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An Evil Twin: A Case Report

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ABSTRACT
Acardiac Acephalus is a rare complication of monozygotic twins where Acardiac twin, also known as TRAP (Twin Reversed Arterial Perfusion Sequence) results from abnormal placental vasculature.
Here we present a case of 27-year-old female with natural conception, Gravida 4, para 2, having 2 live children by vaginal deliveries, one abortion at 2 months of amenorrhea a year ago, presented to labor room at 27 weeks 3 days of gestational age, with labor pain and had sonography suggestive of monochorionic diamniotic pregnancy complicated by TRAP sequence.
KEYWORD: *Acardiac Acephalus, TRAP sequence, Monochorionic Twins CASE REPORT*

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A 27-year-old female with natural conception, Gravida 4, para 2, having 2 live children by vaginal deliveries, one abortion at 2 months of amenorrhea a year ago, presented to labor room at 27 weeks 3 days of gestational age, with labor pain and had sonography suggestive of monochorionic diamniotic pregnancy complicated by TRAP sequence.

At patient's first antenatal visit at around 23 weeks, ultrasonography report showed monochorionic and diamniotic twins, polyhydramnios with deepest fluid pocket of 10 cm and single anterior placenta. Twin A showed normal morphology and growth consistent with 23 weeks of gestation and cephalic presentation with normal cardiac activity and umbilical artery Doppler. However, Twin B on further examination showed moderate to severe edema with no evidence of skull, upper limb or cardiac activity and cystic hygroma. Head circumference and femur length for age determination could not be done. She was then diagnosed with MCDA twins complicated by TRAP sequence with TRAP volume being 640 cc. On subsequent abdominal ultrasonographies, polyhydraminos was seen and after a fortnight, the TRAP volume was found to be 1069 cc. In the pump twin, MCA and DV appeared normal with no signs of fetal anemia. At 25 weeks, patient was suggested to go for Radiofrequency ablation or interstitial laser therapy for occluding TRAP vessel. The patient refused for any intervention and was thus called for weekly follow up.

Unfortunately, in next 12 days, patient went into labor. Patient was admitted with full cervical dilatation and her laboratory investigations like complete blood count, coagulation profile were normal. Twin A was delivered by vertex presentation vaginally and was of about 880 grams and despite immediate transfer to the neonatal intensive care unit (NICU), neonatal demise occurred 12 hours later. Trial of normal vaginal delivery was given for Twin B but absolutely no descent of the abnormal baby was appreciated. Thus in view of cystic hygroma in Twin B possibly hindering with the descent, patient was taken up for emergency Cesarean section where the Twin B was delivered as a soft tissue mass with no head, thorax but only lower limbs with lower abdomen present weighing about 1500 gms. The placenta was found to be of monochorionic type.

Figure 1 shows the Acardius acephalic fetus after C-section at 27 weeks, weighing 1500gms.

I. DISCUSSION

In TRAP sequence, there is abnormal artery to artery or venous to venous communication in the placenta where normal twin acts as the pump twin which donates blood to the recipient twin. This results in reversal of flow in the umbilical cord vessels of the recipient twin, oxygenated blood enters the fetus, through the umbilical artery and deoxygenated blood leaves through umbilical vein. As a result of an imbalance in the inter-fetal 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 pelvis and lower extremities. Fully de-saturated blood then flows in a retrograde fashion to the upper body and head, leading to faulty development of the heart and upper torso, which are either completely absent or severely deformed.

The acardiac twin is not viable, but the intrauterine period is dangerous for the well-being of the pump twin as well as it can be compromised through at least three mechanisms:[1]

congestive heart failure and polyhydramnios of the pump twin due to the increased blood flow;

Preterm premature rupture of membranes (pPROM), preterm labor and preterm delivery, caused by uterine overdistension, since the acardiac twin is often bigger than the pump twin

Hypoxia and intrauterine growth restriction of the pump twin, caused by the deoxygenated blood that comes back to the pump twin through vascular anastomosis.

Schatz (1899) divided the acardiac twins into 2 broad categories: one with and one without any signs of cardiac tissue. Das (1902) further categorized TRAP sequence classification into 4 groups still in use in modern literature[2,3,4]. These are as follows:

Acardius acephalus: Most common of all the types(60-75%), there is absence of head, upper extremities, and thoracic organs with fair development of lower extremities and pelvis.

Acardius amorphous: It presents as a mass of tissue with umbilical vessels (10%) and the second category having some rudimentary cardiac activity type. The former was referred to as holoacardius and the latter as hemiacardius.[5]

Acardius acormus: Very rare(5%) and characterized by only presence of the cephalic pole, while the body, if present is a shriveled mass. The head can be attached to the placenta either directly or through the umbilical cord.

Acardius anceps: Approximately 10% , it is the most morphologically developed acardius, with a recognizable body shape and extremities and a rudimentary development of head and face.

The typical ultrasound features are: gross differences in biometrical measurements of twins, in particular regarding abdominal circumference; absence of a morphologically normal heart in one twin associated with several other malformations in head, trunk, upper and lower extremities; presence of subcutaneous edema and fluid collections in the anomalous twin. Pulsed Doppler study of umbilical cord revealed reversed arterial flow toward rather than away from the anomalous fetus. Single umbilical artery can also be seen. The exact weight of the acardiac twin cannot be calculated using the standard formulas based on ultrasound biometry (such as Hadlock's), because of the usual lack of anatomical structures; the following formula has been proposed to estimate the weight of acardiac fetus: weight (g) = 1.2 × (longest length in cm)2 - (1.7 × longest length in cm)[6]. Wong et al focused on the use of the abdominal circumference in evaluating the acardiac fetus size and suggested a role of abdominal circumference ratio as a prognostic factor.[7] In majority of cases, pump twin develops high

cardiac output failure including cardiomegaly, pericardial effusion and tricuspid regurgitation with polyhydramnios and thus echocardiography is necessary for surveillance of cardiac function in pump twin. [8]

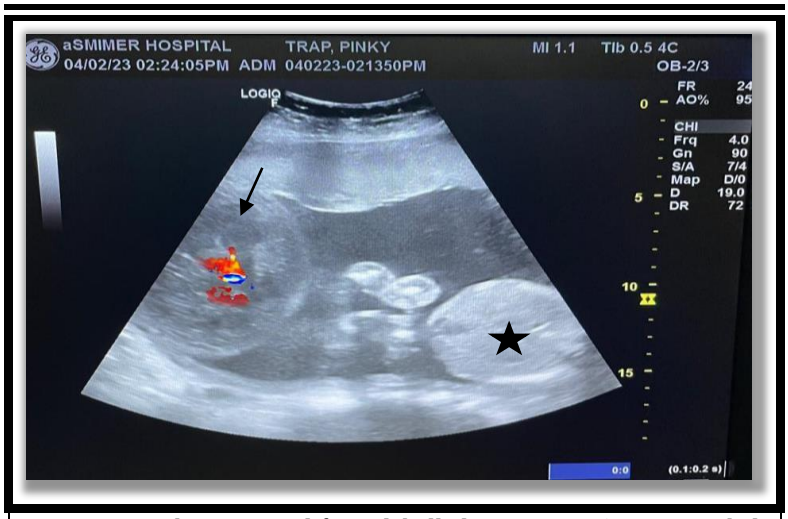
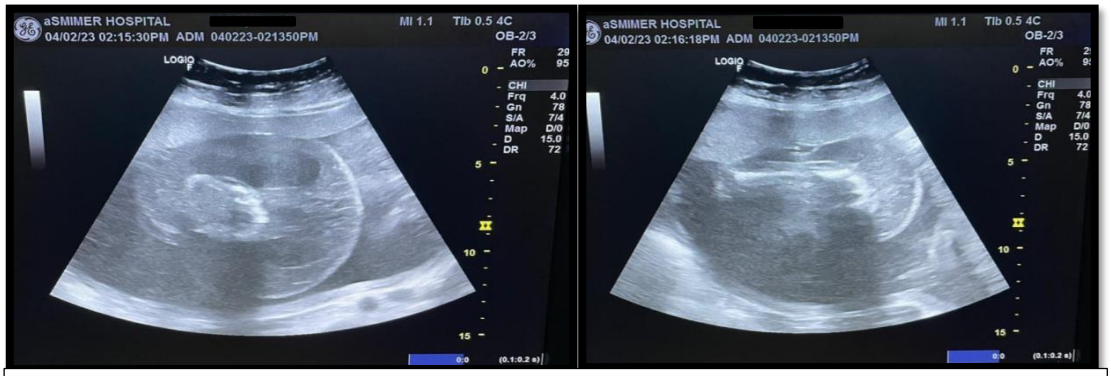


Figure 2 USG shows normal fetus, labelled as Fetus A, (seen as cephalic presenting part near the internal os, represented by star)
Fetus B located cranially to the normal fetus A, shows heterogenous and amorphous anatomy with adjacent cystic hygroma and hydrops and abnormal blood flow, represented by an arrow.

Fetus B shows reversal of blood from umbilical artery of



Fetus B in longitudinal scan and axial scan on 2D USG shows distorted, underdeveloped anatomy and seems to lack normal upper limb. Cephalic and thoracic architecture well formed. Abdomen, lower limb and pelvis seen. Findings consistent with Acardius Acephalic Fetus B with severe adjacent cystic hygroma.

The prognosis of conservative management is excellent if the estimated weight of the acardiac twin is less than one fourth of the pump twin. This risk is 100% when the weight of the acardiac twin exceeds 70% than that of the pump twin. At this point, some form of minimally invasive intervention is warranted to interrupt the vascular communication between the twins.

1. Intrafetal Ablation: The intrafetal approach has the goal of ablating the pelvic vessels or the abdominal aorta of the acardiac twin, which can be easily identified on color Doppler ultrasound. Greater than 90% survival of the pump twin is seen in TRAP Sequence treated with radiofrequency ablation. Recently, high intensity focused ultrasound (HIFU) has been used for occlusion of the blood flow in the acardiac twin.
2. Cord occlusion: Cord occlusion has been attempted by ultrasound-guided injection of thrombogenic materials into the umbilical circulation of the acardiac twin.

3. Ligation of the umbilical cord, laser coagulation, bipolar diathermy, and monopolar diathermy are the other available options to interrupt the vascular anastomosis between the two twins.

A study conducted by Moore et al. [6] reviewed the perinatal courses of 49 acardiac twin pregnancies, showed perinatal mortality to be 55% and the mortality was primarily associated with prematurity. When the twin weight ratio was above 70%, the incidence of pump-twin congestive heart failure was 30%, polyhydroamnios was 40%; and preterm delivery was 90%. Whereas the rates of congestive heart failure, polyhydroamnios and preterm delivery were 10%, 30%, and 75 %, respectively, when the twin weight ratio was less than 70%.

II. CONCLUSIONS

In midst of rising trends of twin pregnancies due to rising numbers of assisted reproductive technologies, this particular case where the risk of mortality of pump twin is very high, early diagnosis of acardiac twin by ultrasonography and colour Doppler is essential for timely management and preventing complications of pump co twin. Latest studies encourage intervention in the first trimester of pregnancy. As for the technique of choice to interrupt the vascular supply to the acardiac twin, USG guided laser coagulation and radiofrequency ablation of the intrafetal vessels are usually the preferred approaches.

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