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Case Report

Conservative management of a case of a cardiac aniceps twin reversed arterial perfusion sequence misdiagnosed as vanishing twin: a case report

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ABSTRACT

Twin reversed arterial perfusion (TRAP) sequence is a specific, rare and severe complication of monochorionic multiple pregnancy, which is characterized by one normal fetus (pump twin) and another with no cardiac activity and variable degree of deficient development of the head and upper limbs. The management of these pregnancies are a real challenge due to the parasitic hemodynamic dependence of the acardiac twin on the pump twin. The aim of management is to maximize the chances of survival of the pump twin with some intervention. The preferred management suggested is elective ultrasound-guided laser coagulation or radiofrequency ablation of the umbilical cord vessels at 11-13 weeks when the survival is 70-75%. Delay in intervention until 16-18 weeks is associated with spontaneous cessation of blood flow in the acardiac twin in 60% of cases and in about 50% of these there is death or brain damage in the pump twin. However, pregnancies diagnosed late need to follow a tailored approach with expectant management. The authors report a case of TRAP twin sequence which on ultrasound was diagnosed as a case of vanishing twin at 20 weeks. Subsequently on follow up ultrasound, it was diagnosed as a trap sequence with the pump twin developing polyhydramnios at 30 weeks. She was counselled regarding the prognosis of the pump twin and she opted for conservative management. Weekly ultrasound and color doppler were done. The amniotic fluid index (AFI) decreased gradually at 33 and 35 weeks, there were no signs of congestive cardiac failure of the pump twin on Doppler studies and she had a spontaneous delivery of a healthy male child and an acardiac aniceps fetus at 36 weeks and 5 days. The perinatal mortality of a pump twin managed conservatively ranges from 35 to 55%. It is essential to diagnose the presence of trap sequence at an early gestational age through improved imaging techniques to plan a timely and effective intervention to salvage most of the pump twins.

Keywords: Trap sequence, Acardiac-twin, Pump twin, Radiofrequency ablation

INTRODUCTION

A twin reversed arterial perfusion (TRAP) sequence is a rare phenomenon occurring in 1% of monochorionic twin pregnancies. The first such case was described by Benedetti in 1533.¹ In 1978 the first description of prenatal diagnosis of an acardiac twin was reported by Lehr and Dire.² The incidence of this condition has been estimated at 1:35,000 pregnancies. However, Van Gemert et al found that in recent years due to better ultrasound diagnosis and the frequent use of assisted reproductive

technologies, the incidence of TRAP has grown to 2.6% of monozygotic twins and 1 in every 9,500-11,000 pregnancies.³ Monochorionic (MC) twin pregnancies show varying degrees of unequal placental share, and most have inter-twin placental vascular anastomosis resulting in communication between the two fetoplacental circulations. This results in inter-twin discordance in fetal size, amniotic fluid volume, structural defects as well as fetoplacental hemodynamics. Twin reversal arterial perfusion is the most severe complication of monochorionic twin pregnancy and occurs when there is

one normal fetus (pump twin) and another fetus with no cardiac activity and variable degrees of deficient development of head limbs and other body parts. Color Doppler in the recipient twin shows reversed pulsatile flow from an umbilical arterio-arterial anastomosis and venous return to the pump twin by a veno-venous anastomosis. The mortality rates of the pump twin have been reported to be as high as 50-70% due to congestive heart failure, severe preterm birth and polyhydramnios, if the trap sequence is not detected early, followed up and timely treated.^{4,5}

CASE REPORT

A 22-year-old gravida-2, para-1 with one live baby attended the labour ward at 36 weeks 6 days with labour pains. This was a spontaneous conception. She was diagnosed as a case of monochorionic diamniotic twin pregnancy with one normal live twin with a normal amniotic fluid index and the other as a vanishing twin at 21 weeks of gestation (Figure 1). The vanishing twin was deformed and had no cardiac activity (Figure 2). She presented to our antenatal clinic at 30 weeks 2 days with polyhydramnios with an AFI of 44 cm. On further examination by ultrasound, the vanishing twin had showed some growth and Doppler revealed blood flow towards the acardiac fetus. The fetus had an incompletely formed skeleton, with no upper limbs, heart or thorax (Figure 2, 3). A diagnosis of trap twin sequence was made and the patient was counselled regarding its prognosis. She did not want to be referred for fetal therapy and wanted to be followed up conservatively. She underwent weekly ultrasound along with Doppler to assess the wellbeing of the pump twin. The AFI of pump twin decreased to 20.9 cm at 33 weeks. At 35 weeks AFI decreased to 18 cm. All other parameters were normal on color doppler. She had a spontaneous labour at 36+6 weeks. and delivered a healthy male child of 2.43 kg followed by the delivery of the acardiac twin which appeared to be male, had rudimentary upper limbs, well developed but deformed lower limbs, a deformed head and absent thorax (Figure 4). Grossly the placenta was single with two umbilical cords. The normal twin's cord was long and that of the acardiac twin short.

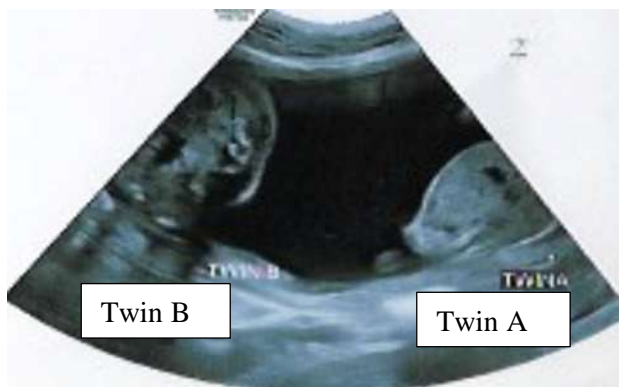


Figure 1: Twin B diagnosed initially as vanishing twin with normal twin A.

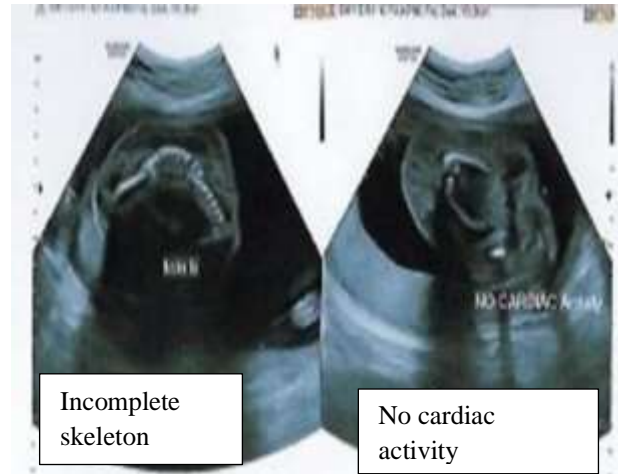


Figure 2: Twin B had an incomplete skeleton with no cardiac activity on USG.



Figure 3: Twin B deformed twin B with no proper head, thorax or upper limbs.

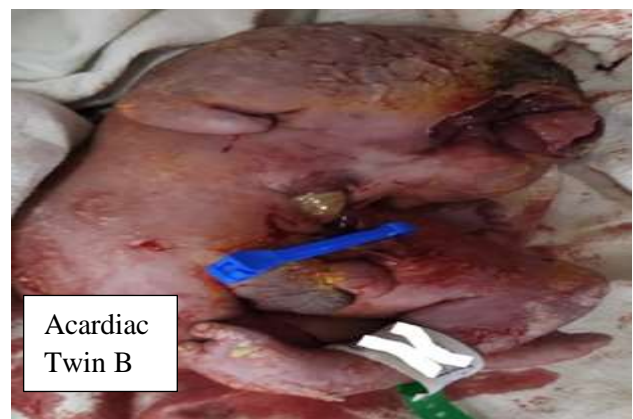


Figure 4: Acardius anceps fetus with developed but deformed lower limbs, partially formed head and face.

DISCUSSION

The etiology and exact pathogenesis of TRAP sequence is still debated. However, two pathways have been

proposed. In early embryogenesis, aberrant vascular pattern causes malformation of the acardiac twin.⁶ Blood pressure imbalance leads to de-oxygenated blood transfer from healthy fetus (donor) to the impaired twin via the umbilical cord. The poorly oxygenated blood maintains a certain growth of near-by located structures like the lower limbs, whereas the supply to the head, upper extremities and thorax is insufficient and there is arrest in development of these structures.^{7,8} Due to some chromosomal abnormality or environmental factors, there is a failure of heart formation and the perfusion support for the acardiac foetus is received through anastomosis between the umbilical vessels.

The prenatal diagnosis of TRAP-sequence is feasible by ultrasound and can be established during the first-trimester screening.^{9,10} The typical ultrasound features are: gross differences in biometrical measurement of twins, particularly the abdominal circumference; absence of a morphologically normal heart in one twin associated with several other malformations in head, trunk, upper and lower extremities; presence of subcutaneous oedema and cystic hygroma in the anomalous twin. A pathognomonic finding is the demonstration of a paradoxical circulation in the acardiac twin, with arterial blood flowing towards, rather than away at Colour Doppler. Intrauterine death of an abnormal monochorionic twin could resemble an acardiac foetus, but maintained growth at ultrasound follow-up shows the correct diagnosis. Persistent intra-fetal blood flow signals on colour Doppler should raise the possibility of a TRAP sequence.

The diagnosis is not always simple. The most frequent confusion is with anencephaly or with fetal demise of one twin. It is easy to erroneously diagnose fetal death in these cases because of absence of cardiac motion and movement in the acardiac twin as happened in our case which was initially reported as vanishing twin. The risks involved, without any intervention, are intrauterine death (25%), Polyhydramnios (50%) and premature birth (80%). The survival rate without any intervention is approximately 55%.^{11,12} The signs of cardiac failure in the pump twin are hydramnios, cardiomegaly, ascites and tricuspid regurgitation.¹³

Based on the morphology of acardiac foetus, four distinct types have been described.¹¹

Acardias acephalus

Total 60-75% of cases, most common type, with well-developed pelvis and lower limbs, but no head, usually no thoracic organs and most often no arms.

Acardius anceps

Total 10% of cases, is the most differentiated type of acardiac twins with well-developed body and extremities, but only a partially formed head and face.

Acardius acornus

Total 5% of cases, only cephalic structures are detectable with a close umbilical cord insertion.

Acardius amorphous

Total 20% of cases, foetus is represented by a shapeless mass of tissue containing no recognizable structures.

This classification only provides a morphological description, has no prognostic value and does not provide information on management options. Some factors that have been identified as markers of pump-twin poor prognosis is congestive heart failure identified by hydrops or polyhydramnios (biggest pocket > 8 cm), delivery before 32 weeks, a big acardius twin defined as acardiac-to-pump twin weight ratio > 70%, discrepancy in pump/acardiac umbilical venous diameter (UVD) ratio and the presence of a well-developed body and upper extremities in the acardiac twin.

The exact weight of the acardiac twin cannot be calculated using the standard formulas based on ultrasound biometry, because of the lack of proper anatomical structures; the following formula has been proposed:

Weight (g) = $1.2 \times (\text{longest length in cm})^2 - (1.7 \times \text{longest length in cm})$.

Wong et al focused on the use of abdominal circumference in evaluating the acardiac foetus size and proposed a role of abdominal circumference ratio as a prognostic factor.¹⁴ If the ratio of abdominal circumference of acardiac twin to normal twin is ≥ 1.0 , it is considered significant.

There are controversies regarding elective versus therapeutic treatment of twin reversed arterial perfusion sequence. Lewi et al found spontaneous flow arrest in 21% of cases during 16-18 weeks. Pagani et al reported a case series in which treatment was given after onset of poor prognostic factors in normal twin.¹⁵ The outcome was good as compared to those who had conservative management; hence they suggested elective intervention due to low sensitivity of prognostic factors to predict intrauterine demise of pump twin.

Over the last two decades, there have been several procedures aimed at establishing a permanent separation of the two blood-circuits of the twins thereby improving the prognosis in the pump foetus. These procedures partly target the umbilical cord vessels using cord occlusion by coil embolization, ligation under fetoscopy, laser coagulation or bipolar coagulation under ultrasound guidance. Using fetoscopy for cord ligation or laser coagulation has a technical failure rate of 10%, increased risk of PPRM, intraamniotic infection and bleeding in pump twin.

The intra-fetal ablation approach has the goal of ablating the pelvic vessels or the abdominal aorta of the acardiac twin. This procedure is not influenced by placental location, umbilical cord structure, amniotic fluid volume and position of the acardius. The intra-fetal ablation methods are done by using monopolar diathermy, interstitial laser or radiofrequency ablation. In recent years, endoscopic procedures have been abandoned because of their invasiveness and both intra-fetal laser and Radiofrequency ablation are the most commonly used techniques

When these methods are used at 16-18 weeks' gestation survival of pump twin is about 80%. However, delay in intervention between the diagnosis of TRAP sequence at 11-13 weeks of gestation until 16-18 weeks is associated with spontaneous cessation of flow in the acardiac twin in 60% of cases and in about 50% of these there is also death or brain damage in pump twin. The preferred management is ultrasound-guided laser coagulation or radiofrequency of the umbilical cord vessels within the abdomen of acardiac twin at 11-13 weeks' gestation. The survival is 70-75% which is less than 80% achieved with intervention at 16-18 weeks.

In our case, the acardiac twin was misdiagnosed as vanishing twin at 20 weeks. However, on follow-up the pump twin developed polyhydramnios with growth of vanishing twin following which acardiac twin diagnosis was made on Doppler studies. The patient was managed by weekly ultrasound and colour Doppler. Liquor volume decreased gradually over the next few weeks, the pump twin did not develop features of cardiac failure and the patient delivered a healthy baby near term.

CONCLUSION

TRAP sequence is an extremely rare congenital anomaly, where antenatal diagnosis is feasible and can be established during the first-trimester screening. However not all patients seek antenatal care in first trimester. Diagnosis is not often simple and confusion with anencephaly and fetal demise of one twin exists. Follow up of vanishing twin must be done with Doppler to look for acardiac twin in monochorionic twin pregnancies. Antenatal ultrasounds should be evaluated in detail if there is a presence of any abnormal mass along with a normal fetus. These foetuses are candidates for early intervention and should be referred to a fetal medicine centre.

Though patients who present late can benefit from conservative treatment, the latest studies however encourage intervention in the first trimester of pregnancy at 11-13 weeks. The technique of choice to interrupt the vascular supply to the acardiac twin is by ultrasound-guided laser coagulation or radiofrequency ablation of the intra-fetal vessels.

Although literature suggest the advantages of early intra-fetal laser treatment in trap sequence, our case shows that pregnancies referred late would still require a tailored approach with close ultrasound monitoring for a successful outcome. Many such pregnancies still do not have access to these advanced procedures and need to be counselled regarding the prognosis.

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