

Case Report

Acardiac twin – A unique complication of monozygotic twin

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Acardiac foetus also referred as Twin Reversed Arterial Perfusion (TRAP) sequence is a unique complication of monozygotic twin where the parasitic twin fails to develop heart and upper part of the body. It is thought that the acardiac foetus develops from reversed circulation of artery-to-artery or vein to vein anastomosis without any vascular communication with the placenta. It has an incidence of approximately 1% of monozygotic twins or 1 in 35,000 deliveries. To report a rare event- acardiac foetus born with a monozygotic co-twin in 2008, Dhaka National Medical College and Hospital. An acardiac amorphous twin was delivered by caesarean section at 30 weeks of gestational age with an asphyxiated living co-twin weighing 1000gm. The normal co-twin was apparently devoid of any gross congenital anomaly but expired 48 hours after birth. As the case was undiagnosed prior to delivery, no antenatal attempt was taken to secure the healthy baby's life by interrupting affected twin's circulation.

Keywords: Acardiac foetus, monozygotic twin, circulatory intervention

INTRODUCTION

An acardiac twin is a serious and unique complication of monochorionic multiple gestation. This rare incidence occurs one in 35,000 deliveries and in one in 100 monozygotic twins (Benirschke and Kim, 1973). It is a Twin Reversed Arterial Perfusion (TRAP) sequence, where the acardiac twin usually has a normally formed co-twin and receives blood flow from the heart of the normal partner. The recipient twin is without a normal heart (acardius) and missing other various structures (Van Allen et al., 1983). It occurs due to circulatory disturbance at the time of early embryogenesis. Acardia results from an artery to artery, often accompanied by vein to vein placental shunt, whereby the arterial pressure of one twin overpowers that of other twin. Arterial blood flow of the first twin perfuses the second twin, whose blood then flows in reverse (Jones, 1997). The blood reaching the recipient twin preferentially goes

to the iliac vessels and thus perfuse only the lower part of the body, leading to disorder of the growth and development of the upper part of the body, thus fails to develop head, arms and a heart. Lower limbs or limb-bud may be seen. Tissues of internal structure like, lung, brain matter and bones can be found. Failure or disrupted growth of the head is called acardius acephalus; a partially developed head with identifiable limbs is called acardius myelacephalus; and failure of any recognizable structure to form is acardius amorphous. It survives by leeching blood from the co-twin. The normal foetus pumps blood for itself and its acardiac twin, this causes extreme stress on normal twin's heart. The circulatory load may be so large that the normal twin eventually develops high output heart failure and death may occur. Without treatment the pump twin dies 50 to 70 percent cases (Specific types of parasitic twin, Acardiac twin or

Figure1a



Figure1b



Figure1c



Figure 1(a-c). Showing ultrasonographic images of acardiac foetus (here it was interpreted as demise foetus) and its normal viable co-twin. (a) Showing the head circumferences of both the acardiac foetus and the normal co-twin. Head circumference of the acardiac foetus is incomplete. (b) Abdominal and head circumferences of normal co-twin. (c) Thoracic and abdominal cavity of the acardiac foetus. Lung is seen in the thoracic cavity and absence of cardiac pulsation. There was moderate amount of ascitic fluid in abdominal cavity. Head circumference was collapsed due absence of major part of skull bone.

TRAP sequence, Wikipedia, the free encyclopedia).

CASE REPORT

A lady of 26 years normotensive, nondiabetic of middle

class socio-economic family, para-1, gravida-2nd was referred to Dhaka National Medical College Hospital at her 30 weeks of pregnancy with labour pain. Her pregnancy was spontaneous. It was not a booked case. An ultrasonography was done according to the advice of a general practitioner to see the pregnancy profile which

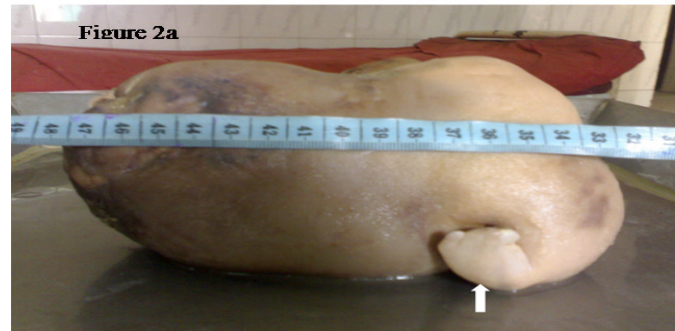


Figure 2(a-c). Are the pictures of the amorphous mass of acardiac twin: (a) the length of the acardiac foetus (15 cm) and arrow showing a foot at one end. (b) Straight arrow shows head like structure of the acardiac foetus without any facial structure, presence of hair. Curved arrow shows jelly-like material present on the body. (c) Ventral aspect of the acardiac foetus.

was interpreted as twin pregnancy with single foetus demise. On examination, no systemic abnormality was found to the patient. Per abdominal examination revealed her symphysis-fundal height 35 cm. Foetal heart sound was auscultated at one site (at right hypochondrium) of the abdomen. Labour was prolonged and maternal distress appeared. Ultimately the babies were born by Caesarean section. The first baby was an acardiac foetus. It was a structure less; amorphous fleshy mass about 15 cm in length. Its cardiac activity, definite head, facial structure and extremities were absent. A foot-like structure was present at one pole. Hair was present on the head like pole. There was a jelly-like substance at one part of the body. No distinct umbilical cord

connection of the acardiac foetus was found. Autopsy of the acardiac twin was performed which revealed part of the cranium, brain matter, lung tissue, urinary bladder and rudimentary pelvic bone and absence of heart.

The living foetus was in another amniotic sac. It was a male foetus of 1000g with APGAR score 4 in 1st minute and 6 in 5th minute. There was a single placenta with one cord attached centrally. Apparently the living baby had no congenital anomaly but asphyxia was present and the baby expired 48 hours after birth in Neonatal Unit. The mother developed no complication and was discharged from the hospital on the 6th postoperative day of caesarean section. (Figures 1, 2, 3, and 4)

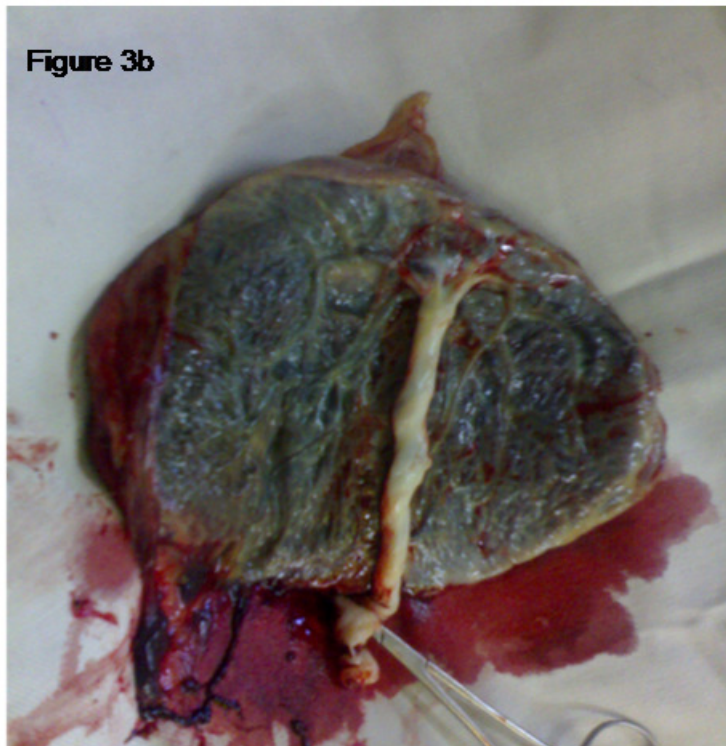


Figure 3. Showing (a) the living co-twin and (b) the single placenta with single umbilical cord

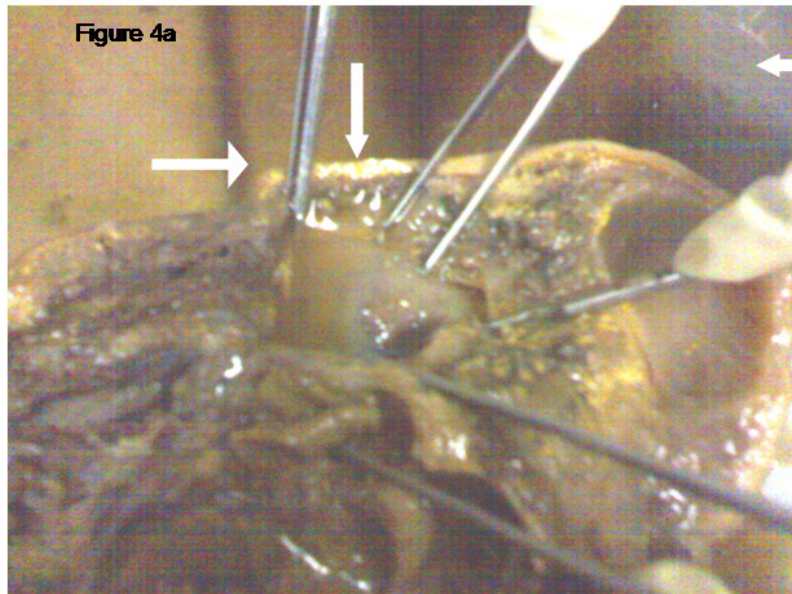


Figure 4a and b. Are the pictures of the autopsy of acardiac foetus, (a) arrow from right, left and above showing urinary bladder, lung tissue and site where heart is absent. (b) Bisectioned head with brain matter.

DISCUSSION

An acardiac foetus can be diagnosed by anomaly scanning in pregnancy. But sometimes an acardiac twin may be mistaken for a dead anomalous twin, especially when the cranial structures are present. On the other hand, when the acardiac twin has minimum resemblance to a human form, confusion with amniotic band syndrome can occur. The present case was not detected as an acardiac foetus during antenatal period, as it was an unsupervised case and the only ultrasonography done in the second trimester of pregnancy was misinterpreted as twin pregnancy with single foetus demise.

The acardiac twinning, also referred as the Twin reversed arterial perfusion (TRAP) sequence likely

represents an extreme manifestation of twin transfusion syndrome (Al-Malt et al., 1991). An acardiac twin is perfused by normal co-twin by means of reversal of circulation through large artery to artery and vein to vein anastomosis and has no direct communication with the placenta (Nyberg et al., 1990; Izquierdo et al., 1991, 1992). In present case no vascular communication was found between the acardiac foetus and placenta and presence of one umbilical cord was seen connected with the single placenta and normal co-twin.

When one of the twins is acardiac the normal foetus is only expected to have a 20 percent or less chance of survival. The prognosis is directly related to the respective weight of the recipient to the pump twin. The higher the weight of the recipient twin, the more likely is

the development of cardiac insufficiency and mortality in the pump twin (Moore et al., 1990). The complication of the pump twin of an acardiac foetus includes congestive heart failure, polyhydromnios, preterm labour and death in 50-70 percent cases (Sonneveld and Correy, 1992). Here also the recipient twin dies soon after birth due to prematurity and may be due to heart failure.

To save the life of a normal co-twin, circulatory anastomosis needs to be interrupted. Multiple methods of interrupting the affected twin's circulation have been attempted with variable success. These include endoscopic ligation of umbilical cord, Foley cordosats, needling to block umbilical cord circulation with coils or fibrin or glue material (Holzgreve et al., 1994). However, these are highly invasive; require expensive equipments and highly skilled operators. A simpler technique was developed for the intrauterine management of acardiac twins by injecting multiple pieces of standard surgical suture soaked in 96 percent ethanol into the umbilical cord. Finally Sepulveda reported the successful treatment with the injection of absolute alcohol, which is a potent sclerosing agent into the intra-abdominal portion of the umbilical artery (Sepulveda et al., 1995). Currently, Laser or bipolar coagulation is the method of choice in second half of the pregnancy. Recently, radio frequency ablation appears successful with minimal complication. All these techniques lead to increased survival of remaining twin and reduced risk of maternal complication. However, the injection of absolute alcohol into the intra-abdominal portion of the umbilical artery is minimal invasive, simple, safe and low cost with high efficacy. Experience suggests that this method may be one of the best choices for the intrauterine treatment an acardiac twin. Unfortunately in present case acardiac foetus was not diagnosed during antenatal period and no attempt was taken to interrupt the affected twin's circulation.

CONCLUSION

Acardiac twin is a rare event. Last 25 years record of Dhaka National Medical College Hospital shows no incidence of acardiac foetus except the present case.

As the prognosis is mostly fatal on the part of the co-twin, therefore it requires early identification during antenatal check-up and counseling of parents. Affected twin's circulation should be interrupted in-utero by method that is minimal invasive, simple, safe, low cost with high efficacy. At the same time, intensive foetal surveillance for the living baby is necessary.

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