

Case Report

Parasitic acardiac twin: a rare anomaly of monochorionic TRAP sequence

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ABSTRACT

Acardiac twin is a rare complication of monozygotic multiple pregnancy. Vascular anastomosis between twins, with single placenta leads to transfer of deoxygenated blood from umbilical artery of donor to the recipient twin in reverse direction through its umbilical artery which is responsible for abnormal growth. Acardiac twin dies in utero or immediately after birth with a 50-75% mortality of normal donor twin due to congestive heart failure secondary to strain of perfusing acardiac twin.

Keywords: TRAP sequence, Acardiacus, Twin pregnancy, Monochorionic twin

INTRODUCTION

Acardiac anomaly in twins was first described in the 16th century by Bendetti in 1533.¹ It affects 1% of monozygotic twins, or 1:35,000 deliveries². A rare condition where one twin is acardiac but receives perfusion from the second twin through arterial to arterial anastomosis present in placenta with blood flowing in reversed direction in parasitic twin therefore also called as Twin Reversed Arterial Perfusion (TRAP) sequence. It is a rarest variety of Twin to Twin Transfusion Syndrome (TTTS) where the acardiac twin is a true and complete parasite.

CASE REPORT

A 24 year old unbooked second gravida G2 P0 A1 was referred to our institute as an 8 months pregnancy with dyspnoea in labour. On examination uterus was over distended and fundus was palpable up to xiphisternum. On sonography she was diagnosed to have twin

pregnancy with first live foetus in breech presentation and second dead foetus with abdominal cysts and limb reduction defects. A single large placenta was localized at the fundo-posterior position. There was no history of fever, rash, radiation or any other teratogenic exposure in any of her trimesters. As the first twin presented as breech with features of fetal distress by cardiotocography she was taken for the caesarean section. A healthy girl of 1600 grams who cried immediately with APGAR '8' was delivered showing features of intra-uterine growth restriction. It was kept in nursery for observation and developed respiratory distress on day two. Second twin was grossly anomalous (Figure 1) weighting 1100 grams, had thin cord with single umbilical artery (Figure 2).

Monster had large head, tortuous spine, low set ears, absent neck and thoracic area, omphalocele and undeveloped upper and lower limbs. Anal and uro-genital structures were not formed. There was single large monochorionic diamniotic placenta with

polyhydramnios in donor segment and severe oligoamnios in parasitic twin. Skeletal deformation is shown in X-ray which confirms well-formed head, absent thoracic cage, heart and limbs, except partially developed right hand and abruptly ending vertebras (Figure 3). Autopsy revealed thick oedematous skin, absence of any intra-thoracic and abdominal viscera. Umbilical cord showed single artery and vein. Umbilical artery supplied blood directly to foetal aorta which leads to cranial development in our case (Figure 4). Many straw coloured fluid filled cavities were seen in abdomen and single large at the cephalic end.



Figure 1: Acardiac monster and normal female twin fetus.



Figure 2: Umbilical cord showing single umbilical artery and vein.



Figure 3: X-ray of acardiacus showing developed head, shoulder girdles, right humerus and abruptly ending vertebral column.

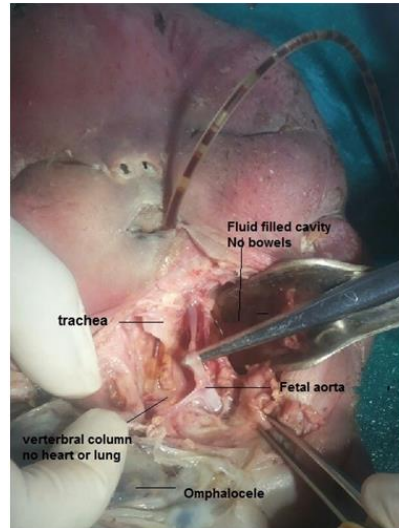


Figure 4: Autopsy showing absence of intra-thoracic and abdominal viscera and umbilical artery directly entering to foetal aorta.

DISCUSSION

Incidence of twinning is increasing with use of artificial reproductive techniques. Monochorionic / monozygotic twin has a high prevalence of structural anomalies as late embryonic division can cause malformations due to unequal distribution of inner cell mass, prezygotic meiotic error or error in postzygotic mitotic nondisjunction. Acardiac parasitic twin is unique to monochorionic pregnancies. Benirschke and Kaufmann,³ has reported different karyotypes in monozygotic co-twins resulting from either a) Late postzygotic non-disjunction at the time of embryonic splitting or b) It may be dizygotic aneuploidy with fusion of two separate placentas with vascular anastomosis.

Acardiac twinning should be suspected whenever foetal death is found with monochorionic pregnancy first trimester USG. Colour Doppler of umbilical artery shows reversal of flow and confirms the diagnosis of TRAP sequence.⁴ In absence of Doppler one can diagnose TRAP sequence by presence of single placenta, one fully developed foetus with polyhydramnios and occasional cardiomegaly, while the other dead but grow in grossly malformed fetus with either absent cranial structures or limbs. Artery to artery shunts develops in placenta through which deoxygenated arterial blood from healthy twin nourishes the acardiac through its umbilical artery (reverse of normal). Normally umbilical arteries join the common iliac vessels and blood preferentially goes to iliac vessels developing lower limbs. In our case single umbilical artery was found directly joining the aorta and developing the cephalic section well.

Depending of development acardiac twin is classified as acardius ancephalus: when head is poorly formed, acardius acephalus: if head is completely absent (commonest 60-75%), acardius acormus when only head

is present (Rarest), acardius amorphous: formless blob, no recognizable organs.⁵

Optimal management option between expectant and prenatal intervention is debated. Expectant management consists of serial antenatal surveillance and timed delivery when signs of heart compromise appear. Invasive treatment requires counselling about the prognosis of surviving twin, where embolization, fetoscopic ligation or coagulation of vessels supplying acardiac twin is done. Sullivan et al. advocates expectant management with foetal surveillance. Prognosis of donor twin is inversely related with acardiac/donor weight ratio. Complication rate is high if "Twin weight ratio" is greater than 70%. Malpresentation and fetal distress are major contributors for high operative delivery rate.⁶ In our case caesarean section was done due to first breech presentation and second foetal death. One foetal death increases the chances of complications in second twin. Normal foetus also has high risk of mortality (50 - 75%) due to anaemia, congestive cardiac failure and growth restriction. Long term cardiac and neurodevelopmental sequelae are high due to vascular imbalances in utero.

CONCLUSION

One dead foetus in multiple pregnancy case should be evaluated for acardiac or TRAP sequence by doing Doppler early in pregnancy when success of invasive management is better.

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