

Twin Reversed Arterial Perfusion Sequence (TRAP Sequence) - The Acardiac/Acephalic Twin: A Case Report

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Abstract: Twin Reversed Arterial Perfusion is an unusual form of twin to twin transfusion syndrome (TTTS) occurring in monochorionic twin pregnancies. There is an artery to artery or vein to vein communication between both the twins. One twin is normal and the other twin has an absent cardiac structure which is haemodynamically dependent on the normal (pump twin). Since the acardiac twin has no well-developed organs above the umbilicus survival is difficult. The prognosis is bad for the pump twin with mortality ranging from 50-70%. Antenatal diagnosis with ultrasonography and Doppler verification of reversed flow in the umbilical cord of acardiac foetus confirms the diagnosis. Early intervention during the antenatal period by radiofrequency ablation of major blood vessel of acardiac foetus can save the pump twin from high output failure. We report a case of acardiac, acephalic twin with hydramnios because of its rarity.

Keywords: Donor and recipient twin, TRAP sequence, twin pregnancy, acephalic, acardiac twin

INTRODUCTION

TRAP sequence is a rare complication seen in monochorionic twins with an incidence 1 in 35,000 pregnancies and 1% of monochorionic twin pregnancies [1].

There is a vascular communication artery to artery vein to vein between both the twin foetuses [2]. One twin is the donor which pumps blood to the recipient via the umbilical artery. As lower part of recipient twin receives oxygenated blood it is well developed. Due to supply of deoxygenated blood to upper part of the body, it is not well developed, thus the chances of survival are nil. Prenatal diagnosis by colour Doppler and ultrasound helps to salvage the donor twin by ablation of umbilical artery of recipient twin [3]. The prognosis of the pump twin is variable with mortality rate ranging from 50%-70% [4].

We report a case of acardiac, acephalic twin with hydramnios because of its rarity. We take an opportunity to discuss various management options.

CASE REPORT

A 20 year old, G2A1 with twin gestation with 36 weeks of gestational age presented to the Department of Obstetrics & Gynaecology with complaints of labour pains since 2 hours, draining per vaginum since 1 hour. She was unbooked case with no prior investigations.

Her clinical examination revealed over distended uterus, globular in shape, fundal height corresponding to 36 weeks of gestation. First twin was in breech presentation. Uterus was acting mildly. Liquor appeared to be less. One fetal heart sound was auscultated. Cervix was fully effaced with 2cm dilatation. Presenting part - breech at -2 station. Membranes were absent and clear liquor was draining. Pelvis was gynaecoid.

Emergency bedside ultrasound revealed twin foetuses, monochorionic monoamniotic placenta, upper segment, anterior; grade 2. Twin A -breech; good foetal heart sounds present. Twin B- exact presentation could

not be made out. Foetal heart sounds absent. AFI -7cm - IUD of Twin B .Other investigations were normal.

Twin A, a male child of 2.5 kg was delivered by caesarean section and was found to be normal without any external malformations and was admitted in neonatal care unit.

Twin B was child of 1.45 kg with absent development of cephalic pole, heart and upper limbs. It had well-formed lower trunk and lower limbs. It had an abdominal stump without any intraabdominal organs; there were two well-developed lower limbs which showed massive, diffuse, soft tissue oedema. Bony abnormalities of lower limbs present. Sex of the baby could not be determined clinically. Placenta was weighed 500 gms; monochorionic monoamniotic.

Patient was transferred to ward in a satisfactory condition and was discharged from hospital on 6th postpartum day with healthy male baby, was advised to come for follow up after six weeks.



Fig. 1: Acardiac Acephalic twin with well developed trunk and lowerlimbs

DISCUSSION

In the TRAP sequence, the acardiac or recipient twin is dependent on the normal donor or pump twin for its blood supply via trans-placental anastomosis and retrograde perfusion of acardiac umbilical cord [3, 5]. Deoxygenated blood from the donor or pump twin enters the umbilical artery of the acardiac twin in reverse direction, resulting in better perfusion of the lower body and extremities. The upper part of the body of the recipient is poorly perfused leading to its absent or poor development [6]. The donor twin is responsible for the pumping and maintenance of circulation of both twins causing a circulatory overload of the donor twin resulting in heart failure and polyhydramnios [7, 8].

Classification

Depending on the extent of cephalic and truncal development it is classified as follows [3, 9]

Type I – Acardius Acephalus: Most common variety, where head and upper limbs are absent. Our case belongs to this type.

Type II – Acardius Anceps: It is a highly developed form where brain tissue is present and body and extremities are well developed.

Type III – Acardius Acormus: Rarest form where head is present neck and upper extremities; abdomen and lower limbs are not fully developed; umbilical cord attached to head region

Type IV—Acardius amorphous: Least developed. No distinguishable head or trunk, differs from teratomas only by attachment to umbilical cord.

9% of pump twins have congenital anomalies [1]. Perinatal mortality in donor twin is 50 -55% due to circulatory causing high output cardiac failure or prematurity secondary to polyhydramnios [10]. Doppler verification of reversal of flow of blood in the umbilical cord of the acardiac twin confirms the diagnosis of TRAP sequence [11].

The indications for therapy are cardiac dysfunction, polyhydramnios, hydrops of the pump twin or large weight of the acardiac twin [12]. Prenatal management to save the donor twin involves radiofrequency ablation of major blood vessel in acardiac twin so that the donor twin need not pump blood to the acardiac twin.

Other methods of management are by endoscopic ligation, laser coagulation or bipolar cord cauterisation [13, 14].

CONCLUSION

TRAP sequence is an example of twin to twin transfusion syndrome seen in monochorionic twin pregnancy. Improved imaging techniques enabled early and accurate diagnosis of the condition in the first trimester. Improving the outcome of the pump twin by early intervention and monitoring is important. Chromosomal anomaly of the pump twin should be excluded prior to foetoscopic procedure due to risk of congenital anomalies.

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