Online Journal of Health and Allied Sciences
Peer Reviewed, Open Access, Free Online Journal
Published Quarterly : Mangalore, South India : ISSN 0972-5997
Volume 12, Issue 2; Apr-Jun 2013

Case Report:
Acephalic Acardiac Fetus

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Citation
Wader JV, Kumbhar S, Whawal V, Joshi SS. Acephalic Acardiac Fetus. *Online J Health Allied Scs.* 2013;12(2):15. Available at URL: http://www.ojhas.org/issue46/2013-2-15.html

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Submitted: Apr 11, 2013; Accepted: Jun 30, 2013; Published: Aug 25, 2013

Abstract: Acardia (Twin Reversed Arterial Perfusion Sequence) is a rare & one of the most bizarre congenital malformations occurring in multiple pregnancies. It affects 1 in 35,000 pregnancies. Usually one twin shows absence of heart & other is structurally normal. The acardiac twin presents with many bizarre anomalies such as lack of head, thorax & upper extremities. We report a case of autopsy study of acardiac twin with severe malformations. The case has been presented for its rarity.
Key Words: Acardia; Congenital malformations; Autopsy

Introduction
Acardia (complete absence of the heart) also referred as the twin reversed arterial perfusion sequence (TRAP) is a rare & one of the most bizarre congenital malformations occurring in multiple pregnancies.
The incidence is 1 in 35,000 pregnancies in general & 1 in 100 monozygotic twin pregnancies.[1]
Usually one twin has acardia & the other is structurally normal.
The acardiac twin (the recipient) is transfused by the normal co-twin (the donor or pump twin) through arterial to arterial & venous to venous anastomosis between the twins.[4] This blood flow is characterized by reversed arterial perfusion. Hence the name TRAP. The acardiac twin presents with many bizarre anomalies such as lack of head, thorax & upper extremities.

Case Report
We report a case of autopsy study of an acardiac twin with severe malformations.
A 23 years old female 2nd gravida with 29 weeks gestation (has one live issue) was referred to the department of Obstetrics- Gynecology. The ultrasonography (USG) report documented intrauterine twin pregnancy with one fetus of 24 wks gestation & the other showing absent head & a large multiloculated cyst engulfing the thoraco-abdominal cavity of the fetus.
Repeat USG revealed both the fetuses to be dead & confirmed rest of the findings. There was no history of consanguineous marriage & no any bad obstetrics history. Pregnancy was terminated by instilling ethacridine (Emcredil), and of the expelled twins, one was anomalous and the other was without any anomaly.
Autopsy study of the anomalous fetus revealed a grossly abnormal baby with easily identifiable lower limb. A large fluid filled cystic sac measuring 8 x 7 cm is seen in place of head & thorax. Head & thoracic region could not be identified. Baby weighed 150 gms & measured 35 cm in length. Upper limb were represented by arms & hands with no forearms. Both sided upper & lower limbs revealed club-hands & club- feet. Some of the digits were missing. External genitalia were poorly developed.
The big sac was filled with clear fluid along with some of the abdominal organs viz. intestines, both the kidneys & both the testes. Rest of the abdominal organs viz. liver, spleen, pancreas, adrenals, stomach & all the thoracic organs including heart & major blood vessels were absent. Brain tissue was not identified both grossly & microscopically. (Image 1) X – ray of whole body of the fetus revealed absent skull, cervical spines & both sided radii & ulnae.(Image 2)

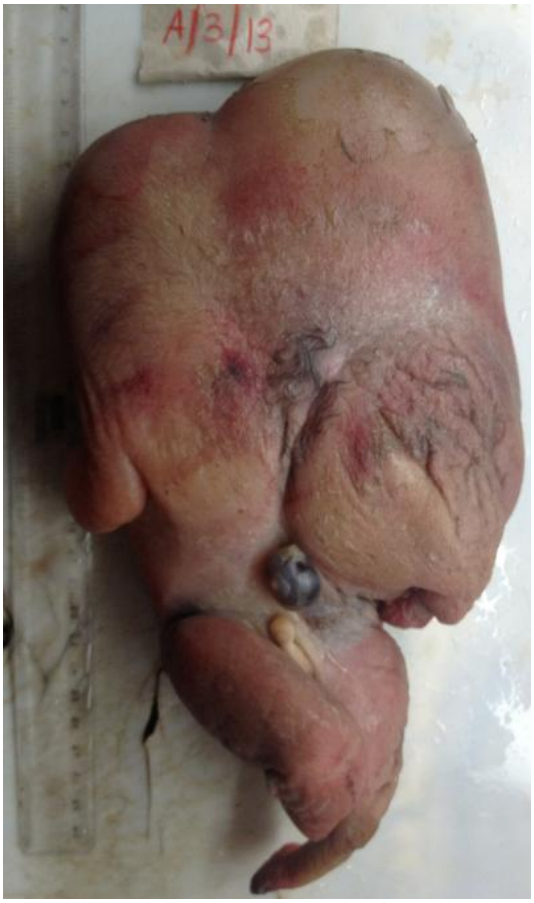

Image 1: Acephalic, acardiac, fetus

The fetus was diagnosed as Acephalic Acardiac Fetus.

Discussion

Acardiac monster was 1st described by Beneditti in 1533. Twin reversed arterial perfusion (TRAP) was defined by Greenwald in 1942.[6] Fnutiger (1969) reported that acardias were predominantly female. Spence (2001) found only 29% cases to be female.

The acardiac twin (recipient twin) is a haemodynamically disadvantaged develops severe anamolies that are incompatible with life.

Four categories of this condition have been described depending upon the degree of cephalic & trunkal mal development.[7] The earlier is the gestational time more perfusion occurs, the more severe damage to the affected fetus is seen.

1. Acardiac Anceps: This lesion has relatively well developed fetus. Parts of the brain, skeleton of face, meninges & scalp hairs are present. Entire Vertebral column is intact with all four extremities. Oesophagus, stomach, intestines, lungs, adrenals, kidneys, bladder, testicles & ovaries are represented. Diaphragm is absent. Remnants of heart present. Large fluid filled spaces cervical hygromas) on either side of cervical vertebrae are seen.

2. Acephalic Acardiac Fetus: This lesion has absence of head & thoracic organs. Ribs are usually present & Thoracic vertebra are either missing or decrease in number. Remnants of liver, spleen, kidney & intestines are present. Pelvis & lower extremities are usually well developed but arms may be absent.

3. Acardiac Acormus: Rarest lesion. Also called as Bodyless Head.

4. Acardiac Amorphus: This lesion has lump of tissue coverd by skin.It contains bone,cartilage,fat,muscles.In this lesion well developed forefoot with 5 toes , hyperplastic tibia ,normal size femur & part of bony pelvis & vertebral column are present. Generalised odema with several prevertebral lymphatic cysts are also present.

The present case had the features of Acephalic Acardiac Fetus.

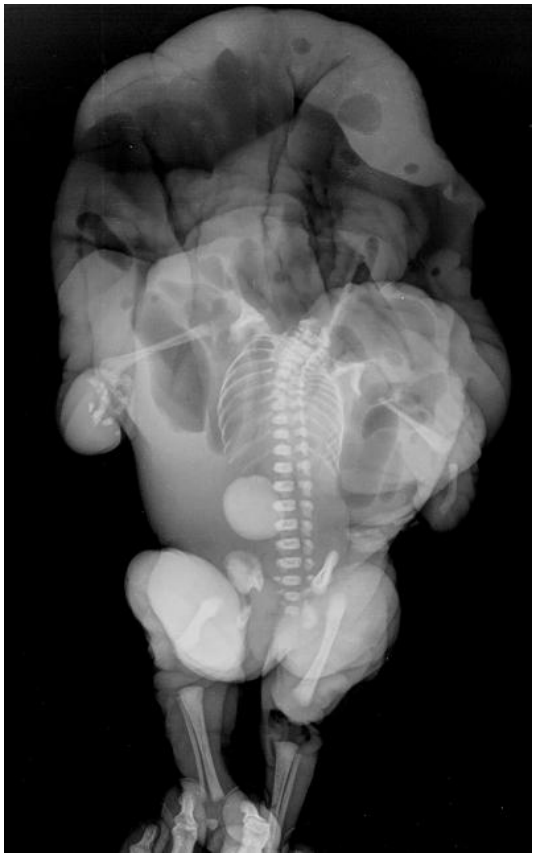

Image 2: X – ray of whole body of the fetus confirming absent skull, cervical spines and radii & ulnae.

Pathophysiology: TRAP sequence is believed to result from abnormal placental vascular anastomoses between the twins leading to imbalance of interfetal circulation.

Of the several theories put forth to explain the abnormal circulation in this syndrome, the most widely accepted explanation is that there occurs sudden reversal in the direction of blood flow within vessels of the affected embryo or fetus and the blood passes through arterio-arterial anastomosis in the placenta retrograde through the umbilical artery to arterial system of the damaged embryo. Then it goes via venous pathways through the venous system, the umbilical vein & placental veno-venous anastomoses back to the normal twin. The pump twin supplies deoxygenated blood via vascular anastomoses to acardiac twin. Inadequate perfusion is thought to be responsible for development of anomalies.

Chromosomal abnormalities associated with such lesion of Acardia includes Trisomies & mosaicism of the affected twin & Donor twin has normal karyotype.[7]

With the use of ultrasonography, transvaginal doppler ultrasonography & doppler velocimetry diagnosis of acardia is possible even in the first trimester of pregnancy by detecting inversion of blood flow in the recipient acardiac fetus (Bonilla Musoles et al).[1]

Most common differential diagnosis for acardiac twin is hydropic, recently demised twin. This condition is excluded

by documentation of blood flow to the twin & the growth of lower extremities. Another differential diagnosis is teratoma. Identification of umbilical cord and developed organs in acardiac twin will exclude teratoma.

Mortality of acardiac twin is universally fatal either in utero or at the time of delivery. Normal pump twin is at increased risk because of high cardiac output, congestive cardiac failure, hydrops and polyhydraminios.[2] Normal fetuses may die of prematurity & respiratory distress syndrome (RDS). The overall mortality of pump twin is approximately 50-75%.[5]

Both fetuses in the present case however died in utero.

Management for the pump twin includes tocolytic agents and volume reduction amniocentesis to treat preterm labour and frequent interval monitoring studies for detection of signs of congestive cardiac failure. If the fetal distress occurs the invasive procedures like hysterotomy ligation of the cord of the acardiac twin or radiofrequency ablation are done.

Early diagnosis of TRAP & appropriate treatment will definitely improve the outcome.[3]

Acknowledgements:

We thank Dr. Sahoo, Head of Radiology, KIMSDU, Karad, for the radiological images.

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