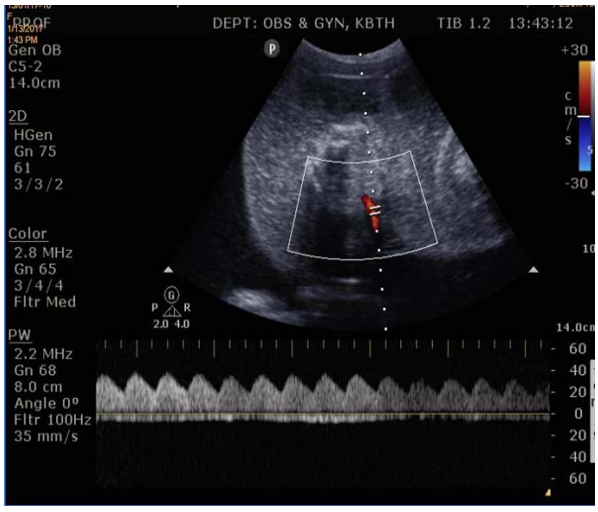

Fig 4 Umbilical Doppler ultrasound of acardiac twin

A diagnosis of TRAP sequence was made and the parents were duly informed and counselled. The patient was admitted and antenatal dexamethasone was administered to aid in lung maturation. She had an amnio-reduction done a week after corticosteroid therapy. Samples of amniotic fluid were taken for karyotyping and a shake test was performed to determine foetal lung maturity. It was negative. She was given indomethacin as tocolysis post-procedure.

Conservative management was continued with twice weekly foetal ultrasound monitoring until she went into spontaneous preterm labour at 31 weeks' gestation. She had an emergency Caesarean section, the findings of which were a live female in breech presentation with a birth weight of 1.26 kg and Apgars of 6 and 7 at 1 and 5 minutes respectively. The baby was morphologically normal. A 3.0kg acephalic mass with rudimentary lower limbs and torso was removed following the delivery of the first twin. **(**Figure 5)

Fig 5 Acephalic acardiac twin

The first twin was admitted to the neonatal intensive care unit (NICU) and managed for prematurity. Investigations done on this baby were normal. The mother was discharged on the third post-operative day and remained well at both her postnatal visits. The baby was discharged from NICU after 5 weeks and remained well with appropriate development during the 6-month follow-up.

Discussion

TRAP Sequence is a rare diagnosis that is considered to be a severe variant of Twin-to-Twin

Transfusion Syndrome (TTTS)[5]. The condition is characterized by a malformed twin which lacks a well-formed cardiac system and so is completely dependent on the other twin for its perfusion. The pump twin is often structurally normal although in 9% of cases, may also have congenital abnormalities[2].

It is called a reversed arterial perfusion sequence because blood from the pump twin flows through the umbilical artery to the recipient or acardiac twin through arterial-arterial anastomosis on the placental surface. The acardiac twin has no direct communication with the placenta. As a result, the acardiac twin is perfused with deoxygenated blood which enters the iliac vessels leading to discordant caudo-cranial development.[6] In addition, the pump twin receives further deoxygenated blood from the acardiac twin through vein-vein anastomosis leading to chronic hypoxia and growth restriction.

Depending on its morphological appearance, the acardiac twin may be classified into four different types[6].

• Acardius acephalus: cranial and thoracic structures are absent but lower limbs are present and distinguishable. It accounts for 60–75% of cases

• Acardius amorphus: no recognizable foetal shape. Accounts for approximately 20% of cases

• Acardius anceps: areas of head, thorax and abdomen are present but are poorly developed.

Seen in approximately 10% of cases

• Acardius acormus: only the head is present. It is quite rare and occurs in approximately 5% of cases.

This case series describes two different types of acardiac twins: the amorphous type in case 1 and the acephalus type in case[2].

As a consequence of these anomalous vascular connections, the pump twin may develop high-output congestive cardiac failure, polyhydramnios leading to preterm labour (and the attendant complications of prematurity) and anaemia. The mortality rate in the acardiac twin is 100% and varies from 50-70% in the pump twin[7]. The prognosis is primarily influenced by the weight of the acardiac twin in relation to the pump twin, with worse outcomes associated with weight differences greater than 70%[8].

Estimating the weight of the acardiac twin is not possible using the standard formulas. We attempted to calculate the weight of the acardiac twin in both cases using the weight regression formula proposed by Moore and Gales[8] and the prolate ellipsoid formula put forward by Oliver et al[9]. We however found a huge disparity in the weight estimation and actual weights in both instances using both formulas. For example, the estimated weight of the acardiac twin in case 2 was 345g (giving a weight ratio of 27%) and 1100g (91%) using the Moore et al and Oliver et al formulas respectively [8,9]. The actual weight of the mass was 3kg. The results of these studies were based on much earlier gestational ages (average gestational ages of 29 and 19 weeks respectively) and so may not be applicable in advanced gestation as in our case series.

In addition, rapid growth of the acardiac twin and the development of hydrops, heart failure, polyhydramnios or abnormal Doppler waveforms in the pump twin are also poor prognostic factors. Although there was evidence of polyhydramnios in both our case

reports, neither of the pump twins showed any other signs of heart failure. Furthermore, the pump twin in Case 1 survived to term. This may be due to the fact that the Acardius amorphous type (described in our first case) has a better prognosis for the survival of the pump twin[10].

Prenatal diagnosis is feasible as early as 11-13 weeks with ultrasound. The diagnosis hinges on the recognition of a normally-appearing foetus with an abnormally-formed co-twin. Placentation is monochorionic, demonstrated by a single placental mass without the twin-peak (lambda) sign. Doppler ultrasound of the acardiac twin shows a reversal of flow[11]. This helps distinguish TRAP sequence from intrauterine foetal demise of a normal co-twin. However, in mono-amniotic pregnancies, the acardiac twin can be easily mistaken for a yolk sac as may have happened in the first case discussed.

In our first case report, the diagnosis was missed during her initial ultrasound done at 10 weeks and subsequently, she had no further sonographic evaluation of her pregnancy. A foetal anomaly scan between 18 and 22 weeks would likely have aided in early diagnosis and management. Had the diagnosis been made earlier, the patient could have then been adequately counselled and offered the options of continuous foetal surveillance or an elective termination based on the prognostic indicators present in the pump twin.

Early detection and assessment of the severity of complications in the pump twin can improve perinatal outcomes. In case 2, the diagnosis was made at 28 weeks. This afforded us the opportunity to provide her with the options of expectant management according to the recommended guidelines with consideration for local limitations.

Expectant management for pregnancies complicated by TRAP sequence involves increased foetal surveillance and foetal intervention[6, 8]. This includes weekly sonographic and echocardiographic surveillance of the pump twin with careful attention paid to its cardiac function[1]. In the event of any evidence of cardiac failure or hydrops such as atrial or ventricular enlargement, early delivery is necessary to improve survival outcomes in the twin.

Antenatal corticosteroid administration between 24-34 weeks is recommended in all patients with pregnancies complicated by TRAP sequence to aid lung maturation[8]. This recommendation was followed in our second case.

In addition, the option of serial reduction amniocentesis or the use of indomethacin may be offered to prevent preterm labour (and the attendant risks associated with prematurity) which may ensue from polyhdramnios[12]. Other authors recommend the use of digoxin to improve cardiac function in the pump twin[13]. The patient in our second case report had amnio-drainage of 200mls of amniotic fluid. Post-procedure, she had tocolysis with indomethacin. Despite these interventions, however, she went into preterm labour at 31 weeks.

The advent of surgical interventions such as laser or radiofrequency ablation techniques to correct the anomalous vascular connection has significantly aided in improving perinatal outcomes. These interventions are most effective if offered before 20-24 weeks[14]. This further emphasizes the importance of early prenatal diagnosis. In settings where prenatal diagnosis is available, it is important to karyotype the normal co-twin before instituting these interventions as up to about 9% have congenital anomalies[2].

Delivery is recommended at 34-36 weeks in a tertiary hospital with a paediatric cardiologist or neonatologist on hand for the delivery[1]. Vaginal delivery is the preferred route and Caesarean section is indicated for obstetric reasons. Both patients in our case series had Caesarean sections. The first patient was offered a Caesarean section due to the uncertainty of the diagnosis and the second due to preterm labour and malpresentation.

Conclusion:

TRAP sequence is a severe complication of monochorionic twin gestation with mortality rates ranging between 50-70% in the untreated pump twin. Prenatal diagnosis is feasible using ultrasound and every effort should be made to recognize the condition early enough to optimize the survival of the pump twin. This is possible even in low resource settings and can result in good outcomes.

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