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Twin Reversed Arterial Perfusion Sequence

Zainab Ali Abdulla Al-Jufairi, MRCOG, Arab Board* Naeema A A Mahmood, MB&CHB, Arab Board** Huda Jaffar Al-Jufairi, MBBS, Arab Board***

Background: Twin-Reversed Arterial Perfusion (TRAP) is a rare complication of monochorionic twins with a prevalence of 1 in 35,000 pregnancies. It is characterized by a structurally normal twin pump perfusing an anomalous recipient twin via an artery-to-artery anastomosis in a reverse direction.

Objective: To highlight Twin-Reversed Arterial Perfusion sequence and its clinical variable presentation.

Design: Prospective review.

Setting: Obstetrics and Gynecology Department, Salmaniya Medical Complex (SMC).

Method: All patients with TRAP sequence, diagnosed by ultrasound were followed up until delivery. Clinical presentation, ultrasound findings, outcome and management were reviewed.

Result: Three patients with a diagnosis of TRAP sequence who had been managed conservatively were included in the study. The first two patients had amorphous acardius and presented with polyhydramnios and both had favorable outcomes for the surviving twin. The third case was a triplet pregnancy, which ended unfortunately with extreme prematurity at 23 weeks of gestation of acephalus acardius and two normal fetuses.

Conclusion: TRAP sequence is a rare complication of monochorionic twins. Accurate antenatal diagnosis is essential to improve the prognosis of this rare entity.

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TRAP sequence is known as acardius or chorioangiopagus parasiticus, it is a rare complication of monochorionic twins. It affects around 1% of monozygotic twins with a prevalence rate of 1 in 35,000 pregnancies[1-2]. The risk of recurrence was estimated 1:10,000[3,4].

TRAP sequence is characterized by a structurally normal pump twin perfusing an anomalous

*Consultant
Obstetrics & Gynecology Department
Salmaniya Medical Complex
Kingdom of Bahrain
E-mail address: mamoamen@batelco.com.bh

recipient twin via an artery-to-artery anastomosis in a reverse direction[5]. The anomalous foetus is either acardiac or has severely anomalous cardiac structure[6].

The typical appearance of the acardiac twin is a hydropic mass without a heartbeat or with only rudimentary pulsatile cardiac structure[1]. Color Doppler sonography is helpful in diagnosing TRAP sequence, which shows reversal of arterial flow from the placenta towards acardiac twin[2,7-10]. The condition could be diagnosed in the first trimester by transvaginal scanning and Color Doppler sonography[2,11,12]. Transvaginal scanning shows increased nuchal translucency and pathological ductus venosus flow in the pump twin[11].

Several theories had been postulated to explain TRAP sequence. The most accepted theory is that artery-to-artery anastomosis between the monochorionic twins in the first trimester is the fundamental event in development TRAP sequence[2,13]. This abnormal circulation may result in early tissue hypoxia with resultant disruption of development of the cardiovascular system and a cascade of disruption of organ development in the recipient twin. Because the blood first perfuses the lower segment of the anomalous twin, the lower limbs and scrotum receive comparatively more oxygen than the upper segment of the body[14,15].

The anomaly is fatal for the recipient twin[2,3,7,16]. The pump twin may develop heart failure because of an increased cardiac demand. Numerous obstetric complications are associated with TRAP syndrome such as hydrops fetalis, polyhydramnios, umbilical cord accidents, preterm delivery or fetal death of the pump twin[12,17].

The differential diagnosis of acardiac fetus includes: anencephalic fetus, twin to twin transfusion, dead anomalous twin, placental tumors and amniotic band syndrome[8,10,18].

The aim of this study is to highlight TRAP sequence and emphasize its clinical variable presentation and management.

METHOD

Three patients with TRAP sequence, diagnosed by ultrasound at the obstetrics and gynecology department were followed up until delivery. Clinical presentation, ultrasound findings, outcome and management were reviewed.

RESULT

Three patients with a diagnosis of TRAP sequence were presented and discussed. The first two patients had favorable outcome while the third case ended with extreme prematurity.

Case One

Forty years old Bahraini lady, Gravida 8, Para 3 with previous four miscarriages was seen in the clinic for antenatal booking at 14 weeks of gestation. Her first three pregnancies ended with full term normal vaginal deliveries. The last four pregnancies ended with spontaneous miscarriage at 6, 10, 12 and 17 weeks respectively. Ultrasound examination revealed a viable fetus corresponding to 14 weeks with normal amount of liquor. Follow up scan at 23 weeks of gestation showed a viable fetus corresponding to date with severe polyhydramnios.

There were no structural abnormalities detected, however, there was 4.3 x 3.8 x 3.6 cm amorphous structure with absent cardiac activity attached by a thin umbilical cord containing two vessels. Doppler examination showed reversal of flow in the umbilical artery.

The impression was monochorionic mono-amniotic twin complicated by TRAP sequence resulting in one of the twin being acardiac. A Follow up scan at 30 weeks showed alive, breech male fetus with evidence of growth restriction (IUGR) and normal liquor. The acardiac twin has increased in size (7 x 6 cm) and there was edematous umbilical cord with high flow, see figure 1. The patient was counseled regarding the prognosis of the surviving twin and the potential risk of hydropic changes as well as intrauterine fetal demise. Because LASER ablation of the feeding vessels was not available in our center, we decided to follow the fetus with growth monitoring and Doppler ultrasound. She was given 2 doses of dexamethasone. A repeat ultrasound at 32 weeks showed no growth of the surviving twin with high edematous umbilical cord and abnormal Doppler study. Therefore, it was decided to terminate the pregnancy by lower segment cesarean section. Male alive infant weighing 1.02 kg was delivered with the acardiac twin. He had uneventful neonatal course and was discharged home at the age of two months. He continued to grow normally.

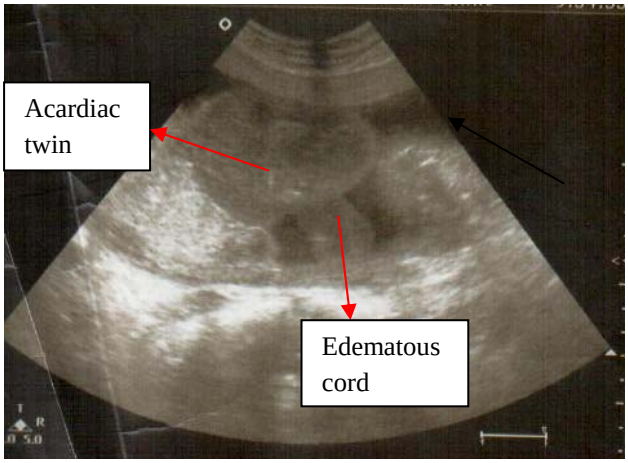

Figure 1: Ultrasound Image Showing Edematous Cord of Twin A and the Amorphous Acardius Twin B

The firm fleshy mass with tiny umbilical cord that was adherent to the placenta weighed 100 grams. The umbilical cord of the surviving twin was edematous and was sharing the same placenta, see figure 2. Histological examination confirmed the diagnosis of amorphous acardius.

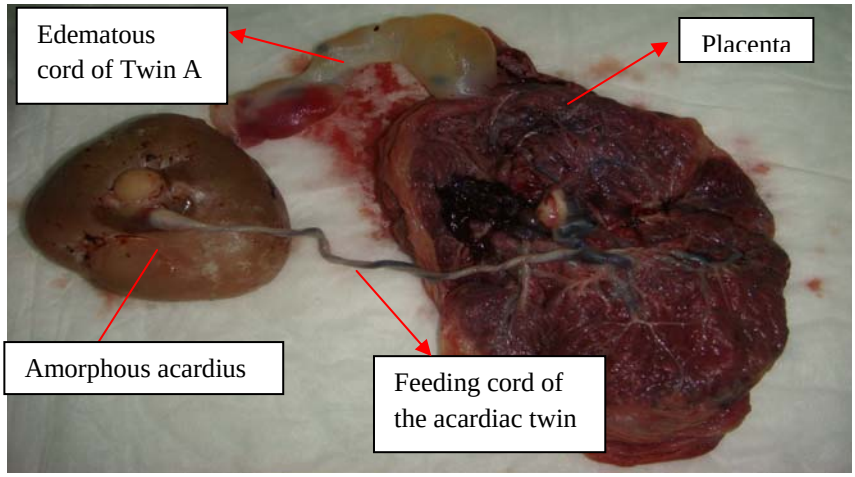

Figure 2: The Amorphous Acardius Attached by a Thin Umbilical Cord to the Placenta

Case Two

A twenty-seven year old Tunisian lady, gravid 2, para 1, referred to our institute in view of polyhydramnios at 25 weeks' gestation. Ultrasound examination revealed monochorionic monoamniotic twin pregnancy. The first twin was normal and corresponding to gestational age. An amorphous acardiac mass with gross edema was seen measuring 4.6 x 4.3 cm with reversed arterial blood flow, see figure 3. The parents were counseled about the possibility of TRAP sequence and the prognosis for the surviving co-twin. A follow up scan two weeks later showed that the surviving twin was growing well and the acardiac twin has increased in size. It measured 6.6 x 5.7 cm and the surrounded edema started to resolve, see figure 4. A third scan was performed a week later which showed similar findings. The patient returned home for her delivery and her husband had informed us that she delivered a normal healthy boy at term with the amorphous mass.

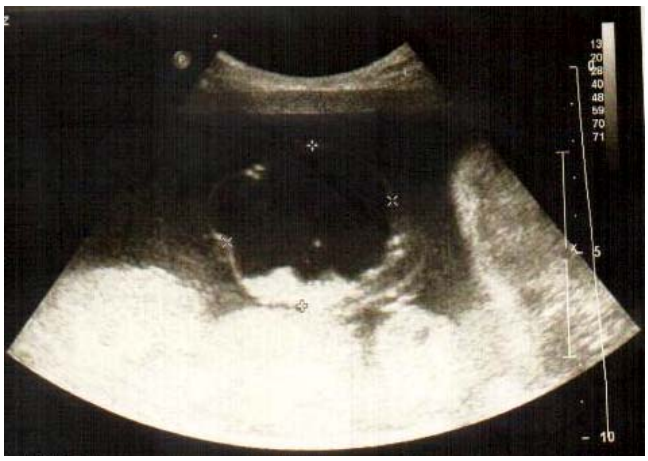

Figure 3: The Cursors Measuring the Diameter of the Acardius Twin, This Was Mainly Cystic at 20 Weeks Gestation

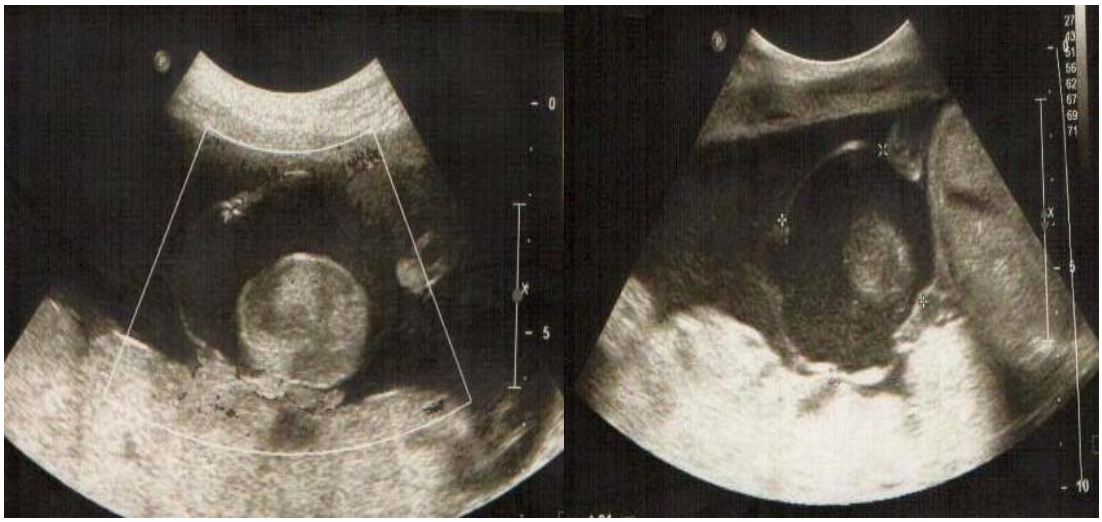

Figure 4: Follow Up Scan at 23 Weeks Gestation Showing Growth of the Amorphous Mass

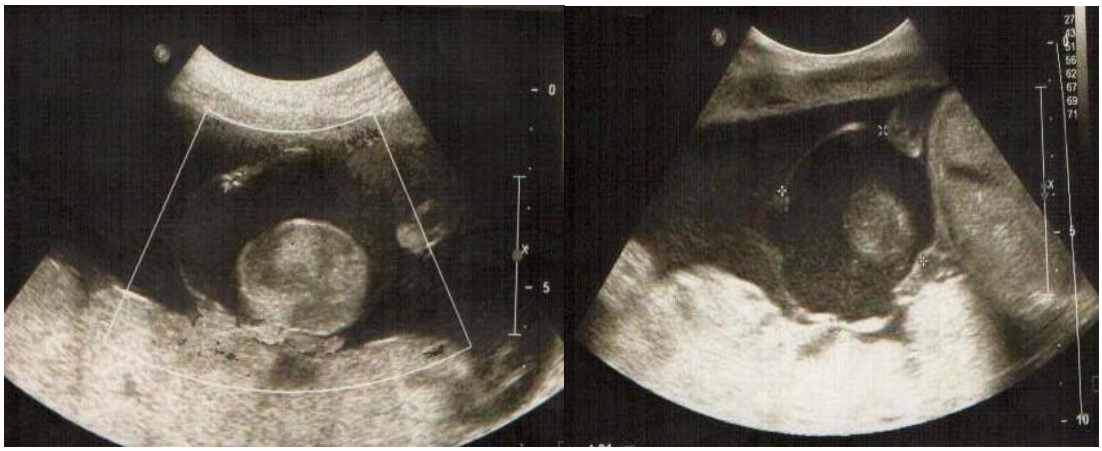

Figure 4: Follow Up Scan at 23 Weeks Gestation Showing Growth of the Amorphous Mass (Case 2)

Case Three

A twenty-five year old, Bahraini, primigravida, presented to the clinic at 20 weeks' gestation with triplet pregnancy. This pregnancy was Clomiphine induced after a year of infertility. Ultrasound examination revealed a dichorionic diamniotic placentaion between twin A and B. The fetuses were active and both corresponded to 20 weeks of gestation with no obvious congenital anomalies and normal amount of liquor. Twin B and C were monochorionic monoamniotic. There was amorphous tissue with probable spine, chest and trunk with an absent cardiac activity and gross edema which measured 9.2 x 6 cm, see figure 5. Doppler examination showed reversal of flow in umbilical artery. A repeat examination at 23 weeks'

gestation showed polyhydramnios with satisfactory growth of both twins, while the acardiac triplet had increased in size to reach 12.2 x 11 cm.

Unfortunately, the patient had preterm delivery at 23 weeks' gestation. The first two newborn were male weighing 570, 500 grams respectively, with no obvious external congenital anomalies and died soon after delivery. The third infant was acephalus acardius with absent head and developed thorax, abdomen and rudimentary lower limbs and weighed 800 grams.

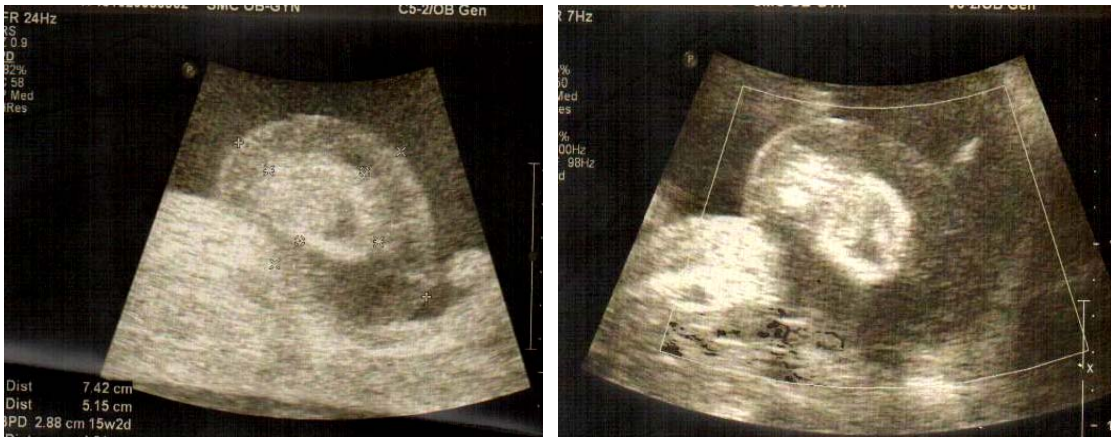

Figure 5: Ultrasound Images of the Acephalous Acardiac Triplet Demonstrating Probable Chest and Trunk

DISCUSSION

TRAP sequence is a rare complication of monochorionic multiple pregnancy. It is classified according to the degree of cephalic and truncal maldevelopment. The first type is acephalus, where no cephalic structures present. The second is anceps where some cranial structure and or neural tissue present. The third is acormus with cephalic structure but no truncal structures present. The fourth type is amorphous with no distinguishable cephalic or truncal structure[19].

The prominent features of the recipient twin are: total or partial absence of cranial vault, holoprosencephaly, 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[3,16,20].

The clinical presentation of TRAP sequence is variable. Two of our patients had presented with polyhydramnios and amorphous acardius co-twin. The first case was complicated by intrauterine growth restriction of the pump twin that necessitated intervention and delivery by cesarean section at 32 weeks, while the co-twin in case two has normally grown pump twin. This favorable outcome for both cases might be because the acardius compared to pump twin weight was less than 70%. However, the outcome of the third patient with triplet pregnancy was poor, as the patient went into preterm labor at extreme prematurity as the acardius to pump twin ratio was 160% (800:500 g). This finding was similar to the conclusion of Moore et al who reviewed 49 cases of acardiac twins and reported that perinatal outcome was related

to the ratio of the weight of the acardiac twin to the weight of the normal twin. When twin weight ratio was above 70%, preterm labor, hydramnios and congestive heart failure in the pump twin were found[21].

The perinatal mortality for the pump twin without intervention is 50-70%[5]. The treatment objectives are to either control the polyhydramnios or treat the cardiac insufficiency in the pump twin or to interrupt the circulation between the twins[2,17]. Several therapeutic modalities have been implemented. These include elective termination of both twins, intervention limited to management of polyhydramnios, digitalization to try to treat cardiac failure in the pump twin and hysterotomy to remove the abnormal twin. Recent therapeutic options targeted at interrupting the vascular anastomosis between the twins under ultrasound guidance using fetoscopy or by hysterotomy. This could be achieved by laser coagulation, unipolar diathermy, bipolar diathermy, radiofrequency, alcohol, cord coagulation or cord embolization[1,6,12,15,22-24].

Tan and Sepulveda recommended that intra-fetal ablation is the treatment of choice for TRAP sequence because it is simpler and more effective in prolonging pregnancy than cord occlusion[18]. On the other hand, Hecher et al found that fetoscopic laser coagulation of placental anastomosis or the umbilical cord of the acardiac twin was successful in 82% by laser alone and in a further 15% by laser coagulation in combination with bipolar forceps. The success rate is better in the early second trimester and less successful in the third trimester because the umbilical cord is more edematous with higher content of Wharton's jelly[24].

Because minimal invasive therapy for the vascular anastomosis was not available in our center, we decided to follow these pregnancies with growth monitoring and Doppler ultrasound of the surviving twin. Two of them had 'alive pump' twins but the third had very premature delivery without any survivors. The last patient could benefit from minimal invasive therapy as the acardius to pump twin weight ratio was more than 70%. She also could benefit from early diagnosis. High index of suspicion could assist in early diagnosis and accurate intervention.

CONCLUSION

Twin-Reversed Arterial Perfusion (TRAP) sequence is a rare complication of monochorionic multiple gestation. Accurate antenatal diagnosis is essential to improve the prognosis of this rare entity. Though many patients can benefit from conservative treatment, minimally invasive treatment modalities for the vascular anastomosis improve the outcome of the pump twin.

We recommend a special training on minimally invasive therapy, LASER, coagulation diathermy, for TRAP sequence and similar conditions as twin-to-twin transfusion syndrome.

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