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Ectopia cordis-beating heart outside the chest

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Abstract
Ectopia cordis is a rare heart lesion that results from an abnormal development of the primitive heart outside the embryonic disc in the early stage of development. It represents a form of pericardial defect but is further characterized by a partial or complete displacement of the heart outside the thorax. Congenital heart diseases like tetralogy of Fallot (most common), ventricular hypoplasia, transposition of great vessels (TGA), tricuspid atresia, Ebstein's anomaly, common atrium, may be present. Multiple extra-cardiac defects have also been occurs this like with meningocele, encephalocele, cleft lip, palate deformities and ventral wall defects. The prognosis is very poor in these cases.

Keywords: Ectopia cordis, anomaly, malformations

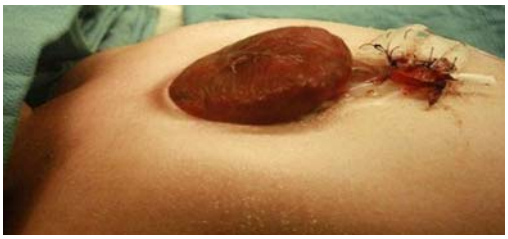
1. Introduction

Ectopia cordis is a rare congenital anomaly associated with the heart positioned outside the thoracic cavity either partially or completely. The ectopic heart can be found along a spectrum of anatomical locations includes the neck, chest or abdomen and the heart is protrudes through a split sternum to outside the chest. The designation ectopia cordis was first proposed by Abott in 1898, although cases of patient with similar defects had been described for decades with other designation.

The heart is located in the fifth intercostal space, resides in the space between the lungs and lies on the diaphragm with its apex towards the left side. The prevalence is 1:126000births and occur more in female infants.

Classification

- Cervical Ectopia cordis:-Exposed heart is located in the neck of infants and is about 5%of the total cases.
- Thoracic Ectopia cordis:-Exposed heart is located in the anterior of the sternum and is occur in about 65%of total cases.
- Thoracoabdominal Ectopia cordis: - Heart is located in the area between thorax and the abdomen and constitutes about 20%of total cases.
- Abdominal Ectopia Cordis:-The heart is located in the abdomen of the infant and in 10% of the total cases.



Etiopathogenesis and Clinical Manifestation

The exact etiology of ectopia cordis is not known. It is largely attributed to the improper development of chest cavity during the embryonic stage due to failure of ventral body wall formation and lateral body walls failed to initiate fusion. Mechanical theory proposed that ectopia cordis is due to the early rupture of chorion and /or yolk sac. The chorion and yolk sac causes compression of thorax and which stops the midline fusion. The defective ventral wall is not capable of providing the necessary shield protection to the heart. It can be also associated with chromosomal abnormalities, like Trisomy18 and Turners syndrome. Amniotic band syndrome is a mechanical cause of ectopia cordis. It occurs when the fetus becomes entangled in fibrous string like amniotic bands in the womb, restricting blood flow and affecting the baby's development that leads to malformation deformation or disruption.

Other organs may also have formed outside the skin. The heart is found outside the chest in the neck or abdomen. The ectopic heart is not protected by the skin or sternum. Other organs may also have formed outside the skin and many heart defects may occur. Most babies born with this condition will die during their first few days. Still births are common. Along with the evident malformation of the heart neonates may develop spinal defects, cleft palate and pulmonary atresia. Patients have C curvature on the thoracic spine as well as kyphosis.

Defects associated with ectopia cordis includes;

- Intra cardiac defects
- Atrial septal defect
- Ventricular septal defect
- Teratology of Fallot
- Tricuspid atresia
- Double outlet right ventricle
- Non – cardiac malformations
- Pentalogy of Cantrell
- Omphalocele

Diagnosis

- Prenatal Sonographic screening:-it can be diagnosed in uterus as early as the first trimester.

- Combination of sensitive urinary pregnancy tests, transvaginal ultra sound and serum HCG estimation in the first trimester.
- Radiographic analysis:- The frontal chest radio graph includes

- Abnormal cardiac position and configuration
- Sternal defects
- Widening of the superior mediastinum
- Air lucency that surrounds the ectopic heart
- Wide separation of sternal ends of the clavicle.

Treatment

There have been limited treatment option due to the uncommonness of the condition and speedy mortality of the infant shortly after delivery. Surgery is available for an infant that is thriving at birth but it is very complicated and risky. Covering of the naked heart and abdominal contents with silastic prosthesis (polymeric silicone substances have the properties of rubber but are biologically inert) is done instantaneously. If any intra cardiac defect is present it is corrected first prior to the replacement of abdominal contents. After a successful operation to provide coverage of the heart further aim is to repair the intra cardiac effect and to reconstruct the chest wall or an orthotopic heart transplantation. Systemic antibiotics and inotropic drugs can be administered to prevent infection and to support the heart and to improve the blood supply.

The overall surgical objectives can be concluded as

1. Closure of the chest wall defect (either by doing the primary chest wall closures or by using bone or cartilage or artificial prosthesis)
2. Closure of the sternal defect
3. Repair of the associated omphalocele
4. Placement of the heart in to the thorax
5. Repair of the intra cardiac defect.

Prognosis and Survival

The prognosis depends on three factors,

a. Location of the defect
b. Extent of intra cardiac defects
c. Associated abnormalities

In most case the prognosis is poor, most infants are still born and others will die shortly after delivery due to hypoxemia, cardiac failure, inflammations and infection. But some studies suggested a better prognosis with surgery of thoracoabdominal ectopia cordis or less severe pentalogy of Cantrell.

Outcomes and achievements.

- The first attempted repair of EC was done by Cutler and Wilens in1925
- Koop in 1975 achieved the first successful repair of thoracic EC in two stages.
- Dobell *et al* reported two stage correction
- Khaled *et al* reported single stage repair in 2003(France)
- Goncalves *et al* reported successful repair of uncomplicated EC in June 2007(Brazil).

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