Case Report

Twin Reversed Arterial Perfusion (TRAP) Syndrome - A Case Report

Hem Prabha Gupta¹, Iti Chowdhary², Shivani Singh³

¹Professor, ²Junior Resident, ³Assistant Professor,
Dept. of Obstetrics of Gynaecology, Era’s Lucknow Medical College & Hospital, Lucknow (UP) India.

Corresponding Author: Hem Prabha Gupta

Received: 06/04/2015        Revised: 29/06/2015        Accepted: 06/07/2015

ABSTRACT

Twin reversed arterial perfusion (TRAP) syndrome is a rare condition. It occurs in monochorionic Twin pregnancies resulting in Co-existence of a normal ‘pump’ twin and acardiac twin. In this case report, a 23 yrs old female gravida 2 para 1, with no living issue reported on 11.07.2014 as a case of 8 months pregnancy with previous caesarean section. In this institution her complete antenatal profile was done, ultrasonography showed twin gestation with one live fetus and another anomalous fetus. She was hospitalized for safe confinement and she was monitored by weekly colour doppler studies and biweekly non stress test. She had Caesarean section on 16.08.2014 as she started leaking and she was a case of high risk pregnancy. First baby was normal, second was a malformed fetus, developed only below the chest. Post natal and neo natal period of the first baby was uneventful. The patient was discharged on 10th post operative day in good health. The patient was asked for regular check up and till now the baby is growing well.

Keywords: Acardiac Twin, Fetal anomaly, Twin reversed arterial perfusion syndrome.

INTRODUCTION

TRAP occurs in monzygotic Multiple Pregnancies with an incidence of 1 in 35000 pregnancies. [1] TRAP occurs when one Twin is lacking a functioning cardiac system and it receives blood from the normally developing “Pump Twin” in a monochorionic pregnancy. This enormous demand of the “Perfused Twin” puts the normal pump Twin at the risk of cardiac failure and if not treated, the normal fetus dies in 50-70% cases. The ‘perfused twin’ which receives denatured blood by reverse flow from ‘pump twin’ develops various abnormalities and incomplete development.

The etiopathogenesis of this anomaly is abnormal placental vascular communication between the twins, leading
to imbalance of interfetal circulation. Reverse blood flow in the umbilical artery of acardiac twin causes atrophy of the heart and maldevelopment of the other organs. [2]

**CASE REPORT**

A rare case of twin reversed arterial perfusion syndrome was observed in Gynaecology and Obstetrics department of Era’s Lucknow Medical College and Hospital, Lucknow. The patient Mrs. N, 23 old woman, Gravida 2, Para 1, with no living issues, reported in the outpatient department for safe confinement on 11/7/14 with chief complaints of Amenorrhea 8 months. There was no history of bleeding or leaking per vaginum. Her abdominal findings were: Fundal height- 28-30 weeks size, Symphysiofundal height-30 cm, longitudinal lie, Cephalic presentation, Multiple fetal parts were felt. On auscultation only one FHS was localized (150 bpm) in right spinoumbilical line. Second FHS could not be localized. On general examination, patient was afebrile, Pulse rate-86/minute, Respiratory rate-20/min, Blood pressure-130/80 mm Hg. No pallor, icterus, cyanosis or lymphadenopathy was present and there was no oedema over feet. All the antenatal investigations were within normal limit.

**USG showed:** twin intrauterine pregnancy with one live intrauterine fetus in cephalic presentation and the parameters were corresponding to gestational age of 31 weeks with normal doppler studies. Another intrauterine fetus lying towards the right side of the uterine cavity was having malformed anatomy showing both thighs, both legs, lower vertebrae, lower abdomen with cord entering the umbilicus. There was abrupt ending of the fetal body at upper abdominal region and the head was not visualised. Acardiac acephalus fetus was diagnosed. The gestational age of this fetus was estimated by femur length only, it was 28 weeks of gestation. Single placenta with thin membrane in between the two fetuses was present. On the basis of clinical and radiological findings, final diagnosis of TWIN REVERSED ARTERIAL PERFUSION (TRAP) SYNDROME was made.

**Figure - 1 Normal Baby - the co-twin of Acardiac Acephalus Fetus**

**Figure - 2 Acardiac Acephalus Fetus with umbilical cord attached to common Placenta**

Patient was kept in the hospital for monitoring and her weekly colour Doppler studies and biweekly non-stress test was done and her coagulation profile was studied periodically as one fetus was dead. She underwent caesarean section for premature rupture of membranes at 35 weeks of gestation on 16/8/14. First baby was live and healthy weighing 2.03 kg (figure 1). Second baby was malformed - acardiac acephalus
developed below the chest only with cord attached to the common placenta. It weighed 1.2 kg (figure-2). Acardiac Acephalac Fetus had absence of upper part of the body with well developed lower extremities and genitalia (figure - 3). Skiagram of this fetus showed abruptly ending upper part of the vertebral column (Figure - 4). Fetal autopsy was not done as it was refused by the patient. First baby was born with Apgar score 10 without any congenital anomaly. Postnatal and Neonatal period was uneventful and the patient was discharged on 28/8/14. The patient came for follow up after 2 months and baby was growing well.

**DISCUSSION**

TRAP Syndrome is a rare Obstetrics condition unique to monozygotic monochorionic Twin pregnancies, in which there is co-existence of Normal pump Twin (donor) and acardiac recipient (perfused Twin). It is due to abnormal anastomosis between the interfetal circulation. As a result of this imbalance in the circulation, the caudal part of the perfused fetus receives blood with relatively more nutrients and oxygen than the upper torso, resulting in the better development of the pelvis and lower extremities of the perfused fetus. On the other hand, fully deoxygenated blood flows in the retrograde fashion to the upper part of the body resulting in maldevelopment of heart, head and upper torso which may be either completely absent or markedly deficient. [3] The various abnormalities which can result are:

- a) Acardiac anceps - when head is partly formed
- b) Acardiac acephalus - when head is absent
- c) Acardiac acormus - when only head is present
- d) Acardiac amorphous - when unrecognisable amorphous mass is present. [4, 5]

It has been hypothesized that TRAP sequence is caused by a large artery to artery placental shunt often accompanied by vein to vein shunt within the single shared placenta, therefore the perfusion pressure of the donor twin exceeds that of the recipient twin, who thus receives reverse flow of de-oxygenated blood from its co-twin. The used blood reaches the recipient twin through its umbilical arteries and preferentially goes to its iliac vessels, thus only lower body is perfused and disrupted growth of upper body results. The twin with normal cardiac activity may also be hydrops or malformed fetus in 9% cases and is at high risk for CHF which may result in preterm delivery and
perinatal mortality in approximately 55% - 75% of cases.

Acardiac fetus was first described by Beneditti in 1533 and Benedictus in 1539 and later by Geoffroy in 1836. [6]

Other theory postulates that there may be severe genetic or other primary cardiac embryogenesis defect. Chromosomal disorder has also been reported in 50% cases of TRAP syndrome. Numerous obstetrical complication have been reported, that are hydrops fetalis, polyhydramnios and preterm delivery. Recipient twin can have many abnormalities for example, absent cranial vault, absent facial structures, Anophthalmia and micro-opthalmia, absent liver and gall bladder, oesophageal atresia, absent lung and heart. [7]

Many authors have reported pregnancy outcome which can be influenced by the ratio of weight between the pump and recipient twin. When Acardiac and pump twin weight ratio exceeds 50% the prognosis for normal fetus is worst. [8] Although in our case the weight of normal fetus was 2.03 Kg (fig 1) and that of acardiac fetus was 1.2 Kg (fig 2).

Management of twin pregnancy with acardiac fetus is a challenge, as the continuous growth of acardiac fetus is deleterious to the normal fetus. Optimal management is controversial. Sullivan et al advocated expectant management in all cases. [9] They reported 90% survival in pump twin in 10 pregnancies with acardiac twin, managed expectantly, they cautioned against aggressive intervention and recommended expectant management with close fetal surveillance. Many methods of management have been suggested including termination of pregnancy. Serial ultrasound to monitor the well being and signs of decompensation is done, medical management and serial amniocentesis for the management of polyhydramnios and hysterotomy to remove the anomalous fetus. The aim of prenatal treatment is to stop blood flow to the acardiac twin without affecting the pump twin and to improve the outcome of the pregnancy. Plat et al in 1983 were the 1st to suggest the occlusion of circulation to the acardiac twin. [10] Minimally invasive interventions are through cord occlusions which have been attempted by Cord Ligation and laser coagulation, while intrafetal ablation is done by alcohol or monopolar diathermy and radiofrequency. [11] Interruption of vascular communication and the invasive treatment should be reserved for cases with poor prognosis for pump twin. In our case the conservative management was done by close supervision, serial ultrasonographic evaluation, Doppler studies of the fetus, weekly co-agulation profile, Biweekly Biophysical Profile study and non-stress test. The pregnancy was terminated for Obstetrical reasons. The indication for cesarean section was premature rupture of membranes with previous c-section with bad obstetrics history.

**CONCLUSION**

TRAP sequence is a rare complication of monochorionic multifetal pregnancy. An Acardiac Twin should be suspected in all monochorionic malformed fetuses. Ultrasound findings of twins showing discordant or malformation along with reverse flow in umbilical artery are diagnostic of an acardiac Twin. Expectant treatment with close supervision deserves consideration in all cases of TRAP sequence but the risk factors should always be kept in mind. The invasive treatment should be considered in cases with poor prognostic factors only, as it requires expensive equipments and trained personnel. Awareness of such cases amongst the Obstetrician, Paediatrician, Radiologist and Anesthetist is also important.
REFERENCES

How to cite this article: Gupta HP, Chowdhary I, Singh S. Twin reversed arterial perfusion (TRAP) syndrome - a case report. Int J Health Sci Res. 2015; 5(8):658-662.

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