Accepted Manuscript online: 2024-08-12 Article published online: 2024-09-10

Rare Case of Twin Reversed Arterial Perfusion *Acardius Acephalus* Subtype Seen in a Community-Based Hospital

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AJP Rep 2024;14:e224–e227.

Abstract

Background TRAP sequence occurs in monochorionic pregnancies consisting of one normal fetus and a non-viable fetus. The pump twin has an increased risk of developing high-output cardiac failure.

Case 32-year-old G4P2012 with TRAP syndrome in current pregnancy presented to triage at 26 weeks with contractions and spotting. She had undergone RFA for selective reduction at another facility. Placental abruption was suspected and patient underwent a cesarean section. Twin A was delivered alive although she subsequently succumbed due to complications of prematurity.

Conclusion This case highlights the importance of early detection and consistent prenatal care in the management of TRAP sequence. Further research of interventions associated with improved outcomes should be encouraged.

Keywords
- TRAP
- complications
- monochorionic monoamniotic twin
- fetal

Introduction

Twin reversed arterial perfusion (TRAP) sequence occurs in monochorionic pregnancies consisting of one structurally and functionally normal fetus (pump twin) and a nonviable fetus (acardiac twin). The prevalence of this condition is around 2.6% in monochorionic pregnancies, with literature suggesting that incidence could be higher due to the increase of assisted reproductive technology and first-trimester scans. While this sequence is being detected earlier on and with more accuracy, its etiology still remains unknown.[1]

TRAP sequence consists of an acardiac twin with limited lower extremity development and, in some cases, with rudimental upper extremities. The acardiac twin's blood supply is hypoxic blood and provided by the normal twin. The anastomoses are either arterio-arterial or less commonly, veno-venous. This leads to the formation of a reverse arterial perfusion of the acardiac twin as poorly oxygenated blood enters through the umbilical artery and exits through the umbilical veins.[2] The pump twin has an increased risk of developing high-output cardiac failure, including polyhydramnios and hydrops.[1] The cardiac complications are expected due to the increased work that the pump twin's primordial heart is subjected to.

The following report discusses a case of monochorionic monoamniotic twin gestation with TRAP sequence *acardius acephalus* subtype.

Case Presentation

A 32-year-old African-American patient G4P2012 (gravida 4 para 2 aborta 1) presented to the emergency department with

received
April 7, 2024
accepted after revision
August 4, 2024
accepted manuscript online
August 12, 2024

DOI https://doi.org/10.1055/a-2384-8058.
ISSN 2157-6998.

Thieme Medical Publishers, Inc., 333 Seventh Avenue, 18th Floor, New York, NY 10001, USA

periumbilical abdominal pain and scant vaginal bleeding. Her past medical history was significant for a previous spontaneous abortion, asthma, anemia, chronic hypertension, hypothyroidism, occasional tobacco use, daily marijuana use, and alcohol use disorder. Pregnancy was confirmed 1 week prior at another hospital, though no prenatal care had yet been established. Initial transabdominal ultrasound showed a single live intrauterine 14 weeks and 5 days gestation, with an estimated fetal weight less than the 3rd percentile but no anatomical abnormalities. Further evaluation revealed a second intrauterine gestation with evidence of fetal motion, cardiac activity, and a large cystic hygroma. Transvaginal ultrasound confirmed a twin intrauterine gestation.

The patient established prenatal care at the prenatal clinic of Wyckoff Heights Medical Center in Brooklyn where a second-trimester ultrasound confirmed a monochorionic/monoamniotic pregnancy after pelvic examination had revealed uterine size–date discrepancy. Twin B had anencephalic acrania complex with complete absence of cephalic and cervical structures and absence of bilateral upper limbs. The thoracic component was markedly edematous with a small central pulsating structure appearing to function as the heart (►**Figs. 1**–**3**).

A month later, bedside ultrasound in the emergency department showed twin B had no fetal heart rate. The patient was discharged with routine obstetric appointments. She was lost to follow-up and when contacted, she stated she was scheduled for termination at another institution. She returned 6 weeks later, at 26 weeks and 1 day with contractions, increased discharge, and scant spotting over the prior few hours. She reported having undergone a procedure to reduce the acardiac twin at another facility since her last visit. She had undergone radiofrequency ablation (RFA) of the malformed twin's umbilical cord for selective reduction. Vaginal examination showed 5-cm dilation of the cervical os, 80% effaced, and −3 fetal station. Spontaneous rupture of membranes occurred with clear then dark red fluid. Fetal tracing showed contractions every 2 to 3 minutes and a fetal heart rate of 140 bpm. Suspected placental abruption and twin A's transverse lie prompted an emergency cesarean section. Twin A was delivered alive and twin B demised. Gross anatomy and histologic examination showed a monoamniotic, monochorionic placenta with chorionitis and hemorrhage suggesting abruption. The acardiac twin was only grossly examined (►**Fig. 4**).

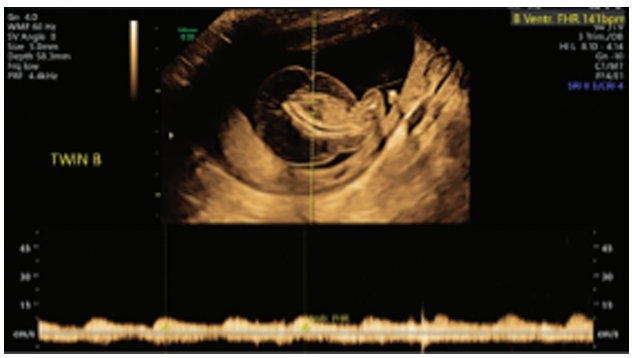

Fig. 1 TRAP fetus with cardiac activity. See marked edema of the thoracic structure. TRAP, twin reversed arterial perfusion.

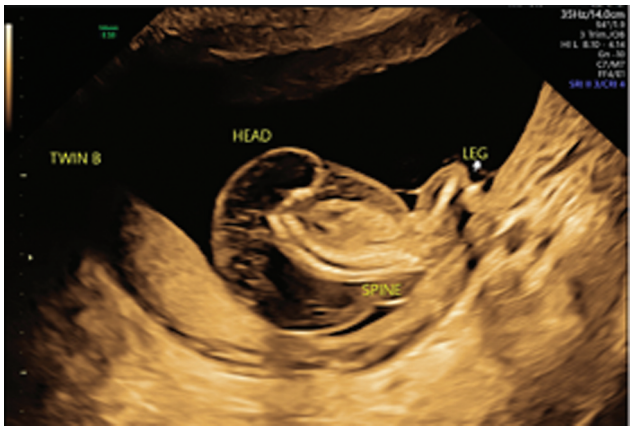

Fig. 2 TRAP fetus, marked edema. TRAP, twin reversed arterial perfusion.

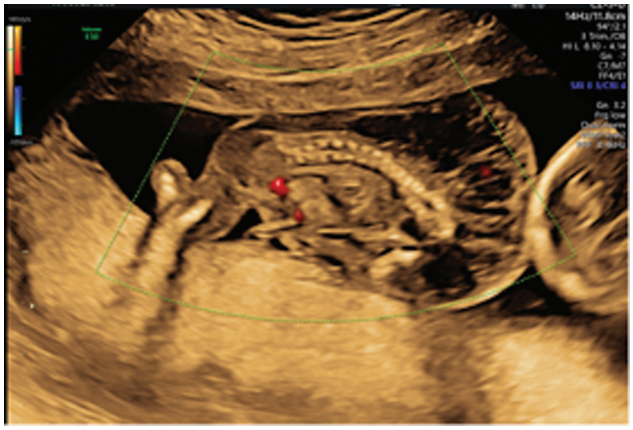

Fig. 3 TRAP fetus, placenta. TRAP, twin reversed arterial perfusion.

Twin A, born alive, was intubated immediately due to low tone, shallow breathing, generalized cyanosis, and Apgar scores of 1 at 1 minute, 4 at 5 minutes and 6 at 10 minutes. Transferred to the neonatal intensive care unit for perinatal depression and severe respiratory distress. During hospital stay, the patient developed bilateral grade IV intraventricular hemorrhage, hydrocephalus, necrotizing enterocolitis, and was pronounced dead on day 30 after cardiopulmonary arrest secondary to severe pulmonary hemorrhage associated with severe respiratory distress syndrome.

A week later, the patient presented to the emergency department complaining of blurry vision, headache, and general malaise. She was diagnosed with postpartum preeclampsia with severe features, and treated accordingly with magnesium sulfate infusion, hydralazine injection, and nifedipine extended-release. During this stay, she received mental health support due to anxiety and concern about twin A's poor prognosis.

Discussion

TRAP sequence is considered to be one of the most severe forms of twin-to-twin transfusion syndrome. The most commonly utilized methods of assessment are color Doppler

Fig. 4 Placenta, membranes, umbilical cord, and TRAP fetus with lower extremities. TRAP, twin reversed arterial perfusion.

or pulse wave Doppler velocimetry studies. In the absence of these tools, the presence of major malformations and the lack of a heart (or presence of a very rudimentary one as in our case) together with extensive edema of the fetus are strong indicators of an acardiac twin consistent with TRAP sequence.[1] In addition, the complications that can be noted on the pump twin are vitally important to detect as they play a major role in the timing of delivery. These include but are not limited to growth restriction, high-output heart failure, subcutaneous edema, and hydrops.[3]

The diagnosis of acardia is efficiently achieved during the second or third trimester of gestation by transabdominal ultrasound. In rare instances, the presence of TRAP sequence is only discovered at birth. On the other hand, there have been cases in which the diagnosis was done early in the pregnancy (11–12 weeks gestation) by transvaginal ultrasound. Early diagnosis is an important goal in patient management as it allows for early elective termination of the acardiac twin, thereby increasing survival chances of the pump twin.[1] Two other reportedly highly accurate diagnostic methods are magnetic resonance imaging and three-dimensional ultrasound. The latter is faster, less expensive, and has proven to be a great diagnosis method.[4,5] One report also points out the importance of karyotype testing to determine any abnormalities in the pump twin but the association between the two is still under study.[2]

Prognosis of this condition is generally poor. The acardiac twin's mortality is inevitable while the pump twin's mortality is still high (50–55%), due to the previously mentioned complications and their prematurity.[1]

Management may be conservative or interventional. The former approach uses expectant management, while the latter uses procedures aiming at stopping blood flow to the acardiac twin. This is achieved by two main techniques: RFA and bipolar cord coagulation (BCC).[1]

RFA is a minimally invasive technique that uses radio waves to heat the inserted needle and stop blood flow in the umbilical cord of the unhealthy fetus. In a 2013 study by Lee et al, the overall fetal survival rates of the healthy twin are $76.55 \pm 8.01\%$; specifically, in TRAP sequence it lowers to $\sim$80% with a mean gestational age of 36 weeks.[6] The commonly reported complications of this procedure include preterm delivery, preterm premature rupture of membranes (PPROM), and miscarriage.[7] There seems to be evidence of decreased mortality when the conservative approach is used.[8] BCC is another minimally invasive procedure to stop blood flow through the umbilical cord of the unhealthy twin. This procedure uses bipolar forceps to perform fetal reduction under ultrasound guidance. According to Weber et al, BCC use in TRAP sequence is more indicated after the first trimester, while at $<$12 weeks gestation, intrafetal laser could be a more beneficial procedure. In the same article, the survival rate of BCC reported is 82%.[9]

The first evaluation of our patient led to an initial report of a single intrauterine pregnancy and the diagnosis of a twin gestation was made at the beginning of the second trimester. It was also noted that one of the fetuses was without a cephalic pole. Because of irregular prenatal visits and the practice of seeking opinions and care at different institutions, the diagnosis and management of TRAP sequence were delayed. After the initial ultrasound that highlighted the physical abnormalities of twin B, the patient had undergone RFA at a different hospital, resulting in the termination of the acardiac fetus. This was followed shortly by preterm premature rupture of the membranes and labor which are well-known potential complications associated with RFA. Another notable finding in our case was the normal fetal karyotype in the normal fetus. Further studies are necessary to determine whether TRAP sequence is associated with any genetic abnormalities.

Conclusion

This case report highlights the importance of early detection and consistent prenatal care in the management of TRAP sequence. Although the early second trimester ultrasound detected abnormalities in the twin pregnancy, intermittent prenatal visits and care across different facilities contributed to a lack of care coordination. As a result, more definite intervention by RFA was pursued later in the pregnancy instead of potential earlier selective reduction.

While the RFA did successfully terminate the acardiac twin, the subsequent PPROM and preterm labor underscore the risks of invasive procedures. This case adds to the existing data on complications of this condition and of its management. Maternal complications including during the postnatal period cannot be overstated. Unfortunately, in this case, the surviving infant was delivered alive but succumbed due to the complications of prematurity. Further research aimed at improving diagnostic accuracy at an earlier gestational age may help in offering the patients earlier interventions with potentially improved maternal and neonatal outcomes.

Conflict of Interest
None declared.

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