

## Special forms in twin pregnancy- A cardiac twin/ twin reversed arterial perfusion (TRAP) sequence case report

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### Abstract

Acardiac twinning or Twin Reversed Arterial Perfusion (TRAP) is a rare anomaly occurring in monozygotic multiple pregnancies. The mortality in recipient acardiac twin is 100% whereas the normal donor twin is also at an increased risk of morbidity and mortality. This is a case of acardiac twins, preterm delivery occurred at 27 weeks and 3 days resulting in emergency caesarean section because of no progress. Dead twin was the presenting twin. The first twin was a male recipient fetus with head and chest odema and amblical hernia. The second fetus was male donor twin with severe respiratory distress admitted to neonatal intensive care unit and later passed away after 12 hours.

**Keywords:** monozygotic twin, TRAP twin (twin reverse arterial perfusion), amnioreduction

### Introduction

#### Case report

A Libyan 30 year old patient from Karsa (west Derna), she was in her fifth pregnancy and a mother of four children, referred to our clinic as a case of intrauterine fetal death. A transabdominal ultrasound performed confirmed monozygotic diamniotic twin pregnancy. The normal viable fetus had a biparietal diameter of 19 weeks and 5 days and normal fetal anatomy. Non viable dead twin a heterogeneous mass without abdominal and thoracic organs was distinguished together with diffuse edema (hydrops fetalis) with an umbilical hernia. The couple was counseled about risk of chromosomal abnormalities (9%), hemodynamic complications, which could lead to the pregnancy loss, premature birth, polyhydramnios and possibility of intervention and its risks<sup>[1]</sup> Need for referral was advised however couple declined referral to specialist center and conservative approach was adopted.

Pregnancy continued with follows up weekly, for vital signs, urine analysis fundal height, abdominal girth, and ultrasound examination every two weeks for growth and wellbeing of surviving twin.

The patient came back for a routine ultrasound examination at 22 weeks of pregnancy. The ultrasound examination showed structurally normal living twin with increased amniotic fluid volume largest pole was 8.5 cm and dexamethasone was given at 24 weeks.

Close follow-up of the patient continued with serial ultrasound examinations. Increasing amniotic fluid warranted intervention. Therapeutic amniocentesis for amnioreduction performed at 26 weeks and follow up continued as out patient. Preterm delivery

occurred 10 days later (at 27 weeks and 3 days) resulting in emergency caesarean section because of no progress. Dead twin was the presenting twin. The first male twin weighted 2.8 Kg, with head and chest odema, appear as one mass and there is amblical hernia (examphols). The final diagnosis was TRAP syndrome (Picture 1&2). The second fetus was male weighing 1.2 Kg (pump twin) severe respiratory distress admitted to neonatal intensive care unit and later passed away after 12 hours.



**Fig 1:** Ventral aspect shows head as a mass and exophalos



**Fig 2:** Dorsal aspect shows the head and neck as single mass with trunkal edema

### Discussion

TRAP syndrome is a rare obstetric condition unique to monozygotic monochorionic twin pregnancies in which there is co-existence of a normal pump (donor) twin and an acardiac recipient (perfused) twin [2, 3]. The incidence of TRAP syndrome is 1 in 35,000 pregnancies, 1 in 100 pregnancies of monochorionic monozygous twins, and 1 in 30 pregnancies of monochorionic triplets [2, 5]. Most cases are observed in twins also seen in 8% of triplets [4]. The risk of multiple pregnancies increases with higher order [5].

Two criteria seem to be necessary for the development of a TRAP sequence. The first is the presence of an arterio-arterial anastomosis and the second a discordant development or an intrauterine demise of one of the monochorionic twins, allowing for reversal of blood flow. In dichorionic twin pregnancies, intrauterine demise of 1 twin in the first trimester invariably leads to a "vanishing twin," whereas in monochorionic twins, existing anastomoses may actually prevent vanishing and sustain further cell growth and some degree of differentiation of the demised twin. Whenever a single intrauterine demise is diagnosed in a monochorionic twin pregnancy in the first trimester, the possibility of TRAP sequence should be kept in mind, which can be easily diagnosed by demonstrating the reversed blood flow in the umbilical artery with color Doppler flow [6].

The acardiac twinning include: hemiacardius (if the heart is incompletely formed) and holoacardius (if the heart is absent). Another type of classification includes: acardius anceps (when the head is poorly formed), acardius acephalus (if the head is absent), acardius acormus (presence of head only) and acardius amorphous (unrecognizable anatomy).

Prenatal diagnosis by the conventional 3/4D Doppler sonography. The acardiac twin – TRAP should be suspected in monochorionic twin pregnancies in which one of the fetuses is normally developed and in the other fetus, no structure or cardiac activity can be detected. The performance of karyotype for the pumping twin is mandatory.

The estimated fetal weight discrepancy (higher than 0.70) between the weights of the acardiac and pumping twin is indicators of an unfavorable prognosis. The higher the discrepancy the greater risk of polyhydramnios (deepest pool >

8cm) and premature delivery [7, 9]. The signs of cardiac failure in the pumping twin are polyhydramnios, cardiomegaly, ascitis, tricuspid regurgitation [1]. The acardiac twin and fetal hydrops in the pumping twin increased abdominal circumference. On Doppler ultrasonography (diastolic wave inversed on the umbilical artery, flow inversed on the ductus venosum) it will indicate cardiac failure in pumping twin.

Without intervention risks of intrauterine death (25%), polyhydramnios (50%), premature birth (80%) and with survival rate of approximately 55% [1].

The management implies serial ultrasound examination of the pumping twin in order to early detect the signs of fetal hydrops or the alternation of the flows. The administration of corticosteroids between 24 weeks and 34 weeks is important to optimize lung function in surviving twin, given the risk of premature birth. Over the years, many efforts have been made to treat TRAP. The interventions that can be performed are the following: laser ablation, bipolar cord coagulation, radio frequency ablation (RFA). If it is decided that intrauterine surgery is needed, there are two types of approach, such as umbilical cord occlusion by bipolar procedure (between 17 and 25 weeks of pregnancy), ligation (after 26 weeks of pregnancy) and selective fetal reduction [2, 4, 5].

Both methods can be approached through fetoscopy under ultrasound guidance, optimum timing of the procedure isn't clear in the literature. Both methods show similar success rates. The maneuver is chosen based on the surgeon's experience, gestational age and the choice itself. Fetuses between 18 and 27 weeks gestation, which are prone to a poor prognosis, are the main candidates for surgery. In a study by Lee *et al.* (2007), a survival rate of over 90% of healthy fetuses after radiofrequency fetal reduction of acardiac twins was reported. The aim of the treatment was to safely stop the blood flow to the acardiac twin, to ensure the safety of the pumping twin and to eliminate as much as possible the risk of heart failure or death. With regard to childbirth, a key issue was to avoid vaginal childbirth.

Chromosomal disorder has been reported in 50% of cases of TRAP syndrome [10, 11]. A case of TRAP syndrome reported in triamniotic dichorionic triplet pregnancies conceived by in-vitro fertilization [12]. Monochorionicity always highlights risk of TRAP. Our case was monochorionic diamniotic twins. Chromosomal analysis and doppler ultrasound were not done in our case due to their lack in our center and couple refusing referral to other centers.

### Conclusion

This is the first case reported in Alwahada hospital. The obstetrician and ultrasonographer should be aware of TRAP twin especially in twin/multiple pregnancies so that timely proper measures can be taken to rescue the pump twin. TRAP should be suspected in cases of monochorionic twin pregnancies when no structure/ cardiac activity is noticed in one of the twins while the other one is normally developed (dead of co-twin), (color Doppler sonography is mandatory) with reversed blood flow in the umbilical artery. Cardiac complications, which can lead to cardiac failure might appear early in pregnancy.

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