

# Expectant Management of an Acardiac Twin Pregnancy

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**Abstract:** ***Definition:** A complication of monozygotic twin pregnancies in which one fetus develops normally (pump twin) and the second twin (recipient twin) demonstrates cardiac mal development ranging from complete absence of heart tissue to some formation of rudimentary myocardia. **Etiology:** Probable reversal of circulation in the anomalous twin resulting from anastomosis of circulation between the twins **Pathogenesis:** Malformation of the heart and heart dependent endodermal organs in the anomalous twin. **Associated anomalies:** In the perfused twin: total or partial absence of cranial vault, holoprosencephaly, anencephaly, absent facial structures, anophthalmia, microphthalmia, cleft lip, cleft palate, absent or rudimentary limbs, diaphragmatic defects, absent lungs and heart, esophageal atresia, ventral wall defects, ascites, absent liver and gallbladder, edema of the skin and single umbilical artery. In the pump twin: evidence of congestive heart failure (ascites, pleural effusions, polyhydramnios, skin edema). **Differential diagnosis:** Cystic hygromas, singleton pregnancies with intraamniotic tumors, pseudoacardiac twin. **Prognosis:** Uniformly lethal for the recipient twin; perinatal mortality rates reported up to 50 percent in pump twin. **Recurrence risk:** Not increased. **Management:** Serial ultrasound for evaluation of growth and signs of congestive heart failure in the normal twin.*

**Keywords:** Twin reversed arterial perfusion syndrome (TRAP), Acardius, acardiac monsters, Mono chorionic twins.

**Prevalence:** 0.3:10,000 pregnancies, 1% monozygotic twin pregnancies.

## 1. Introduction

Acardiac twinning is a very rare complication of multiple pregnancy which affects 1 in 100 monozygotic twin pregnancies and 1 in 35000 pregnancies overall. It is more common in twin pregnancies where the twin fetus which circulates the blood to it self and the cardiac twin is called pump twin and the other twin who receives the blood is called recipient twin. The exact patho-physiology which causes this fatal fetal anomaly is not known yet. But a common explanation for this condition is abnormal placental vascular anastomosis termed as Acephalus Acardiac Anomaly, also known as the Twin Reversed Arterial Perfusion theory (TRAP). In this condition arterial to arterial and venous to venous anastomosis in the placenta permits twin to twin transfusion and reversal of the blood flow in the umbilical vessel and the aorta of the recipient twin. The heart is deficient either by secondary atrophy or more possibly a primary pathology though currently unknown. As result cranial tissues are less likely to be perfused with oxygenated blood than caudal tissues. In addition to the absent heart and cranial and brain deformities variable degrees of somatic developmental destructions are present.

## 2. History

Mrs. SS, 24 year old mother who is in her third pregnancy with a history of previous two uncomplicated vaginal deliveries, referred at 24 weeks of period of gestation from Base Hospital Nikaveratiya for further management of twin pregnancy complicated with an anomalous fetus. This pregnancy was medically uncomplicated but she has the history of consanguinity (1<sup>st</sup> degree relatives). On examination her general condition was fare, size of the fundus was 36 weeks which is larger for the period of gestation. Presentation was difficult to determine due to the gross polyhydramnios. One fetal heart sound heard with a

rate of 140/beats per min. her routine investigations were within the normal range.

On admission the ultrasound scanning revealed twin pregnancy with one morphologically normal fetus and an acardiac-acephalic co-twin with reversed arterial perfusion. The biometry of normal fetus is

- Abdominal circumference- 24/52 weeks
- Biparietal diameter- 25/52 weeks
- Head circumference- 25/52 weeks
- Femur length- 24=1/52 weeks

No signs of exsanguinations in the pump twin.

The recipient fetus was motionless. In the sac of the pump twin, there was evidence of polyhydramnios but the pump twin had no evidence of ascites, pleural effusions, or skin edema. Doppler studies in the umbilical arteries of the pump twin were normal. In the anomalous twin, there was an anterior midline opening in the area of the upper thorax. No cephalic structures were developed.

## 3. Diagnosis

Whenever a twin pregnancy is detected by ultrasonography, and the twins have a discordant or a grotesque malformation; acardia should be strongly suspected. Serial ultrasounds are indicated to assess these complicated pregnancies.

Doppler velocimetry has been used to investigate the TRAP syndrome. Sherer et al reported the use of the Doppler velocimetry of the umbilical cord in the TRAP syndrome, reporting a markedly abnormal peak systolic to end diastolic velocity (S/D) ratio. Our case, probably due to the early gestational age, failed to demonstrate any Doppler changes.

#### 4. Management

The major perinatal problems associated with acardiac twinning include congestive heart failure and hydrops in the normal twin, hydramnios, and preterm delivery. Increasing perinatal morbidity and mortality has been associated with the relative size of the affected twin. Evidence for this is suggested by Moore et al. In a review of 49 cases of acardiac twinning, they found an overall perinatal mortality of 55 percent. To assess the effect of the anomalous twin's size on perinatal outcome, the weights of the twins were expressed as a ratio called the Twin Weight Ratio (TWR). The TWR is the wet weight of the acardiac twin divided by the weight of the normal twin. Preterm delivery, hydramnios and pump twin congestive heart failure in the normal twin were seen more commonly if the TWR was above 70%. This implies that the increased perfusion demands of the relatively large acardiac twin, compared to the normal twin, is related to prognosis. This data may be useful the counseling and management of these complicated pregnancies.

#### 5. Delivery of Twins

The pregnancy was planned to carry out till viability of the surviving twin with weekly ultrasound scanning. The poor prognosis of the acardiac fetus was explained to the mother and counseled.

At a POG of 27 weeks the patient had pre labour preterm rupture of membranes with grade 1 meconium stained liquor.

First of the twin delivered vaginally, baby girl weighing 760 grams with the Apgar of 10 at 5 minutes and transferred to the Special care baby unit for further management. Later developed Infant Respiratory Distress Syndrome and transferred to NICU for further management.

Acardiac twin weighing 740 grams delivered vaginally 1 hour after the first co-twin. Placenta delivered completely with two cords attached.

#### 6. Autopsy

Autopsy findings confirmed the diagnosis of a monochorionic diamniotic placenta with a vascular shunt and TRAP sequence. Both twins were female fetuses. The anatomy of the pump twin was entirely normal. The recipient twin had an edematous cephalad cystic pouch above the thorax. Histologic examination confirmed that the longitudinal openings in the anterior superior surface of the fetus represented developing cartilage, bone and skeletal muscle partially lined by skin. The internal examination revealed an empty thorax containing only edematous tissue and fluid. No diaphragm was seen. The abdomen contained a portion of small intestine with a cecum, appendix, large bowel and stomach. No evidence of a liver, spleen or pancreas was seen. Two normal kidneys and adrenals were present.

#### 7. Discussion

While the antenatal diagnosis of TRAP has been reported by several authors the pathogenesis of the TRAP sequence remains controversial. Evidence suggests that intrauterine

growth is achieved by perfusion from the normal twin via a large blood vessel anastomosis in the placenta. Perfusion of the anomalous twin occurs by reversal of flow through the umbilical vessels of that twin. The arterial blood that enters the body of the affected twin is presumably under reduced oxygen tension, which may cause disruption of organ morphogenesis. It has also been suggested that the primary defect is a failure of embryo logically paired structures, including the heart, to fuse in the midline, with subsequent maldevelopment.

#### 8. Types of acardiac twins

Types of acardiac twins have been categorized based on the degree of cephalic and truncal maldevelopment.

##### a) Acephalic

No cephalic structures present

##### b) Anceps

Some cranial structures and/or neural tissue present

##### c) Acornus

Cephalic structures but no truncal structures

##### d) Amorphus

No discernible cephalic or truncal structures

The acardiac acephalus fetus has no cephalic development at all. The acardius anceps fetus has cranial structures and/or neural tissue development. The acardius acornus fetus demonstrates cephalic structures but has limited or no truncal development. The fourth type, and most seriously mal developed, is the acardius amorphus fetus, which retains virtually no cephalic or truncal organization.

#### 9. Interventional procedures

Attempts to improve perinatal outcome by interruption of the circulation to the anomalous twin have been proposed. Platt et al speculated that termination of the circulation to the abnormal fetus might reduce the amount of hydramnios and subsequently prolong pregnancy. Seeds et al have suggested selective feticide in these pregnancies, but that the injection of a lethal substance would also endanger the normal twin. Recently, Fusi et al reported that the pump twin remains at increased risk of sudden death even without ultrasound evidence of cardiac failure, possibly due to acute disseminated intravascular coagulation. Hamade et al reported a steel coil placement in the abdominal wall of a acardiac fetus under ultrasound guidance.

Management of TRAP syndrome should include serial ultrasound to assess the growth rate and cardiovascular status of the normal twin.

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Antenatal Ultrasound pictures of the acardiac twin



Autopsy pictures of acardiac twin





Surviving twin and mother

