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Acardiac Amorphous Twin Pregnancy-A Rare Case Report

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ABSTRACT:
Background: Acardiac anomaly is a rare complication of multiple pregnancies. It is a hemodyamically disadvantaged nonviable twin which occurs in association with a twin reversed arterial perfusion sequence (TRAP). In TRAP, blood flows from an umbilical artery of the pump twin in reverse direction into umbilical artery of the perfused (or acardiac) twin via an arterial to arterial (AA) anastomosis. Its blood is poorly oxygenated and results in variable degrees of deficient development of the head, heart, and limb structures.**Case report:** A 21 year primigravida presented with pain abdomen and leaking pv attended to the GOPD. USG at 35wks 3days showed one healthy fetus and the other large heterogenous hyperechoic lesion with fetal parts on right side poorly visualised ? acardiac twin.**Discussion:** Imbalance of the interfetal circulation, the caudal aspect of the perfused fetus receives blood with relatively more nutrients and oxygen than the upper torso, resulting in better development of the pelvis and lower extremities in the acardiac fetus. Fully desaturated blood flows in a retrograde fashion to the upper body and head, leading to mal development of the heart, head, and upper torso, which may be either completely absent or severely deficient. Therefore, on USG it appears as a heterogeneous mass, simulating a teratoma or intrauterine fetal demise.
KEYWORDS: Acardiac Twin

INTRODUCTION:

Acardiac Twin is a rare complication in about 1% monozygotic multiple pregnancies [(1)] in 1 out of 35,000 deliveries [(2,3)] first described in the sixteenth century. The most widely accepted theory on the pathogenesis of acardius is the occurrence of a twin reversal arterial perfusion sequence (TRAP). TRAP refers to a rare, unique complication of monochorionic twin pregnancies in which a twin with an absent or a non-functioning heart (acardiac twin) is perfused by its co-twin (pump twin) via placental arterial to arterial (AA) anastomoses. [(3,5)] The acardiac twin usually has a poorly developed heart, upper body and head. The pump twin is at risk of heart failure and problems related to preterm birth. [(3,5)] Two criteria must be fulfilled for the development of a TRAP sequence. The first one is an anastomosis and the second is discordant development or in utero demise of one of the monochorionic twins, allowing for the blood flow reversal. Not infrequently, chromosomal abnormalities are identified in acardiac twin, whereas the pump has a normal karyotype. The diagnosis can be reliably made on ultrasound scan in the first trimester. [(7)] TRAP sequence is characterized by a grossly abnormal fetus that grows, may even show movements, but has no functional cardiac activity of its own. There are four types of acardiac twins. In the **acardiac-acephalus**, the thoracic organs and the fetal head are absent, **acardiac-acromas** only the fetal head develops, **acardiac-amorphous** consists a mass of tissue without recognizable human parts and the **acardiac -myelacephalus** the head and one or several extremities develop normally.

CASE REPORT:

A 30 year old lady G2P1 was referred to MKCG MCH, Berhampur, Odisha at her 37th week of twin pregnancy with a history of caesarean section complaining of pain abdomen and leaking per vaginum. Pre-natal ultrasonographic (USG) screening (2 times) showed one fetus with inconspicuous anatomy and structure. There were no signs of decompensation and biometry corresponded to 36 weeks of gestation. Her blood group was 'B' (+)ve. Routine investigations of pregnancy and coagulation profiles were done, which were within normal limits. She was treated with close fetal monitoring and a repeat USG done which showed one healthy fetus of 2.2 kg and a and a heterogeneous hyper echogenicity of 20 x 20cm inside the amniotic cavity separate from the placenta showing evidence of cord insertion, suspected to be either a demised fetus or abnormal placentation with a single amniotic sac with live active fetus apart from the mass. Amniotic fluid was 5.2cm. Caesarean section done due to monochorionic twin pregnancy with reduced liquor. A healthy female baby with APGAR 10 with birth weight of 2.3kg was delivered. The umbilical cord had three vessels; the length was 48 cm with no signs of decompensation. An amorphus mass of 20x20cm that could be an acardiac twin was

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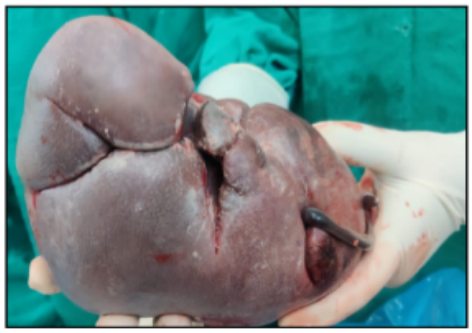
delivered with a single umbilical artery. Liquor was clear and reduced. There was a single placenta that was later examined and found to be monochorionic type. Amorphous mass was sent to pathology department for evaluation. Pediatric evaluation of the live baby was done and found fit. The mother's postoperative period was uneventful and was discharged on the 7th Post. OP Day with a healthy female baby and advised to come for follow up.



Acardiac Twin

DISCUSSION:

In the TRAP sequence, the normal twin 'pumps' or 'donates' blood to the abnormal twin, which is called the 'recipient' or 'perfused' twin through abnormal artery-to-artery communications in the placenta. [5] There is a reversal of flow in the recipient twin, with relatively oxygenated blood flowing from the abnormal anastomosis to the umbilical artery; the flow then proceeds cranially, leaving the fetus via the umbilical vein; hence the term, TRAP sequence. This finding can be confirmed by pulsed Doppler of the umbilical artery of the recipient twin, which will reveal reversal of flow on the spectral waveform. In 75% of cases, the umbilical cord of the recipient twin contains a single umbilical artery, which was also seen in this case. As a result of imbalance of the interfetal circulation, the caudal aspect of the perfused fetus receives blood with relatively more nutrients and oxygen than the upper torso, resulting in better development of the pelvis and lower extremities in the acardiac fetus. Fully desaturated blood then flows in a retrograde fashion to the upper body and head, leading to mal-development of the heart, head, and upper torso, which may be either completely absent or severely deficient. [5] This case was with these typical features. Therefore, on USG it appears as a heterogeneous mass, simulating a teratoma or intrauterine fetal demise. The acardiac twin usually has a dorsal cystic hygroma but in this case it was not present. The etiology of cytogenic discovery in TRAP twins is unclear. Polar body fertilization was described in an acardiac twin pregnancy, but zygosity testing excluded polar body fertilization as a likely cause of TRAP. [8,9] The development of acardiac fetuses in one of a pair of dizygotic twins in certain animal species who share anastomoses after fusion of two separated placenta has been described and these could be equivalent patho-mechanism in humans. The principal perinatal problems associated with acardiac twinning such as pump twin congestive heart failure, maternal hydramnios and preterm delivery did not occur in our case.

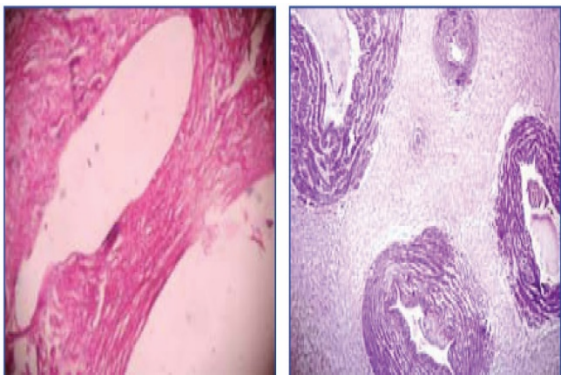


Fig 1: Rudimentary heart showing cystic spaces and myometrium
Fig 2: Umbilical Cord with two veins and one rudimentary artery

Damani reported a similar case with good perinatal outcome of the healthy co-twin without any antenatal intervention. [11] In a case series by Chandramouly and Namitha [3], they described two cases of acardiac twin with different outcomes. The first patient went into premature labor secondary to polyhydramnios at 27 weeks of gestation and delivered an amorphous mass and a normal-appearing fetus which died soon thereafter. The second patient was followed-up with serial USG examinations and she had a successful outcome, with the delivery of a normal fetus and an amorphous mass at term. Chromosomal anomalies may be present in up to 50% of cases of acardiac fetus.[2] Chromosomal analysis of acardiac twin showed trisomy[2]. Only a few abortion in the late first trimester have been reported and no case of trisomy[2] in which the fetus survived into the second trimester has been described. [10]

Chromosomal analysis of the acardiac fetus was not done in our case, but to evaluate the impact of aneuploidy in acardiac twin pregnancies, cytogenic investigations should be carried out routinely. The acardiac twin is non viable and the majority of efforts in management are focused on maintaining viability of the donor twin including close surveillance for development of hydrops. Several treatment modalities are practiced in these cases including palliative treatment which involve prolongation of pregnancy by serial amnio drainage and maternal administration of indomethacin for preterm labour.(7)

CONCLUSION:

In TRAP sequence, the normal twin 'pumps' blood to the 'recipient' twin via abnormal artery-to-artery communications in the placenta. Reversal of flow in the recipient twin, with relatively oxygenated blood flowing from the abnormal anastomosis to the umbilical artery; the flow then proceeds cranially, via the umbilical vein; hence, TRAP sequence. This can be diagnosed earlier by USG confirmed by pulsed Doppler of the umbilical artery of the recipient twin. Prenatal treatment involves occlusion of blood flow to the acardiac twin by endoscopic (fetoscopic) ligation or laser coagulation of the umbilical cord, bipolar cord cauterization, or intrafetal radiofrequency ablation. The indications for prenatal treatment include polyhydramnios, cardiac dysfunction, hydrops of the pump twin, or a relatively large weight of the acardiac twin. Though none of these modalities were available or tried in our case, we could achieve a good outcome only by monitoring the patient with serial USG and termination at term.

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