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■ Case Report

Twin Reverse Arterial Perfusion Sequence (Traps): Acardius Acepalus with Externalized Intestines – A Case Report

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Abstract

Background: Twin reversed-arterial perfusion sequence (TRAPS) is a unique complication of monozygotic monochorionic multiple gestation, and the cause not known with certainty. In TRAPS, one twin develops normally ('pump twin'), whereas the other develops without normal heart (acardiac twin).-The 'pump twin' supplies blood to the acardiac twin and for itself via a retrograde blood flow through the umbilical artery. An acardius is therefore a parasitic twin since it receives its entire blood supply from the pump twin. Acardius acephalus, as a form of acardiac twin is a rare abnormality occurring in multifetal pregnancy particularly monozygous monochorionic twin gestation, and even rarer is the occurrence of externalized intestines with an associated omphalocele. It is usually associated with many structural malformations incompatible with intrauterine and extrauterine survival. Reports on acardius acephalus fetal anomaly are scanty. We report a case of acardius acephalus with co-existing omphalocele and externalized intestines in an undiagnosed twin delivered by caesarean section to a 34-year-old multipara who presented with reduced fetal movements. **Objective**: To report an acardius acephalic twin with omphalocele and externalized intestines as the second of an undiagnosed twin pregnancy delivered to a multipara by caesarean and review of important literature. **Method**: A case report of multipara who presented with reduced fetal movements and was diagnosed to have twin gestation with an acardiac twin and managed at the maternity unit of the Federal Medical Centre Gusau, Zamfara State in which relevant history, physical examination and radiological investigation were evaluated. The case was documented, and local and international literature search conducted and reviewed. **Conclusion**: Acardius acephalus may have additional malformations in the other organs. The current case, the second in our centre, shows yet other striking gross and severe malformations characterizing acardius acephalus.

Key words: Acardiac, acephalus, omphalocele, externalized intestines, twin reverse arterial perfusion sequence (TRAPS)

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INTRODUCTION

Monozygotic multiple gestations can result in a dangerous condition TRAPS, which affects 1 in 35,000 births[1] or 1% of monozygotic twins.[1-4] TRAPS, also known as the artery-artery twin disruption sequence, causes the development of the so-called acardiac twin, also referred to as the chorioangiopagus parasiticus, and the acardiac monster.[5] It has been proposed that in a monozygotic placenta with artery-to-artery and vein-to-vein anastomoses, blood is perfused retrogradely from the hemodynamically dominant twin (the 'pump twin') to the hemodynamically inferior twin (the 'recipient twin').[2] It is believed that postcapillary blood, which is deficient in

nutrients and oxygen, is pumped from the more typical pump twin into the acardius, where it deteriorates existent structures and disrupts the morphogenesis of developing structures.[5]

It has been suggested that TRAPS occurs because of occurrence of an omphalocele obstructing venous in the acardiac twin.[6] This suggestion has been refuted to play an aetiologic role despite the frequent occurrence of omphalocele in acardiac twins, however [6] indicated both reversal of the circulation and an omphalocele obstructing veneous return could contribute to the development of an acardius.[7] This condition is associated with a very high mortality rate of 50 to 70% in the pump twin and almost 100% in the acardiac twin.[8-10]

CASE REPORT

A 34-year-old booked $G_4P_2^{+1}$ who was unsure of date presented in the labour ward at term with a complaint of reduced fetal movements of 12 hours. She had no history of antepartum haemorrhage, drainage of liquor or febrile illness. Her first and last pregnancies were spontaneously conceived and normal. The babies were alive and growing normally. Her second pregnancy however was an uncomplicated spontaneous miscarriage at 8 weeks' gestation. She had no significant past medical and family history noted. At presentation, she was anxiously looking, not in any distress, not pale, afebrile, anicteric, not dehydrated, no pedal oedema.

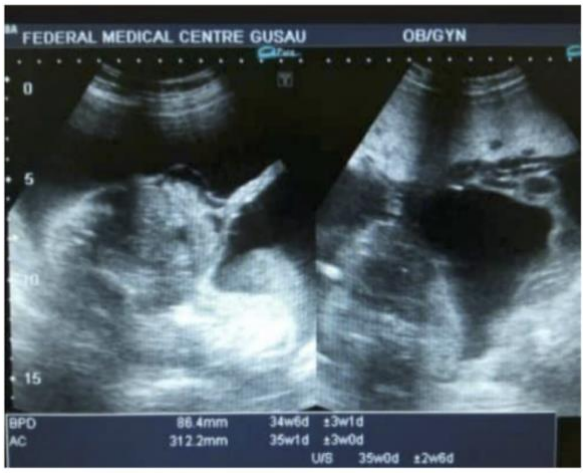

Fig. 1 Sonographic image of the acardius showing an omphalocele with the umbilical cord attached to the sac.

She had booked the pregnancy at a primary health care centre in the second trimester and had 2 visits that were uneventful. Her general condition was satisfactory, and her vital signs were normal. The abdomen was enlarged and shiny and tense, and the symphysio-fundal height was 40cm, fetal parts were difficult to palpate, and the fetal heart tone was distant. There was no uterine contractions and vaginal examination revealed no cervical changes.

An ultrasound scan revealed that the first twin was normal while the second malformed twin showed the absence of a head, chest, upper limbs, heart, and the presence of omphalocele major with the presence of loops of intestines. Polyhydramnios was also observed on ultrasound (figure 1). Cardiotocograph was non-reassuring.

In view of the non-reassuring CTG, the patient was delivered by an urgent caesarean section of a grossly normal male fetus with good Apgar scores and an acardiac macerated fetus. In the acardiac fetus, the structures superior to the abdomen were non-existent. The abdomen has externalized small intestinal loops because of the rupture of the omphalocele sac and there was no umbilical cord attached. It weighed 750g and had a rudimentary penis and no scrotal sac and anal canal. Both lower limbs were present, though the left was shorter, however, the digits in both feet were poorly defined (figure 2). The placenta was monochorionic with two umbilical cords attached and that of the acardiac twin measured 80mm.

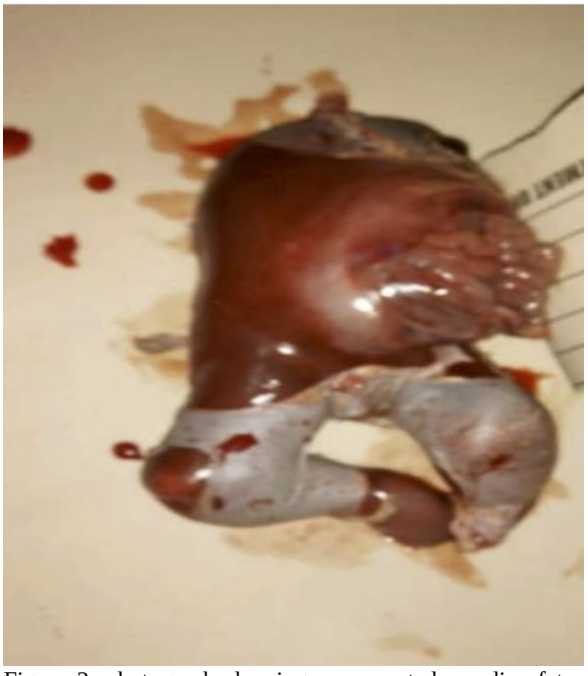

Figure 2. photograph showing a macerated acardiac fetus with externalized intestines and no attached umbilical cord.

The X-ray of the acardiac fetus showed absence of the head, neck and thoracic cavity and both upper limbs. The tibia and fibula of the left lower extremity as well as the ipsilateral hemipelvis were non-demonstrable, while the remaining bones showed normal density and outlines and soft tissue

irregularity and extrusion was noted on the right side of the abdomen. Given the gross anatomical and radiological findings described, we sub-classified the acardiac twin as an acardius acephalus with externalized intestines because of the ruptured omphalocele sac (shown in figure 3).

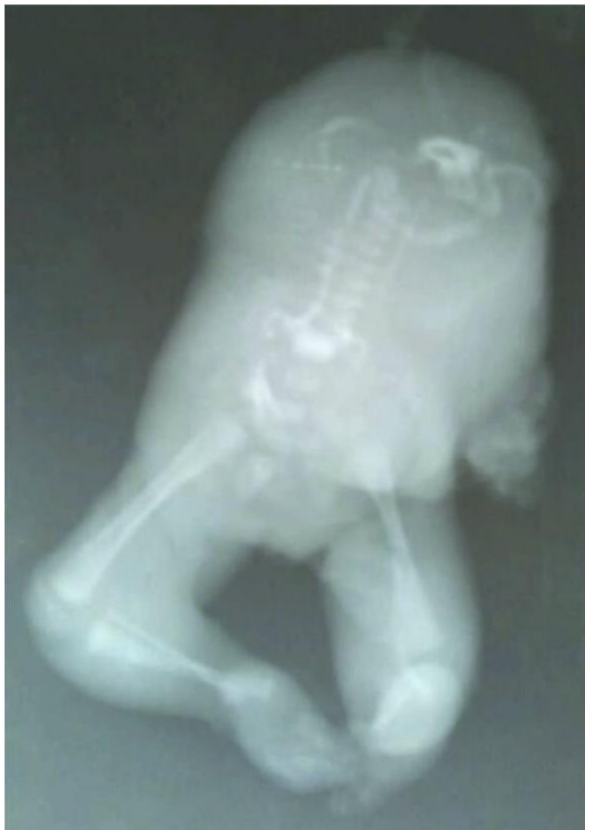

Figure 3. Roentgenogram of acardiac fetus showing malformed skeleton

DISCUSSION

In our department, this was the second case of acardius recorded, the first[8] was in 2015**.**

Monochorionic (MC) twins are the only type of twins with the complications of twin-twin transfusion syndrome (TTTS) because, in contrast to dichorionic twin pregnancies, the circulatory systems of the greater majority MC twins have placental anastomoses that result in complex vascular connections between the two foetuses.[11, 12] Artery-to-artery anastomoses, which are less common than arteriovenous connections, result in severe abnormalities when the perfusion pressure of one twin surpasses that of the other, resulting in reversed arterial flow. The recipient's iliac vessels receive blood from the donor twin, causing the lower body to be well-perfused than the upper body. This disorder known as the TRAPS causes the acardius fetal malformation.[11]

Acardiac twins demonstrate a complex constellation of structural malformations and these were thought to occur as a consequence of reversed blood flow and eventually causing deterioration of pre-existing structures and disruption of the morphogenesis of developing structures due to result of delivery of oxygen and nutrient-poor postcapillary blood to the acardiac twin.[10] Although chromosomal abnormalities have been identified in some acardiac fetuses, acardiac twinning is believed to be a developmental rather than a genetic disorder since the co-twin is usually morphologically and genetically normal.[13] No two twin fetuses have the same morphological appearance due to the variability in the timing of disruption of morphogenesis**.**[8]

The skin appears to be the sole structure consistently present in an acardiac twin; however, hair, upper and lower limbs long bones, vertebrae, and the large intestines are often present, other organs are rarely present in acardius.[5]

Several sub-classification patterns exist for acardiac twins with each categorizing the various combinations of dysmorphogenesis to different extents. Acardiac twins are classified[3, 5, 13, 14] into pseudo-acardiac (hemiacardius) – when the heart is incompletely developed or holoacardius - heart is absent. Other classification includes acardius ancephs – when the head is poorly developed, acardius acephalus -head is absent, acardius acormus – a rudimentary head, and acardius amorphous – an indistinguishable amorphous mass or a structure that has no resemblance to typical human form. Another subclass was described by Simmonds and Gowen as cardius mylacephalus—amorphous mass with some level of development of one or more extremities.[5] The index case fits into the acardius acephalus type with omphalocele and externalized intestines.

Reports of acardiac twins with externalized intestines are rare and to the best of our knowledge only one case was reported by Shawn et al and they hypothesized that the acardiac twin might have developed an omphalocele early during pregnancy.[5] TRAPS can lead to a wide range of pregnancy complications such as polyhydramnios, preterm labour, cord accidents, dystocia, uterine rupture, hydrops fetalis, intrauterine fetal death, preterm birth, fetal cardiac failure, anaemia, TTTS.[5, 10] The complications in the index case where polyhydramnios, fetal compromise in the pump twin and IUFD in the acardius.

Thus, early detection of an acardius by

ultrasonography and monitoring of the normal twin's cardiac status and well-being is essential.[8, 12] Several interventions including radiofrequency ablation and intrafetal laser have been used for in-utero interruption of the placental vascular anastomoses or selectively terminating the acardiac twin.[1, 2, 11, 12, 15] However, monochorionic twins complicated with an acardius do not necessarily need invasive treatment all the time.[11, 15] The choice of treatment is dependent on the size of the acardiac twin in relation to the 'pump' twin[15] as seen in the index case, although had presented late, where the acardiac twin was small resulting in reduced stress on 'pump' twin's heart with consequent improved survival of the latter without need for invasive treatment.

Another factor to consider in treatment choice is the presence of any form of cardiovascular impairment in the pump twin and the treatment should be considered before 16 weeks gestational age and ideally in centers with expertise in such treatments.[11, 15]

CONCLUSION

Complex structural malformations in acardius acephalus remain inconsistent and incompatible with life. The index case, the second in our Centre demonstrates additional abnormality in the form of externalized intestines from ruptured omphalocele underscoring the diverse malformation characterizing acardiac twins.

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