

Case Report

The Case of the Vampire Baby Case Report: Twin Reversed Arterial Perfusion Sequence

Joseph R and Anderson S*Nova Southeastern University-Southwest Florida
Physician Assistant Program, USA***Corresponding author:** Anderson S, Nova
Southeastern University-Southwest Florida Physician
Assistant Program, 3650 Colonial Court, Fort Myers, FL
33913, USA**Received:** August 01, 2016; **Accepted:** August 23,
2016; **Published:** August 26, 2016**Abstract**

Twin reversed arterial perfusion (TRAP) sequence is a rare divergence of monozygotic monochorionic twin gestations resulting in an acardiac twin receiving its blood supply from an otherwise structurally normal twin via placental vascular anastomoses. If not managed, this unusual complication poses increased risks of heart failure in the viable twin, polyhydramnios, and parental mortality. This case report follows a primigravida second-trimester TRAP pregnancy, highlighting the necessity of serial observations and timely management to lessen morbidity and mortality.

Keywords: TRAP sequence; Twin gestations; Placental arterial anastomoses**Abbreviations**

TRAP: Twin Reversed Arterial Perfusion; ED: Emergency Department; ER: Emergency Room; HCG: Human Chorionic Gonadotropin

Case Presentation

A 29-year-old gravida 0, otherwise healthy, African American female presented to the ED with a 3-day history of worsening lower abdominal pain. The patient described the pain as sharp, severe, right lower quadrant pain radiating to the lower back. The patient denied nausea, vomiting, vaginal bleeding, dysuria, hematuria, change in bowels, and fever. She was stable with normal vital signs at time of presentation. Her last menstrual period was 2 months prior to the ED visit. A positive urine pregnancy test was obtained with subsequent quantitative beta-HCG. The transvaginal ultrasound revealed a twin gestation of approximately 7 weeks. Though twin pregnancy was confirmed, because there was no appreciable heartbeat noted from either embryo at the time, the attending ER physician recommended dilatation and curettage. The patient declined. She was discharged and instructed to follow-up with her obstetrician.

During the initial prenatal visit, a heartbeat was discovered in one twin, the other was termed a “vanishing twin.” The mother was advised that this twin would not develop further and would likely be “reabsorbed,” by the surviving twin. At the 17-week fetal anatomy ultrasound, a monochorionic-diamniotic gestation was confirmed; however, perfusion to the nonviable twin was detected. Furthermore, growth of the abnormal twin was notable in comparison to prior prenatal and ED ultrasounds. Though the anomalous twin was growing, it was amorphous with no distinct limbs or organs. Subsequently, the patient was referred to maternal fetal medicine and the diagnosis of twin reversed arterial perfusion sequence-acardius amorphous, was established.

The remainder of the pregnancy included genetic counseling and weekly visits to specialists at a regional children’s hospital. Each visit consisted of a detailed ultrasound with measurements of crown-rump length, biparietal diameter, femur length, and abdominal

circumference of the viable fetus; echocardiogram; amniotic fluid index; measurements of blood flow to the acardiac twin, donor twin, and placenta; as well as documentation of the growth of the viable twin relative to the nonviable twin. At the initial visit, the ratio of recipient to donor twin growth was 0.30; this increased over the course of weekly ultrasounds up until delivery at 38 weeks with a final ratio of 0.41. A ratio greater than 0.70 signaled stress to the viable twin’s heart and would have necessitated intervention.

Antenatal corticosteroids were administered at 34 weeks to aid fetal lung maturity and labor was induced at 38 weeks with a Foley balloon catheter and Oxytocin drip. An African American-Hispanic boy was delivered vaginally, weighing 2466.4 grams with Apgar scores of 8 and 9 at one and five minutes, respectively. After failed attempts at vaginal delivery of the abnormal twin, the mother was taken to the operating room for dilation and evacuation. It was essentially a mass, weighing 907.2 grams with no identifiable head or extremities. The mother suffered a third-degree vaginal tear and was catheterized for three days to prevent urinary retention. The remaining hospital stay was uneventful and both mother and baby were discharged four days postpartum.

Discussion

TRAP remains an uncommon anomaly exclusive to monochorionic multifetal gestations with an incidence of 1 in 9,500-11,000 pregnancies and 2.6% of monochorionic twin pregnancies [1-2]. Aside from these statistical findings, there are no known risk factors.

Two theories explain this phenomenon, dysmorphogenesis versus presence of arterio-arterial anastomosis [2-3]. The less popular theory, dysmorphogenesis, defines the underlying pathology as a defect in early organogenesis, resulting in a fatal malformed cardiac system [3]. Though arterial anastomosis contributes to the reversed blood flow, it is not responsible for the acardia. The arterio-arterial anastomosis concept describes a reversed umbilical artery blood flow as the principal cause of acardia. In this model, anastomosis causes deoxygenated arterial blood to flow from the donor to the

Table 1: 4 Types of TRAP morphologies.

Acardius Acephalus	<ul style="list-style-type: none"> Absence of head, upper extremities, and trunk. Maintenance of lower limbs, genitalia, and abdominal organs Most common type
Acardius Anceps	<ul style="list-style-type: none"> Present, but incomplete facial features and cranial structures. Rudimentary limbs and organs Most developed type
Acardius Amorphous	<ul style="list-style-type: none"> Heterogenous mass. No head, extremities, or organs Least developed type
Acardius Acormus	<ul style="list-style-type: none"> Head is the only formed structure, no limbs and organs Least common type

Due to the sensitive nature of this case report, the patient declined use of ultrasound imaging or photographs for publication.

recipient twin [3]. Unfortunately, circumventing the placenta results in a lack of proper perfusion for vital organ development, leading to the recipient twin appearing as a heterogeneous mass rather than a normally developing fetus. The acardiac twin essentially becomes a parasite, putting the surviving twin in danger of high-output cardiac failure, polyhydramnios, preterm birth, structural defects, and hydrops fetalis. Twin reversed arterial perfusion holds a perinatal mortality rate of 55% if not treated [4].

The artery-to-artery connection serves as the underlying cause of TRAP sequence. Arterial anastomosis and reverse flow of deoxygenated blood lead to stunted growth of the recipient twin. While the recipient twin mainly receives oxygen-poor blood, there is slightly more nutrient-rich blood flowing to the common iliac arteries and abdominal aorta that typically allow for growth of lower extremities, abdominal, and reproductive organs. This unequal vascular perfusion explains the different morphologies of TRAP [2].

The four types of acardia are classified based on presence or absence of various limbs and organs (Table 1) [5]. The most common type, acardius acephalus is the absence of a head, upper extremities, and trunk but maintenance of lower limbs, genitalia, and abdominal organs. In acardius anceps, the most developed type, facial features and cranial structures are present but incomplete along with rudimentary limbs and organs.

The twin in this case report had acardius amorphous, the least differentiated morphology. In acardius amorphous the recipient twin is essentially an ill-defined mass of tissue with no head, extremities, or organs. The fourth type and least common, acardius acormus, the head is the only formed structure with no limbs and organs [5].

Diagnosis

Early diagnosis and supervision is imperative to prevent mortality of the viable fetus in TRAP. In a cohort study of twenty-three TRAP cases, six untreated pregnancies resulted in intrauterine death before 20 weeks gestation, whereas 82% (14/17) of treated cases delivered a full-term, healthy neonate [6].

TRAP can be diagnosed as early as 11 weeks by 2-dimensional sonography, noting a grossly anomalous acardiac twin [7]. Further evaluation is then warranted with the use of 3-dimensional color Doppler ultrasound where reversed arterial blood flow is visualized and diagnosis confirmed [7].

Management

Once diagnosed, the pregnancy must be monitored with weekly visits to skilled specialists in maternal fetal medicine. Visits should

include detailed ultrasound and measurement of blood flow to the irregular twin. The greatest prognostic factor is the size of acardiac twin relative to the normal fetus. Growth of the recipient twin indicates increased workload and stress to the donor twin's heart. When the ratio of recipient to donor twin surpasses 0.70, intervention is necessary. According to one study of 49 acardiac twin pregnancies, if the twin-weight proportion was greater than 0.70, the chance of preterm delivery was 90%, polyhydramnios 40%, and CHF to the donor twin 30% compared with 75%, 30%, and 10%, respectively, if it was less than 0.70 [4].

Treatment

Antenatal intervention involves fetal surgery that obstructs blood flow to the nonviable twin via endoscopic ligation or laser coagulation of the umbilical cord, bipolar cord coagulation, or intrafetal radiofrequency ablation [8]. Intrafetal laser treatment is recommended at less than 16 weeks gestation, whereas radiofrequency ablation is preferred if greater than 16 weeks due to the higher rate of blood flow [9]. All methods can be performed under local anesthesia and conscious sedation. Survival of donor twin following in utero laser treatment or ablation is 80-90% [6,10]. Maternal complications are infrequent; however, there are still risks of hemorrhage and chorio-amnionitis.

Due to the high rate of preterm labor associated with TRAP, antenatal corticosteroid administration is necessary for all patients between 24-34 weeks' gestation. Early delivery is not required unless compromise to the viable twin is noted. Vaginal delivery is plausible, followed by dilation and evacuation of the anomalous twin if necessary. Cesarean section is specified for standard obstetric indications, e.g. threatening fetal heart tones and low biophysical profile.

Conclusion

This case report described the significance of strict monitoring for survival of the viable twin in TRAP pregnancies. Once TRAP is diagnosed, weekly observations are recommended to assess fetal measurements for signs of fetal distress and growth of nonviable twin, followed by appropriate treatment if warranted.

Both mother and baby in this case study are presently doing well. At 3 months old, the surviving twin has more than doubled its birth weight, weighing 5981.75 grams, and meeting all developmental milestones. Thus, despite its rarity, with proper surveillance and intervention, morbidity and mortality can be significantly reduced in TRAP sequence to yield a normally developed child.

References

1. Gemert MJV, Wijngaard JPVD, Vandenbussche FP. Twin reversed arterial perfusion sequence is more common than generally accepted. *Birth Defects Research Part A: Clinical and Molecular Teratology*. 2015; 103: 641–643.
2. Adamou N, Yakasai I. Twin Reverse Arterial Perfusion (TRAP): Case Report. *OJOG Open Journal of Obstetrics and Gynecology*. 2014; 4: 1072–1076.
3. Sogaard K, Skibsted L, Brocks V. Acardiac Twins: Pathophysiology, Diagnosis, Outcome and Treatment. *Fetal Diagn Ther Fetal Diagnosis and Therapy*. 1999; 14: 53–59.
4. Moore TR, Gale S, Benirschke K. Perinatal outcome of forty-nine pregnancies complicated by acardiac twinning. *American Journal of Obstetrics and Gynecology*. 1990; 163: 907–912.
5. Aggarwal N, Suri V, Saxena SV, Malhotra S, Vasishta K, Saxena AK. Acardiac acephalus twins: a case report and review of literature. *Acta Obstetrica et Gynecologica Scandinavica Acta Obstet Gynecol Scand*. 2002; 8: 983–984.
6. Pagani G, D'antonio F, Khalil A, Papageorgiou A, Bhide A, Thilaganathan B. Intrafetal laser treatment for twin reversed arterial perfusion sequence: cohort study and meta-analysis. *Ultrasound Obstet Gynecol Ultrasound in Obstetrics & Gynecology*. 2013; 42: 6–14.
7. Bornstein E, Monteagudo A, Dong R, Schwartz N, Timor-Tritsch IE. Detection of twin reversed arterial perfusion sequence at the time of first-trimester screening: the added value of 3-dimensional volume and color Doppler sonography. *Journal of Ultrasound in Medicine*. 2008; 27: 1105–1109.
8. Malone FD, D'Alton ME. Anomalies Peculiar to Multiple Gestations. *Clinics in Perinatology*. 2000; 27: 1033–1046.
9. Ville Y, Hyett JA, Vandenbussche FPHA, Nicolaidis KH. Endoscopic laser coagulation of umbilical cord vessels in twin reversed arterial perfusion sequence. *Ultrasound in Obstetrics and Gynecology Ultrasound Obstet Gynecol*. 1994; 4: 396–398.
10. Cabassa P, Fichera A, Prefumo F, et al. The use of radiofrequency in the treatment of twin reversed arterial perfusion sequence: a case series and review of the literature. *European Journal of Obstetrics & Gynecology and Reproductive Biology*. 2013; 166: 127–132.