ECTOPIA CORDIS UP DATE - 2008

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In medicine an ectopia is a displacement or malposition of an organ of the body. Most ectopias are congenital but some may happen later in life.

Ectopia cordis is a birth defect in which the heart is complete or partially outside of the thoracic cavity. In the most common form, the heart protrudes outside the chest through a split sternum. Less often the heart may be situated in the abdominal cavity or neck. Figure 1

Figure1: partial thoracic ectopia cordis

Often other birth defects are also present. This condition is usually fatal in the first days of life. In some cases surgical treatment is possible. The ectopic heart is not protected by the skin or sternum. Other organs may also have formed outside the skin. Often the heart is not formed properly and many other heart defects are associated with this condition including: Tetralogy of Fallot, pulmonary atresia, atrial and ventricular septal defects, double outlet right ventricle(1). Other non cardiac malformations may be present such as cleft palates, spine malformations that can cause kyphosis.

TYPES

Depending on where the defect was located, the cases of ectopia were classified into four groups: cervical, thoracic, thoraco-abdominal, and abdominal.

1)CERVICAL(2)

2)THORACIC: where the heart would lie within the thoracic cavity(3).

3)THORACOABDOMINAL: where the heart would lie somewhere between the thoracic and abdominal cavities (4).

4) ABDOMINAL: where the heart would lie in the abdominal cavity(5).

PREVALENCE:

Ectopia cordis is a rare disease that occurs in 5.5 to 7.9 per million live births. It is a very rare congenital heart malformation.

To know the prevalence of fetal cardiopathy and detection in high risk pregnancies a observational study was made in pregnancies women with 16 old week of gestation.A total of 3500 high-risk pregnancies and were detected 112 cases with fetal cardiopathy (3.2%). The 30% of them had a risk factor of cardiopathy. The most frequent fetal cardiac defect detected were:

1)Arrhythmia in 34 fetus;

2)Septal defects in 30

3)Valvular defects in 17

4)Hypoplasic or absence of cardiac cavities 16

5)Tronco-conus defects 8,

6)Ectopia cordis 3,

7)Cardiac tumor 2,

8)Abnormal drainage of pulmonary veins

The diagnosis increased every year since started study. The prenatal diagnoses suspected in fetal echocardiography were confirmed in 80% of the cases in neonatal period(6).

Etiology: No exact cause has been identified but this condition has also been seen more frequently in Turner Syndrome and Trisomy 18(7) however so far there is no evidence that it is a genetically transmissible disease. The entity is frequenty associated with Cantrel pentalogy. It is a rare congenital anomaly consisting of the following five features:

1) Midline supraumbilical abdominal wall defects

2) Deficiency of the anterior diaphragm

3) Defects in the diaphragmatic pericardium

4) Defects of the lower sternum

5) Congenital cardiac malformations.

PROGNOSIS

Most cases result in stillbirth or death shortly after birth. Some cases of ectopia cordis can be treated surgically but in general involve lengthy and very complicated pediatric cardiothoracic surgery.

The malfunction happens when the sternum forms during the gestation period and the heart begins to grow in the wrong place.

TREATMENT

The handling of the ectopia cordis includes an appropriate prenatal diagnosis through echocardiograph, a multidisciplinary perinatal team, to program the caesarean operation, aseptic handling of the newborn, immediate correction of the wall defect with skin torn piece without trying to correct the costal grill, surgical treatment using a rib graft to create a neo-sternum, specific hemodynamic cares, to reprogramme a correction of associated defects(8).

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