Thoraco-Abdominal Ectopia Cordis: A Rare Entity. Case Report and Review of Literature.

Ashwin V Apte

Department of Surgery, People’s college of Medical Sciences and Research Center, Bhanpur, Bhopal-462 010.

Abstract:
Ectopia cordis is a rare congenital abnormality characterised by partial or complete displacement of the heart outside the thoracic cavity. It can be associated with other congenital abnormalities. The present case was a two hours old male child and a product of non-consanguineous marriage. Child had thoraco-abdominal ectopia cordis. Condition of child detoriated rapidly and succumbed before any investigation and surgical intervention could be performed.

Key Words: Ectopia Cordis Thoracoabdominalis, Sternal Cleft.

Introduction:
Ectopia cordis is defined as complete or partial displacement of the heart out side the thoracic cavity. It is a rare congenital defect in fusion of the anterior chest wall resulting in extra thoracic location of the heart (Hornberger et al, 1996; Kim et al, 1997). It occurs in 5.5 to 7.9 per 1 million live births (Hornberger et al, 1996). This anomaly is classified into five types: cervical, cervicothoracic, thoracic, abdominal, and thoracoabdominal (Kim et al, 1997). The two most common forms of ectopia cordis are the thoracic and thoracoabdominal type (Amato et al, 1995; Hochberg et al, 1995). The thoracic ectopia cordis constitute the classic naked heart with no overlying somatic structures. The thoracoabdominal type is frequently associated with Cantrell’s pentalogy, which include bifid sternum, deficiency of the diaphragm, defect of diaphragmatic pericardium, defect of the anterior abdominal wall, and intracardiac defects (Amato et al, 1995; Abdallah et al, 1993).

Case Report:
A fullterm 2 hours old male boy weighing 2.4 kg was shifted to our hospital with Ectopia Cordis. He was born to a 26 year old mother by caesarian section due to fetal distress. There was no history of consanguineous marriage, antenatal infection, drug ingestion, exposure to radiation etc. Preliminary examination of the baby revealed peri-oral cyanosis with heart rate of 140/min and respiratory rate of 50/ min. The heart of baby was lying outside the thoracic cavity & devoid of pericardium with a Sternal Cleft. The apex of heart was pointing anteriorly. Baby also had associated supraumbilical omphalocele which contained the liver and bowel with a thin membrane covering the omphalocele. The sternum was bifid in lower half, with an inter-ridge distance of 5 cm, through which the heart was protruding for 4-5 cm. (Fig. 1).

Baby was resuscitated on admission and initial management included covering of the heart and omphalocele with sterile-soaked dressing and systemic antibiotics coverage.

The baby deteriorated rapidly and inspite of supportive management baby unfortunately died within few hours of birth before any investigations and surgical intervention could be undertaken. At our hospital performing a medical autopsy is not a routine practice & obtaining consent for the same is difficult,
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Therefore, the underlying cardiac defects and associated malformations could not be ascertained.

Discussion:

Ectopia cordis is a rare and striking congenital heart defect, which was first observed 5000 years ago (Taussing, 1982). The term ectopia cordis was first coined by Haller in 1706. The defect is described as malposition of the heart, partially or completely outside the thorax. According to the position of the misplaced heart, ectopia cordis can be classified into five types: 1) cervical, in which the heart is located in the neck with sternum that is usually intact; 2) thoracocervical, in which the heart is partially in the cervical region but the upper portion of the sternum is split; 3) thoracic, in which the sternum is completely split or absent, and the heart lies partially or completely outside the thorax; 4) thoraco-abdominal, which usually accompanies Cantrell’s syndrome; 5) abdominal, in which the heart passes through a defect in the diaphragm to enter the abdominal cavity. (Kim et al, 1997; Dobell et al, 1982).

Genesis of ectopia cordis has not been fully explained, although several theories have been offered (Dobell et al, 1982). Predominant theory states that there is primary failure of descent & mid line fusion of the lateral body folds (Humph et al, 1999); early rupture of chorion & I or yolk sac causing failure of midline fusion, amniotic band syndrome (Dobell et al, 1982; VanAllen & Myhre, 1985).

The majority of ectopia cordis patients have associated intracardiac defects. Ventricular septal defect, atrial septal defect, tetralogy of Fallot, and diverticulum of the ventricle are the most commonly encountered heart lesions (Amato et al, 1995; Leca et al, 1989). The severity and the complexity of the intracardiac defect contribute largely to the poor prognosis associated with this malformation (Amato et al, 1995).

It has been also observed that the ultimate survival of these patients depend more on the presence or absence of intrinsic cardiac defects rather than surgical techniques. Most of infants are stillborn or die within first few hrs or days of life (Humph et al, 1999; Shamberger, 1998).

The overall objectives of ectopia cordis management are: closure of the chest wall defect, including the sternal defect, repair of the associated omphalocele, placement of the heart into the thorax, and repair of the intracardiac defect (Dobell et al, 1982; Leca et al, 1989).

Ectopia cordis is a rare congenital malformation which may require a staged procedure to achieve a complete repair. The prognosis of this condition has been poor historically. With the advances in all aspects of medicine, the number of infants with ectopia cordis who undergo successful surgical repair and survive is steadily increasing.

Bibliography: