University Heart Journal
Vol. 9, No. 2, July 2013

CASE REPORTS

Ectopia Cordis

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Abstract:

Ectopia cordisis characterized by partial or complete displacement of the heart out of the thoracic cavity The defect is a rare congenital abnormality, occurring in 5.5 to 7.9 per 1 million live births.[1]

Case Report:

A 2650-g female neonate was born on Sept 16, 2014 who presented with an anterior thoracic defect with complete extra thoracic heart. Her gestational age was 38 weeks. The infant was delivered by caesarean section in a medical centre at Manikgonj, Bangladesh. She was transferred immediately to the National Institute of Cardiovascular Diseases, Dhaka.

The infant had an extrathoracic heart covered only by visceral pericardium (complete thoracic ectopia cordis).The sternum was completely splitted with an inter-ridge distance of 5-6 cm, through which the heart was protruding for 4-5 cm and the apex pointing anteriorly.

Since her birth, her activities are normal in relation to any other newborn infants including breast feeding, hands and feet movements etc. Her urinary and bowel systems were functioning normal.

Initially her heart was covered with saline-soaked gauze pads& systemic antibiotics were given.

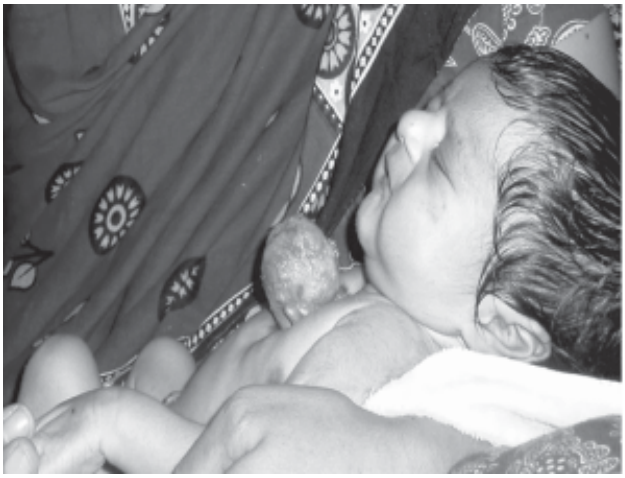
Direct echocardiography showed single ventricle, grade-111 tricuspid regurgitation with severe pulmonary arterial hypertension (PASP-78).

On her 3rd day of birth at the National Institute of Cardiovascular Diseases, there was bleeding from her visceral pericardium for 5 minutes and she lost approximately 200ml of blood.

After 15 days again bleeding occurred followed by a loud scream and cry in the beginning but this time bleeding lasted for 15 minutes and loss of around 400ml of blood. After her blood loss she became very weak and pale. Nutritional support was given by parenteral nutrition.

Her complete blood count showed normal at that time. After 1 hour she was haemodynamically stable and began to intake breast milk of a very low quantity. She resumed her normal activities like before after a total of 6 hours approximately.

Her parents were poor & her condition was very critical. Unfortunately the baby died on her 20th days of birth.



Discussion:

Ectopia cordis is a very rare congenital heart defect, which was first observed 5000 years ago.[2] The defect is described as malposition of the heart, partially or completely outside the thorax. When a multiple pregnancy exists then one fetus is affected only.[3]

Ectopia cordis is rarely associated with chromosomal abnormalities. Its etiology is unknown.[4-6]

It can be classified into five types:

1) cervical, 2) thoracic- cervical, 3) thoracic, 4) thoraco-abdominal, 5) abdominal.[7,8]

Our case seemed to be the thoracic type. The following characteristics were observed in our patient: completely bifid sternum, anterior extrathoracic heart with a univentricular heart & grade -III TR with severe pulmonary arterial hypertension, absence of parietal pericardium.

Ectopia cordis is usually associated with other intracardiac defects like ventricular septal defect, atrial septal defect, tetralogy of Fallot, and diverticulum of the ventricle are the most commonly encountered heart lesions.[9,10]More severe & complex intracardiac defect associated with this malformation leads to very poor prognosis.[9]

This defect may also be associated with other congenital anomalies like anencephaly, hydrocephaly, cranial & facial malformations, cleft lip or palate, abdominal wall defects, neural tube defects, genitourinary or gastrointestinal malformation etc.[1,11,12]

Ectopia cordis can be diagnosed during pregnancy by an ultrasonography, an early plan should be made for elective caesarean section. After delivery infant's heart should be covered with sterile saline soaked gauze pads.

Specific management of ectopia cordis are repair of the intra cardiac defect, placement of the heart into the thorax, repair of the associated abdominal anomaly, then closure of the chest wall defect.

In spite of surgical treatments, few patients with these cardiac malformations survive and most of them die in the first few weeks of life.[13]The prognosis of this condition is usually poor.[14]

Conclusion:

Ectopia cordis is a rare congenital malformation with very few reported survivors after surgical correction.

Despite a poor outcome, surgical repair may have to be attempted in the neonatal period. As The infants who undergo successful surgical repair have chances to survive.

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