

MISDIAGNOSIS OF AN ACARDIAC TWIN IN THE FIRST TRIMESTER: A CASE REPORT

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ABSTRACT Background: Twin reversed arterial perfusion (TRAP) sequence is a rare monochorionic twin pregnancy complication. It should be suspected in the first-trimester ultrasound when one fetus has no cardiac activity in monochorionic pregnancies. Doppler study is essential for early diagnosis of acardiac twin. This manuscript shows the importance of reassessing cases considered as vanishing twins in early pregnancy, to avoid a late diagnosis of acardiac twin, without timely treatment. **Case Summary:** In this case, the assumption of the death of one fetus in the first trimester, delayed the correct diagnosis of TRAP until 25 weeks, when the case was referred to our unit. We opted conservative management with weekly ultrasound surveillance, looking for features of heart failure in the pump twin. After suspecting fetal anaemia, at 32 weeks a healthy baby weighing 2100g was delivered along with an acardiac aniceps with 518g. This is a case of expectant management, for lack of another option, to a successful end. **Conclusion:** Although twin reversed arterial perfusion (TRAP) sequence being a rare monochorionic twin pregnancies complication, it should be suspected in the first-trimester ultrasound, when detected monochorionic pregnancies and one embryo/fetus has no cardiac activity. Follow up in the fetal death of one twin in monochorionic pregnancies must be done with Doppler to look for an acardiac twin. Because pump twin may develop a high-output cardiac failure and intrauterine fetal demise, for continuing pregnancies without poor prognostic criteria, it is important to maintain weekly ultrasound surveillance, with attention to the intervention criteria.

KEYWORDS TRAP sequence, acardiac fetus

Introduction

TRAP sequence is a rare and exclusive complication of monochorionic twin pregnancies, with recent data on the incidence of 2.6% in these pregnancies.[1] It is characterised by a fetus that lacks cardiac structure and activity (acardiac twin), that is perfused and hemodynamically dependent (parasite) on the structurally normal co-twin (pump twin). This anomaly occurs

through an artery-to-artery placental anastomosis, creating a “reversed” circulation, which provides perfusion of mixed or medium oxygenated blood from the pump twin to the recipient twin. It results in poor perfusion and tissue necrosis, with the evolution of a variety of structural abnormalities in the acardiac fetus.

The heart of the acardiac fetus may be completely absent (holo-acardius) or represented by a fundamental cardiac structure (pseudo-acardius). Four distinct morphological types have been described in literature: acardius acephalous: is the most common type (60-75%), with developed pelvis and lower limbs, may have arms, but thoracic organs and head are absent; acardius aniceps: the most differentiated type of acardiac twins with established body and extremities, but only a partially formed head and face (20%); acardius acornus: in which only cephalic structures were detectable, is a sporadic type (10%); acardius amorphous: consists of a shapeless mass of tissue containing no recognizable human structures (5%).[2]

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CASE REPORT:

A 37-year-old, caucasian, spontaneous primigravida was referred to our prenatal unit at 25 weeks when twin reversed arterial perfusion (TRAP) sequence was diagnosed.

Early in the pregnancy she had performed the first ultrasound at ten weeks and five days: twin pregnancy was identified, with a second embryo without cardiac activity. At the 12 week ultrasound, one fetus had a crown rump length (CRL) of 61.3mm, described with normal morphology, 1.6mm of nuchal translucency and low risk in the combined screening test. At this time, the second fetus was not described, assuming a vanishing twin.

The morphological ultrasound at 20 weeks identified a normal female fetus and a heterogeneous mass of 71x24mm, with suggestive bone tissue, considering possible resorption of the second fetus.

At the time she arrived at our prenatal unit (25 weeks), was described as a monochorionic monoamniotic twin pregnancy, with one structurally normal fetus and a second twin anatomically abnormal and acardiac. The first twin presented a fetal estimation weight (EFW) of 846g, in the 50th percentile, normal fetal Doppler study and normal amniotic fluid index. The acardiac mass continued to grow (98x53x57mm), and TRAP sequence was diagnosed by Doppler study, which identified interstitial blood flow in the acardiac mass (figure 1).

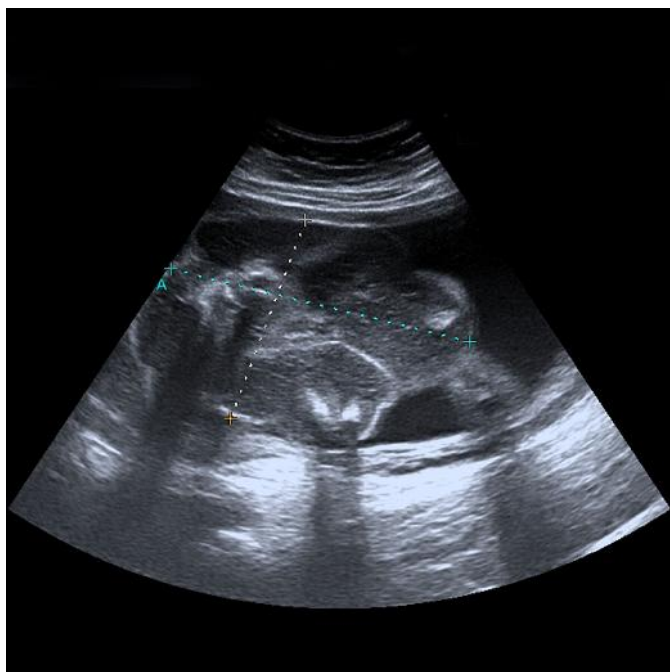


Figure 1: Ultrasound of the acardiac fetus at 28 weeks, measuring 130x98x67mm.

Weekly ultrasound was performed with standard Doppler study and no signs of cardiac failure.

At 29 weeks, the peak systolic velocity (PSV) in the middle cerebral artery (MCA) of the healthy fetus reached 1.54 multiples of the median (MoM), with no signs of hemodynamic decompensation. Due to suspicion of fetal anaemia, a course of antenatal corticosteroids for fetal lung maturation was administered. Since then ultrasound surveillance started to be performed twice a week. The PSV in the MCA maintained under 1.5 MoM until 32 weeks and five days, reaching 1.69 MoM. The remaining Doppler studies were normal. The pump twin

continued EFW in the 50th percentile and at breech presentation. No signs of high-output cardiac failure like polyhydramnios, cardiomegaly, ascites and fetal hydrops were found. The acardiac mass was 176x90x55mm, less than 25% of the healthy twin estimated weight (2073g). After inconclusive neurosonogram, a fetal magnetic resonance imaging (MRI) evaluated the natural brain development of the first twin, at 29 and 32 weeks.

Due to suspected fetal anemia, at 32 weeks and 5 days, she underwent caesarean section. Delivered a typical female live baby, weighting 2100g, Apgar score of 8 / 9 at one and five minutes respectively; and another female acardiac anencephalic: with 518g. The acardiac fetus had a rudimentary structure: incompletely formed skeleton, edematous lower limbs, shortened upper limbs, abdomen with omphalocele and umbilical cord with two vessels, remnants of the chest and head structures (Figures 2 and 3).



Figure 2: Acardiac fetus delivered.

The healthy baby had mild anaemia (hemoglobin: 13.4 g/dl, haematocrit 38.9%), without the need for transfusion. She was admitted to the intensive/intermediate care unit, with a diagnosis of prematurity, hyperbilirubinemia and intra-periventricular haemorrhage grade I and discharged on the 17th day of life, clinically and neurologically well. Maintains neurodevelopment surveillance in our hospital centre.

DISCUSSION

Advances in first-trimester ultrasound have enabled diagnosis of TRAP sequence at earlier gestational age. TRAP should be suspected, when a monochorionic pregnancy is diagnosed with one fetus appearing anatomically normal, and the other lacks cardiac structures and activity. The confirmation is made with Doppler study: pulsatile flow in the umbilical artery towards the acardiac fetus or interstitial blood flow within the mass and supported by the observation of continuous growth of the acardiac twin in serial ultrasound examinations.[3] It may simulate an intrauterine death of one grossly abnormal monochorionic twin or a placental teratoma. In the described case, the first ultrasound identifies the second embryo without cardiac activity, and

Table 1 Timeline of interventions and outcomes.

From Obstetric Ultrasound to Delivery								
Out of our unit			In our prenatal unit					
10w	12w	20w	25w	Once a week	29w	Twice a week	32w	32w
Two embryos one without cardiac activity.	One fetus with normal morphology and low risk in the combined screening test.	One female fetus, normal morphology and a heterogeneous mass of 71x24mm.	Monochorionic monoamniotic twin, one structurally normal fetus (EFW - 846g) and a second twin (98x53x57mm) anatomically abnormal and acardiac. Doppler study identified interstitial blood flow in the acardiac mass.	Ultrasound + Doppler study	Normal fetus: PSV in the MCA = 1.54 MoM. No signs of hemodynamic decompensation. Fetal MRI: normal brain development of the pump twin.	Ultrasound + Doppler study	Healthy fetus: EFW in the 50 th percentile and at breech presentation. PSV in the MCA = 1.69 MoM. No signs of high-output cardiac failure. Acardiac mass: 176x90x55mm, less than 25% of the standard twin estimated weight (2073g). Fetal MRI: normal brain development of the pump twin.	Caesarean delivery because of fetal breech presentation. Healthy female live baby, 2100g AS-8/9. Mild anaemia (hemoglobin: 13.4 g/dl, haematocrit 38.9%), without the need for transfusion. Discharged on the 17th day of life, clinically and neurologically well.
Vanishing twin?		Resorption of the second fetus?	TRAP sequence.		Suspicion of fetal anaemia, a course of antenatal corticosteroids for fetal lung maturation was administered.		Suspicion of fetal anaemia. Delivery decision.	Female acardiac aniceps, with 518g.
AS - Apgar score; EFW - fetal estimation weight; MCA - middle cerebral artery; MRI - magnetic resonance imaging; MoM - multiples of median; PSV - peak systolic velocity; TRAP - twin reversed arterial perfusion; W- Weeks;								



Figure 3: X-ray of Acardiac fetus.

a vanishing twin was erroneously assumed in the first trimester.

The pump twin may develop a high-output cardiac failure and intrauterine fetal demise. This high cardiac output also increases perfusion of the fetal kidneys, resulting in overproduction of fetal urine and polyhydramnios, at risk of premature delivery.[2] The perinatal mortality rate for pump twin without treatment is high, around 55%, due to heart failure and preterm delivery.[4] After in utero therapy, the survival rate of the pump twin increases to 80-90%.

Indicators of poor prognosis have been described. A higher weight of the recipient twin is more likely associated with cardiac insufficiency and perinatal mortality of the pump twin.[2]

When the ratio of the weight of the acardiac twin to the weight of the pump twin is greater than 70%, the risk of preterm delivery is 90%, and of congestive heart failure in the pump twin is 30%. In comparison, when the ratio is less than 70% the corresponding risks are 75% and 10%, respectively. Estimation of acardiac fetus weight can be calculated using the following formula: Weight (grams) = $(1.2 \times \text{longest length } 2) - (1.7 \times \text{most extended lengths})$. [4] The alternative prognostic factor is the ratio of the abdominal circumference of acardiac twin to standard twin when ≥ 1.0 is considered significant.[5]

After diagnosis, the aim of management is to save the pump twin, as mortality is 100% for an acardiac twin. Exclusion of genetic abnormalities (determination of the karyotype) and ultrasound malformations of the pump twin, is advised before the management decision. It has been reported that the pump twin has a 9% risk of aneuploidy. To improve the outcome for the pump twin, at viable gestational age, fetal monitoring should be done with weekly ultrasound and Doppler fetal study. If there is evidence of pre-hydrops signs, the frequency should be twice a week. Fetal surveillance with Doppler studies of middle cerebral artery help in early diagnosis of anaemia in pump twin.[3,6] Fetal neurosonogram should be performed to evaluate brain development of the pump twin if cerebral lesions are suspected a fetal MRI is advised.

The correct and appropriate management is not yet estab-

lished. Antenatal intervention, delivery and expectant management are possible options. It is controversial regarding elective versus treatment of TRAP. Some support elective intervention due to improved outcomes after therapy and low sensitivity of prognosis factors to predict intrauterine fetal death.[7] It seems that the results are better if therapy is done before 16 weeks. Others support expectant management with treatment when sonographic evidence of compromise is present in the pump twin (polyhydramnios, cardiac dysfunction, abnormal Doppler and hydrops), avoiding an intervention that itself can cause miscarriage.[2]

Current, minimally invasive in utero interventions, to occlude vascular supply to the acardiac twin, can be performed through two modalities: extra-fetal (intertwine anastomosis or umbilical cord occlusion) and intra-fetal circulation. Occlusion of the umbilical cord of the acardiac twin can be performed through a fetoscope by laser or bipolar coagulation. The intra-fetal intervention modality is performed by radiofrequency ablation (RFA) or laser therapy, through a needle inserted under ultrasound guidance. RFA coagulates the abdominal wall of the acardiac twin, at the base of the umbilical cord, being preferred at gestational ages greater than 16 weeks. Laser needle is inserted towards fetal abdomen and the target fetal vessels, being preferred until 16 weeks.[3,8]

Current small evidence, suggests ultrasound guided intra-fetal techniques are associated with higher success rates and lower postoperative complications like preterm premature rupture of membranes and preterm labour. Laser and RFA are relatively comparable, with neonatal survival rates of 82% and 85%, respectively.[7,9] The choice of treatment should be based on operator experience, gestational age and accessibility.[3]

The ideal timing of in utero intervention remains controversial; procedures have been reported between 12 and 27 weeks of pregnancy, is usually performed at the beginning of the second trimester (16 weeks). Knowing that the death of the pump twin occurs mostly until 16 weeks, preventive intervention at 12 weeks may be more advantageous. Larger studies are necessary to examine the best timing for preventive intervention with the higher pump twin survival.[2]

In our case, the patient was diagnosed late in pregnancy (25 weeks), at the time she was referred to us we adopted an expectant management with weekly ultrasound surveillance, because of the higher risk of preterm delivery after treatment in late gestational age. No polyhydramnios, signs of cardiac failure or fetal hydrops in the pump twin were identified. The acardiac twin continued to grow but with an estimated weight proportion of less than 25%. Rohilla et al. defends conservative treatment when the acardiac twin is less than 25% the weight of the pump twin and no signs of heart failure. When the weight of the acardiac twin exceeds 70% of the pump twin, invasive intervention is justified.[10]

The timing of delivery depends on clinical evolution and gestational age. TRAP sequence without poor prognostic criteria should deliver at 34-36 weeks of gestation. Caesarean delivery is indicated for general obstetrical indications.[3]

When anaemia of the pump twin was suspected in this case, by Doppler study of middle cerebral artery, a course of antenatal steroids was administered. The sign of fetal anaemia was our criteria to interrupt pregnancy after fetal pulmonary maturation. We delivered a healthy baby at 32 weeks and five days by caesarean section, for breech presentation.

Our case is an example of why ultrasound should be per-

formed by an experienced sonographer to avoid failure in early diagnosis of TRAP. In this sequence of late diagnosis, the approach of expectant management with tight surveillance to decide when to intervene or deliver allows good outcomes.

LEARNING POINTS/TAKE HOME MESSAGES

- Although twin reversed arterial perfusion (TRAP) sequence being a rare monochorionic twin pregnancies complication, it should be suspected in the first-trimester ultrasound, when detected monochorionic pregnancies and one embryo/fetus has no cardiac activity.
- Follow up in the fetal death of one twin in monochorionic pregnancies must be done with Doppler to look for an acardiac twin.
- The appropriate management is not established: elective treatment and expectant management with treatment or delivery, are options. Early in utero therapy reduces the perinatal mortality rate. Intra-fetal intervention with laser or radio-frequency ablation is the preferred modality.
- Because pump twin may develop a high-output cardiac failure and intrauterine fetal demise, for continuing pregnancies without poor prognostic criteria, it is important to maintain weekly ultrasound surveillance.
- Criteria for intervention should be: Onset of Cardiac failure, hydrops of the pump twin or polyhydramnios and acardiac/pump twin weight ratio > 70%

Authors' Statements

Competing Interests

The authors declare no conflict of interest.

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