

A Rare Case of Acardiac Twin: A Case Report

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Abstract

Acardiac twin occurrence is rare. It is a serious complication of monozygotic multiple gestations. This is due to the sharing of blood supply between the twins in monozygotic gestation. This is called the twin reversed arterial perfusion occurring only in monozygotic twins. Monozygotic twins occur when the fertilized egg divides very early 4-8 days after fertilization. In monozygotic twin, acardiac twin is one of the twin that fails to develop head, arms and heart and gets its entire blood supply from the structurally normal pump twin. Acardiac twin has 100% mortality. Pump twin though structurally normal suffers due to heart failure and prematurity and has high morbidity and mortality all due to pumping blood to the acardiac twin. The acardiac twin receives all its blood supply from the pump twin through anastomotic channels, the term reversed perfusion is used to describe this condition because blood enters the acephalic twin through umbilical artery and exit through umbilical vein which is opposite to the normal blood supply. The acardiac twin loses direct vascular connection with the placental villi and receives its entire blood supply from the pump twin. Here we are presenting a primi gravida of 24-26 weeks of gestation with twin gestation of monochorionic di amniotic type in preterm labor. One of the twin was acardiac acephalous and another a pump twin with hydrops.

Keywords: Acardiac twin, Monochorionic twins, Twin reversed arterial perfusion sequence

INTRODUCTION

The occurrence of twin gestations and higher order multiple births have increased as a consequence of use of ovulation induction drugs and other assisted reproductive technology. Fetus in multiple gestations suffers variety of complications such as fetal malformations, preterm births, difference in birth weight, cord entanglement, intrauterine fetal demise, twin-to-twin transfusion syndrome (TTTS) and reversed arterial perfusion sequence. Zygosity is important in determining complications. Monozygotic twins have more complications than dizygotic twins. Acardiac twin occurs only in monozygotic twins. The presence of an acardiac twin occurs in one of every 35,000 twin pregnancies and in 1% of all monochorionic twin pregnancies.¹ Other names given are holocardius,

hemicardius, fetus amorphous. Such cases have been reported in the literature as early as 1533.

Occurrence of acardiac twin is due to twin reversed arterial perfusion sequence (TRAP) occurring early in embryogenesis. There is vascular communication between the twins in monozygotic twins. The vascular communication in acardiac twin is different, in that, the acardiac twin receives blood supply from other twin-pump twin through umbilical artery. The blood in the umbilical artery is mostly deoxygenated. Hence it leads to secondary organ atrophy.² Upper body does not develop at all, hence missing heart and head. All the blood supply to the acardiac twin is derived from the pump twin. The acardiac twin develops only lower part of the body or just a mass of tissue. Hence, the mortality for acardiac twin is 100%.

The pump twin suffers congestive cardiac failure and hydropic changes due to pumping blood to the acardiac twin. Mortality for pump twin is 50-70%. However, early identification and follow-up and treatment improve the survival rate of the pump twin. This article highlights the

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importance of early diagnosis of zygosity of twin. Early identification helps to plan the treatment for improving the survival of the pump twin.

CASE REPORT

A 21-year-old primigravida reported to our hospital at 24 weeks of gestational age with complaints of pain abdomen since one day. Her clinical examination revealed a uterus of 24-26 week's size. Per vaginal examination showed well-effaced cervix of 2 cm dilatation with tense bulging bag of membranes. Her ultrasonography (USG) scan (Figure 1) done on the same day reported twin intrauterine gestation with a single placenta and a thin membrane separating the two fetuses - monochorionic diamniotic twins. Twin B of 25-26 weeks of gestation with good cardiac activity with hydropic changes with grossly increased liquor (AFI-30) was seen on the right. Twin A - acardiac acephalic seen on the left. Within three hours patient progressed to active labor and delivered a first twin - (Figure 2) an alive, female

baby of 1600 g, followed an hour later by the acardiac twin (Figure 3) of 1000 g. The first twin was found to be normal without any external abnormalities. It was shifted to neonatal care unit, but perinatal mortality occurred. The second baby was acardiac/acephalous. The baby had well-formed lower limbs and the lower trunk which was normal. It had absent development of cephalic pole, heart and upper limbs. Both feet showed equinovarus deformity. The X-ray (Figure 4) of acardiac twin shows sudden abruption of the cervical spine.

Placenta was 300 g (Figure 5) with two umbilical cords. The normal twin cord was long and edematous, had three vessels. The acardiac twin had a short cord. Both twins shared the same placenta. Patient was transferred to ward in satisfactory condition and was discharged from hospital on the 5th postpartum day.

DISCUSSION

There are vascular connections in monozygotic twins. TTS is one of the manifestations affecting up to 15%



Figure 1: USG showing acardiac acephalic twin



Figure 3: Acardiac acephalic twin



Figure 2: Pump twin

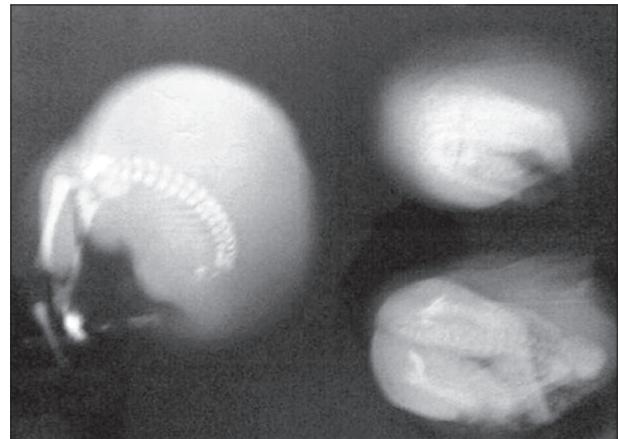


Figure 4: X Ray of acardiac acephalic twin showing sudden abruption of cervical spine



Figure 5: Placenta showing short & long umbilical cord

of monochorionic twins. In this condition there is disproportionate blood supply between the twins. Mortality is high without treatment.

TRAP sequence is one of rare occurrence. Here the pump twin pumps blood in reversed way, through umbilical artery, to the acardiac twin. The acardiac twin suffers 100% mortality.

Acardiac twin is classified according to the degree of cephalic and truncal maldevelopment.³

1. The first type is acardius-acephalus, where no cephalic structures are present. Head and upper extremities are lacking. It is most common variety. This is the type seen in the present case.
2. The second is acardius-anceps where some cranial structure and neural tissue or brain tissue is present. The body and extremities are also developed. It is highly developed form.
3. The third is acardius-acormus with cephalic structure, but no truncal structures are present. The umbilical cord is attached to the head. It is rarest form of the acardia.
4. The fourth type is acardius amorphous with no distinguishable cephalic or truncal structure. It is least developed and not recognizable as a human form with minimal development. This differs from teratomas only by its attachment to an umbilical cord.

A late separation of the embryonic cell mass results in a monochorionic twin pregnancy. In monochorionic pregnancies, anastomotic vessels are established connecting the two circulations. Retrograde perfusion via the anastomotic channel prevents the normal cardiac development due to lack of sufficient oxygenated blood. The cardia, if develops, is either tubular or completely infantile. Thus, the acardiac fetus becomes dependent on the perfusion of the "pump" twin.

The pathogenesis in TRAP sequence include.^{2,3}

1. Deep placental anastomoses in early embryogenesis cause malformation of the acardiac twin. The early pressure flow in one twin exceeds that of other leading to the reversal of flow in the umbilical artery of the co-twin.
2. A primary defect in embryogenesis in one twin leads to failure of cardiac development. The normal twin then perfuses the acardiac twin via artery-artery anastomoses. The anastomoses are not responsible for the cardiac anomaly but are established as a result of it.

Diagnosis of acardiac twin should be made early by ultrasound and Doppler by recognizing the absence of heart and reversal of blood flow in the umbilical artery. Once diagnosed it should be followed up to assess the weight ratio of twins, changes in the pump twin like cardiac failure and polyhydramnios.

A study done Moore *et al.*⁴ concluded that preterm delivery was strongly associated with the development of hydramnios and congestive heart failure in the pump twin. If the twin-weight ratio was above 70%, the incidence of preterm delivery was 90%; hydramnios was 40%; and pump-twin congestive heart failure was 30%. This was in comparison with 75%, 30%, and 10%, respectively, when the ratio was less. This suggests that estimation of the relative weights in acardiac twins provides prognostic information regarding the outcome. Poor outcome occurs with congestive heart failure and hydramnios in the normal twin conservative treatment is done when acardiac twin is small in size. Invasive treatment is required when pump twin is having cardiac failure to improve the perinatal outcome.^{5,6}

Treatment

Minimal invasive procedures like percutaneous insertion of helical metal coil to induce thrombogenesis in single umbilical artery of acardiac twin can be done.^{7,8}

Blocking the vessels by coagulation using Nd:Yag laser and radiofrequency ablation⁹ under ultrasound guidance are now the first line of treatment.

Each pregnancy has to be assessed individually and fetal surgery tailored.

CONCLUSION

Diagnosis of acardiac twin can be made in the first trimester itself by USG and Doppler. Early diagnosis of chronicity of twin pregnancy helps in improving the survival of the pump twin. Prevention of preterm labor and diagnosing cardiac failure in the pump twin is very important. First line

of treatment is by blocking the vessel of acardiac twin by radio frequency ablation by ultrasound guidance. Treatment at appropriate time improves the survival of the pump twin by 95% with an average age at delivery between 36 and 37 weeks.

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