Twin-Reversed Arterial Perfusion Sequence/Syndrome (TRAP): An Insight into the Ultrasonographic Features for Prenatal Diagnosis and Review of Literature

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ABSTRACT

We present two case reports of twin reversed arterial perfusion sequence- a rare type of twin-twin transfusion syndrome wherein there is abnormal development of one fetus which derives its blood supply from the normal fetus. It is present in cases of monochorionic twin pregnancies and is also known as chorangiopagusparasiticus. Prompt diagnosis and intervention are needed to prevent the death of the normal twin. We discuss the importance of diagnosing this entity.

Keywords: TRAP; Twin; Acephalus; Acardiac; Embolisation; Ligation

INTRODUCTION

Twin Reversed Arterial Perfusion Sequence (TRAP) syndrome is a rare obstetric condition unique to monozygotic monochorionic twin pregnancies in which there is coexistence of a normal pump (donor) fetus and an acardiac recipient (perfuse) fetus [1]. The incidence of TRAP syndrome is 1 in 35000 pregnancies, 1 in 100 monozygotic monochorionic twin gestations and 1 in 30 monochorionic triplet gestations [2]. Most cases are seen in twins with only 8% are seen in triplets [3]. The risk increases with multiple pregnancies of a higher order [4]. Diagnosis is necessary for proper prenatal management and can be established by ultrasonography and color Doppler examination of the umbilical artery of an abnormal twin. We present two case reports to highlight the radiological features.

CASE REPORT 1

A 30-year old primigravida female patient presented for the second-trimester anomaly scan. Her blood parameters including complete blood count, hemoglobin, bleeding time, clotting time, TORCH (toxoplasmosis, rubella, cytomegalovirus, and herpes simplex) profile were within normal range. Ultrasonography (USG) with color Doppler found a live fetus with a biparietal diameter (BPD) and femur length (FL) corresponding to the gestational age of 19 weeks 2 days and another heterogeneous mass in the amniotic cavity. On careful examination, the mass was seen connected to the normal fetus via an umbilical artery and consisted of fetal liver and gut loops surrounded by membranes with absent fetal head, thorax and upper limbs (Fig. 1a). Color Doppler study revealed high pulsatility index (PI) and resistivity index (RI) of the involved umbilical arteries (Fig. 1b and 1c). Single anterior uterine wall placenta was visible. The diagnosis of twin pregnancy with one normal fetus and another acardiac acephalic twin was made with a demonstration of TRAP on color Doppler and patient was sent to obstetrics service for further management.

CASE REPORT 2

Another patient came to our department for anomaly scan in the second trimester. USG with color Doppler found one normal fetus with another twin appearing as a heterogeneous mass comprising liver and gut loops without heart and

Figure 1a: Longitudinal ultrasonography showing the heart of pump twin and heterogeneous mass on the side (perfuse twin) without head and upper torso.

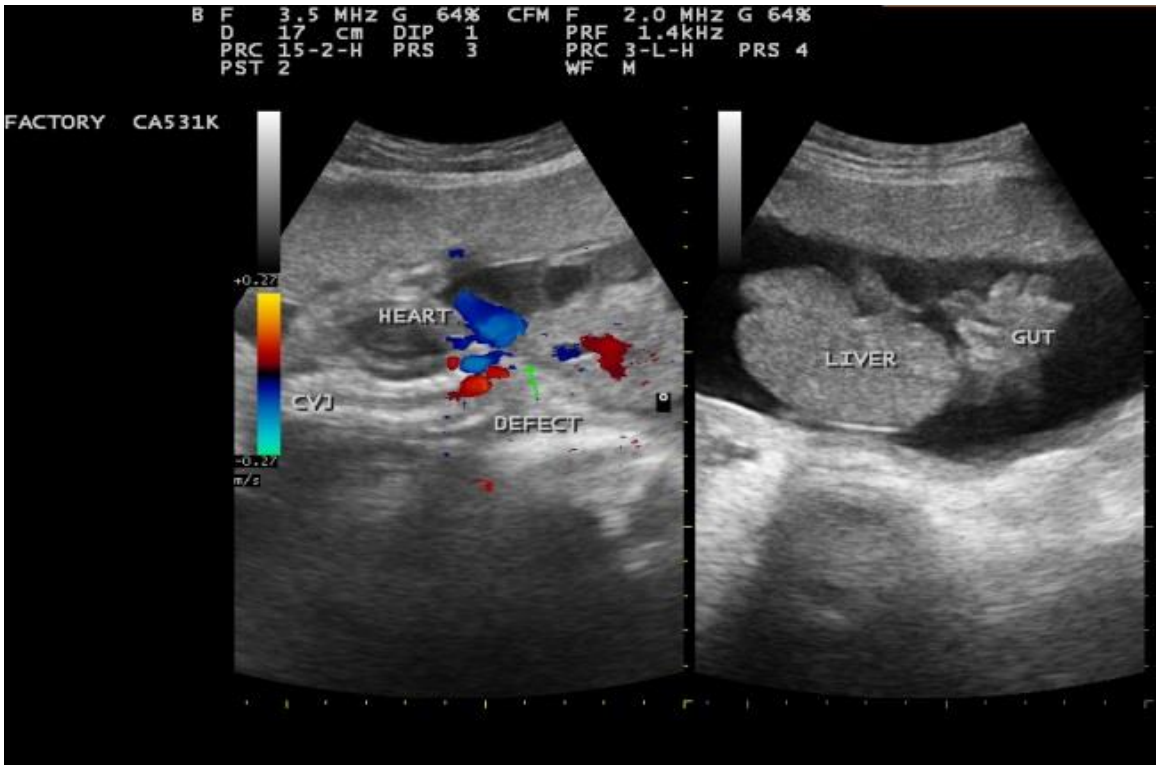


Fig 1b: Color/Pulsed Doppler waveform of umbilical artery of normal twin

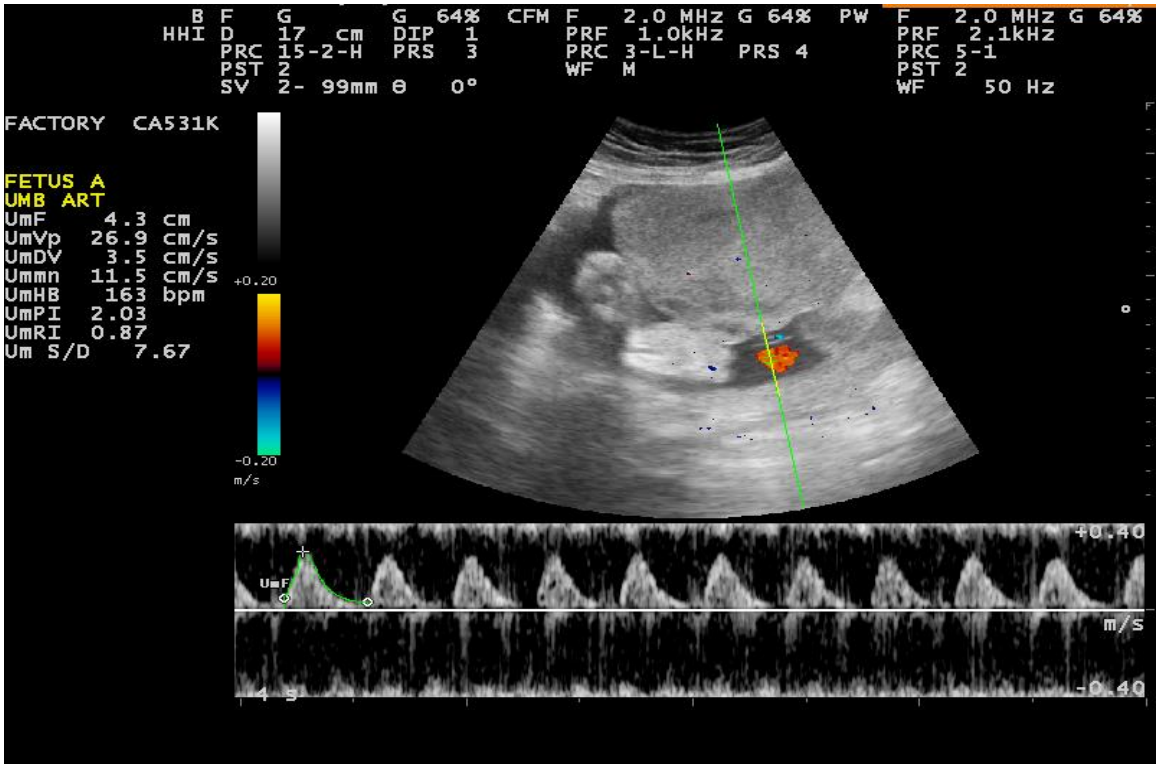


Figure 1c: Color/Pulsed Doppler waveform of umbilical artery of perfuse twin

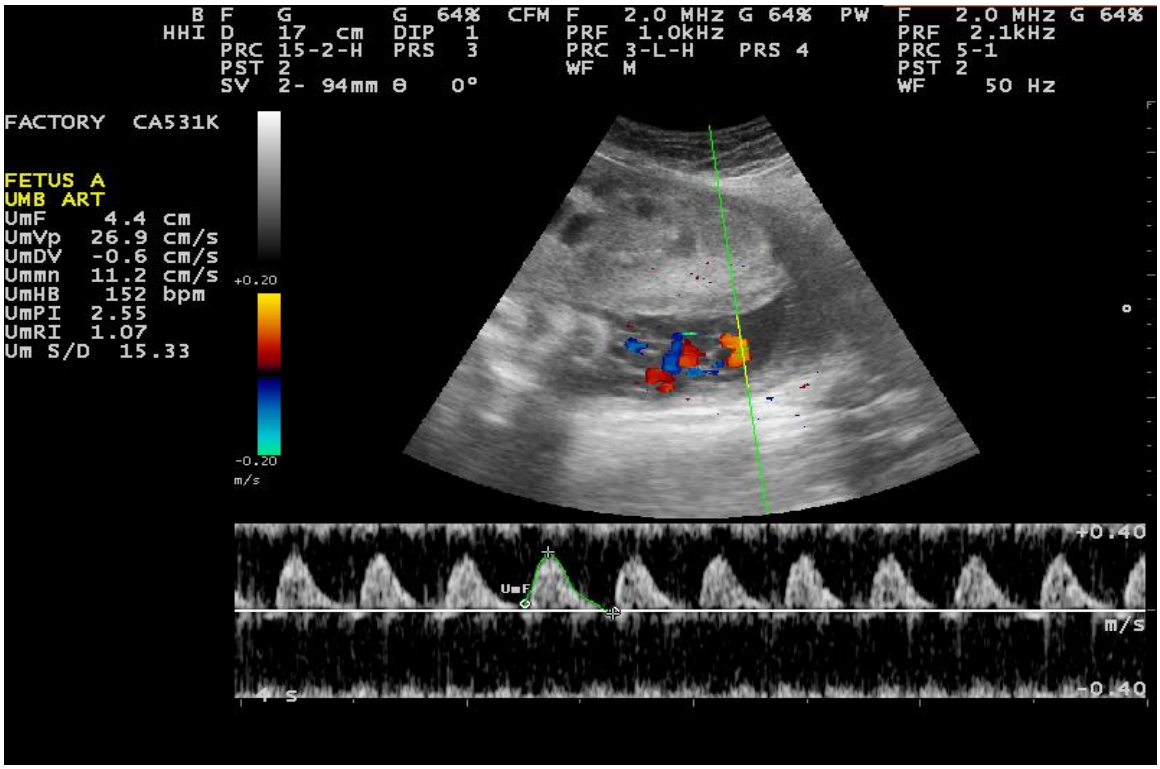
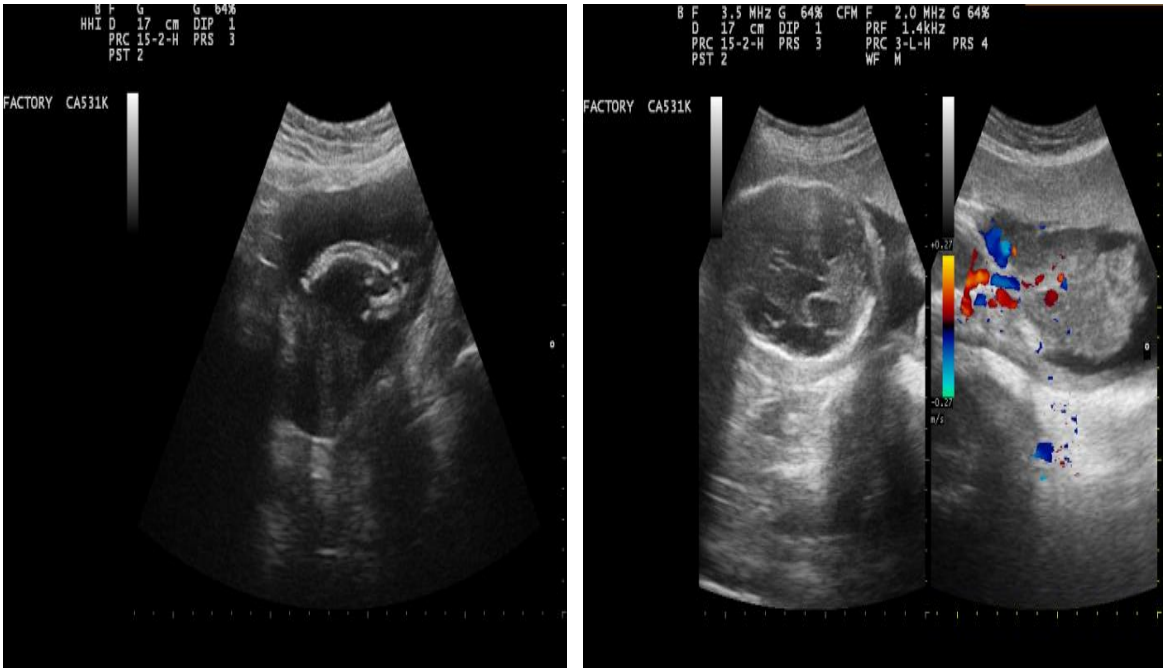


Figure 2a and 2b: Ultrasonography demonstration of acardiac heterogeneous twin and normal twin with normal heart and the connection between two on color Doppler study



TRAP was made and patient referred to obstetrics service for further management.

DISCUSSION

TRAP syndrome occurs in monochorionic gestation in which one twin has acardia (the recipient) with no heart and the other is structurally normal (pump or donor twin). Acardiac twin doesn't send blood to the placenta and all its blood comes from and goes back to the circulation of the pump twin through the vascular connections on the surface of shared placenta. Therefore, it is the structurally normal twin perfusing an anomalous recipient twin via an artery to artery anastomoses in a reverse direction. The reversed flow is through its umbilical artery and exits through umbilical vein, which is opposite to the normal fetal blood supply. The acardiac twin is usually grossly abnormal. The mortality risk of the pump twin is 50-75% due to the result of heart failure (likely due to polyhydramnios or excessive cardiac load) and mortality risk of the acardiac twin is 100%.

The upper half of the body of an acardiac twin is extremely poorly developed and, sometimes not developed at all. Head, cervical spine and upper limbs are usually absent. In contrast, the lower half of the body, although malformed, is better developed. Therefore, on USG, acardiac twin appears as a heterogeneous mass simulating a teratoma or intrauterine fetal demise [5].

The classification of acardiac twin is according to the degree of cephalic and truncal maldevelopment [6]. Various types are:

- Acardius acephalus: No cephalic structure present (head and upper extremities lacking). This is the most common type.
- Acardius anceps: Some cranial structures and neural tissue/ brain tissue are present. The body and extremities are also developed. It is highly developed form.
- Acardiusacromus: Cephalic structure present but no truncal structures are seen (head without a body). The umbilical cord is attached directly to the head. It is the rarest type of acardia.
- Acardia amorphous: No distinguishable cephalic or truncal structure. It is the least developed and not recognizable as a human form with minimal development. This differs from teratomas only by its attachment to an umbilical cord.

Another classification of acardiac twins is as follows:

- Hemi acardius: if the heart is incompletely formed.
- Holoacardius: if the heart is absent

The TRAP sequence is explained by the two main hypotheses:

- Deep placental anastomoses in early embryogenesis cause malformation of the acardiac twin. The early pressure flow in one twin exceeds that of other and leads to reversed circulation in the twin who exhibits perfusion.
- A primary defect in embryogenesis in one twin leads to failure of cardiac development. The normal twin perfuses the acardiac twin via artery to artery anastomoses. The anastomoses are not responsible for the cardiac anomaly [7].

Congenital anomalies are present in about 9% of pump twins [2].When the ratio of the weight of the acardiac fetus to the weight of the pump fetus is more than 70%, the incidence of preterm delivery is 90%, that of polyhydramnios is 40%, and that of congestive heart failure in the pump twin is 30%. In comparison, the corresponding rates are 75%, 30% and 10% respectively, when the ratio is less than 70% [2]. The weight of the acardiac twin cannot be measured by routine parameters and is calculated by the following formula:

$(1.2 \times \text{longest length}^2) - (1.7 \times \text{longest length})$ [8]

Prenatal diagnosis is performed using gray-scale and color-Doppler USG. TRAP syndrome is suspected when a twin gestation presents with discordance and bizarre malformations with retrograde blood flow in acardiac twin demonstrated by pulsed/ color flow Doppler [1]. The various prenatal treatment options for occlusion of blood flow to the acardiac twin so that normal twin is saved are;

- Endoscopic (fetoscopic) ligation of the umbilical cord.
- Laser coagulation of umbilical cord.
- Bipolar cord cauterization or
- Intrafetal radiofrequency ablation [9]

The various indications for prenatal treatment include:

- Polyhydramnios.
- Cardiac dysfunction of pump twin.
- Hydrops of the pump twin or
- Relatively large weight of the acardiac twin.

The obstetricians and radiologists need to be well versed with the condition so that timely diagnosis is made and the normal twin saved from complications.

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