A CASE OF TRAP SEQUENCE WITH IMMENSE ACARDIAC TWIN

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ABSTRACT

Twin-reversed arterial perfusion sequence is a rare complication of monochorionic twin pregnancy in which an anomalous acardiac fetus is hemodynamically dependent on its structurally normal “pump” twin. Early diagnosis is essential for improving perinatal prognosis for the normal twin. In this case report we present a case of TRAP sequence with immense acardiac twin with favorable outcome, emphasizing the importance of ultrasound imaging, follow-up and timing of delivery in this complicated pregnancy.

Key words: TRAP sequence, prenatal diagnosis, monochorionic twins

INTRODUCTION

Twin-reversed arterial perfusion sequence (TRAP) is a rare complication of monochorionic pregnancy. The prevalence of TRAP syndrome is about 1 in 35000 pregnancies, or in 0.3% of monozygotic twin gestations.1-3 The syndrome is characterized by one well-developed structurally normal twin called “pump” twin, while the other twin with absent cardiac structure, called ‘acardiac twin’, is hemodynamically dependent on the “pump” twin. The pump twin perfuses the anomalous recipient twin via an artery-to-artery anastomosis in a reverse direction.1-5 The perinatal prognosis of the pump twin is variable with a mortality rate between 50-70%.1

The usual complications that influence the prognosis are prematurity secondary to preterm delivery, congestive cardiac failure due to increased cardiac demand and polyhydramnion.6,7 Early detection of TRAP syndrome is essential for the appropriate follow-up, aggressive treatment – endoscopic laser coagulation of umbilical vessels, timing of the delivery, and the prognosis of the pump twin.8-11 Grey scale ultrasound and Doppler techniques are crucial for early diagnosis of TRAP sequence.9,12

We report a case of TRAP sequence in monochorionic-diamniotic pregnancy with immense acardiac-acephalic twin and favorable outcome for the pump twin.
CASE REPORT

A 27-year-old woman G3P2 (Gravida 3/Para 2) underwent ultrasound examination at 18 w.g. to detect congenital anomalies. The ultrasound scan revealed a monochorionic-diamniotic pregnancy. One of the twins was described as an acardiac fetus with cystic hygroma, lower limbs, pelvis, and part of the spine was visible. No cranium was identified. Color Doppler was not performed because no blood flow was expected. No structural defects were described in the other twin except absence of visible nasal bone and a frontomaxillary angle of 91.59°. Color and pulse Doppler showed normal parameters of ductus venosus and the tricuspid valve. Amniocentesis was performed and normal female karyotype was found. Monthly follow-up was performed.

The woman was admitted in our department again at 30 w.g. with complaints of dyspnea and irregular uterine contractions. The patient reported that she had not visited her consulting obstetrician for two months. The ultrasound findings were: an immense acardiac-acephalic fetus, the upper part of the body displayed as a cystic heterogeneous mass with diffuse soft tissue edema (22 cm in diameter), underdeveloped chest, hypoplastic ribs, underdeveloped lower limbs (FL – 4.05 cm, 22+ w.g.) and club foot. Mild polyhydramnion was also found. Blood flow in the umbilical cord and in the body of the acardiac twin was detected by color and pulse Doppler. The parameters measured in the pump twin were normal for the gestational age. Normal Doppler parameters were measured in the umbilical artery, ductus venosus and the middle cerebral artery. Echocardiography of the pump twin showed significant tricuspid regurgitation. Careful ultrasound examination of the placenta discovered artery-to-artery anastomosis between the two umbilical cords. Placentomegaly was also described (Fig. 1).

Figure 1. Pathological blood flow in the acardiac twin at 30 w.g.

Figure 2. Acardiac twin after delivery.
Due to the ultrasound findings for heart failure of the pump twin and the woman’s complaints, cesarean section was performed after three days of a dexamethazone prophylactic regimen. The weight of the pump twin was 1070 grams, the APGAR score 5 at 1 minute, and the baby was admitted in ICU. The weight of the acardiac twin was 3560 grams with presence of a solid mass (diameter 24 cm) engaging the upper part of the fetal body (Fig. 2). Histological study revealed acardiac-acephalic fetus, a kidney, an adrenal gland and an ureter, caudal part of the spine, two ribs and underdeveloped lower limbs. Artery-to-artery anastomosis was found in the placenta (Fig. 3).

DISCUSSION

TRAP sequence is a rare complication of monochorionic multiple pregnancy. The etiology and mechanism of its development is not clear. Many authors suggest the hypothesis that disruption of organogenesis occurs secondary to a reversed flow through an abnormal artery-to-artery placental anastomosis resulting in cardiac and other malformations. However, other researchers put forward the hypothesis that initial abnormal cardiac formation during embryogenesis initiates the development of TRAP sequence.

According to the degree of cephalic and truncal maldevelopment, TRAP sequence is divided into four types. The first type is acephalus – no cephalic structures are present. The second type is anceps – some cranial structures or/and neural tissue are present. The third type is acormus – cephalic structures but no truncal structures are present. The fourth type is amorphous with no distinguishable cranial or truncal structures.

In TRAP sequence the normal pump twin donates blood to the abnormal twin called “recipient”. It is possible through an abnormal artery-to-artery or venous-to-venous communications in the placenta. This leads to 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 the fetus via the umbilical vein, which is the opposite of what the normal blood flow pattern to a fetus should be. As a result of imbalance in the interfetal circulation, the caudal part of the perfused fetus receives blood with relatively more nutrients and oxygen than the upper part, resulting in better development of the pelvis and lower limbs. Fully desaturated blood then flows in a retrograde fashion in the upper part of the body, leading to faulty development or missing of the heart, cranium and upper limbs.

Congenital anomalies are present in about 9% of pump twins. Perinatal death of such twins occurs in 50-70% of pregnancies and results in severe prematurity, heart failure, umbilical cord incidents and polyhydramnion. Usually, the weight of the acardiac twin is about 70% of the weight of the pump twin. Proper ultrasonographic diagnosis is essential for achieving favourable perinatal outcome. Recent therapeutic options are 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.

By reporting this clinical case we would like to emphasize the importance of early diagnosis of TRAP sequence. Thus, the management of the condition can be more efficient, leading to favourable outcome for the pump twin.
CONCLUSIONS

TRAP sequence is a rare complication of monochorionic pregnancy which is poorly known by obstetricians. Delays in diagnosis are common because of the TRAP sequence scarce development. Ultrasound diagnosis is essential for the favourable outcome for the pump twin. The use of color Doppler and power Doppler techniques in every monochorionic pregnancy with clinical manifestation of an anomalous twin allows easy establishment of the diagnosis.

REFERENCES