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Case Report

Partial Ectopia Cordis: A Case Report

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ABSTRACT

***Background*:** One-third of all major congenital anomalies are Congenital heart disease (CHD) and Reported CHD prevalence increased over time and in Asian countries is more than western countries.
Ectopia cordis (EC) is a rare congenital anomaly with an estimated incidence of 1:100 000 live births in developed countries. EC is characterized by abnormal heart placement outside the thorax, mostly on the thoracoabdominal side. This form is often associated with pentalogy of Cantrell.
Case report: We report one cases of the ectopia cordis at the Emam Reza Hospital in Mashhad. In this report, a rare case with incomplete pentalogy of Cantrell are described. It was a boy with a large omphalocele with evisceration of the heart. He had normal capillary refill and responded to stimuli. This patient was a male fetus with ectopia cordis with intracardiac anomalies; a large omphalocele with evisceration of the heart; a hypoplastic sternum and rib cage.
Conclusion: Prognosis seems to be poorer in patients with the complete form of pentalogy of Cantrell, EC, and patients with associated anomalies. Intracardial defects do not seem to be a prognostic factor.

Keywords: Congenital heart disease, Ectopia cordis, Neonate

Introduction

One-third of all major congenital anomalies are Congenital heart disease (CHD) and Reported CHD birth prevalence increased over time, from 0.6 per 1,000 live births (95% confidence interval [CI]: in 1934 to 9.1 per 1,000 live births (95% CI: 9.0 to 9.2) after 1995.

In one systematic review included a total study population of 24,091,867 live births with CHD identified in 164,396 infants.

Asia reported 9.3 per 1,000 live births (95% CI: 8.9 to 9.7) then Reported CHD prevalence in Europe was significantly higher than in North America (8.2 per 1,000 live births [95% CI: 8.1 to 8.3] vs. 6.9 per 1,000 live births [95% CI: 6.7 to 7.1]; P < 0.001).

These differences that Observed in the studies may be of genetic, environmental, socioeconomical, or ethnic origin (1).

In some studies, ventricular septal defects (VSDs) is the most common type of CHD.

The incidence of moderate and severe forms of CHD is about 6/1,000 live births (19/1,000 live births if the potentially serious bicuspid aortic valve is included), and of all forms increases to 75/1,000 live births if tiny muscular VSDs present at birth and other trivial lesions are included (2).

Extrathoracic heart (ectopia cordis) is a rare CHD in which the heart is located outside the hest cavity.

The cases are usually categorized according to whether the heart is anterior to the sternum (thoracic type), within the abdomen (abdominal type), between the thorax and the abdomen (thoracoabdominal type), or in the neck (cervical type) (3).

In this article we report one case of the ectopia cordis at Imam Reza Hospital in Mashhad, Iran.

Case report

Upon birth, the infant was breathing sponta-neously with Apgar scores of 8 at 1 minute and 9 at 5minutes. His vital signs were as follows: blood pressure 39/20, temperature 36.6°C axillary, and

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Figure 1. A neonate with ectopic cordis

SpO2 86%.

Continuous positive airway pressure was applied with hood, but he continued to have labored respirations and was intubated. The heart was noted to be partialy extruded out of the chest, and major great vessels was not seen (Figure1).

There was a superficial abrasion on the anterior aspect of the heart with no evident bleeding or oozing, complete absence of the pericardium and there was no skin cover. The cardiac apex pointed towards the chin, the heart being rotated through 90 degrees in the frontal plane so that the diaphragmatic surface was anterior and the anterior surface faced the thorax.

There were no other evident malformations. The femoral pulses were easily palpated and equal. He had normal capillary refill and responded to stimuli.

A sterile, moist coverage was placed, heart. He was started on prostaglandins was administered. Initial diagnostic studies showed pH of 7.08, pCO2 of 61, HCO3 of 24, normal hemoglobin/ hematocrit, normal white cell count with 10% bands, and normal electrolytes and renal function. Chest radiographs revealed hypoplastic, hypoventilated lungs (Figure 2).

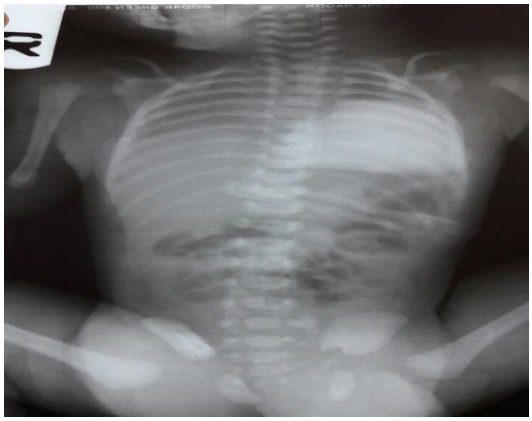
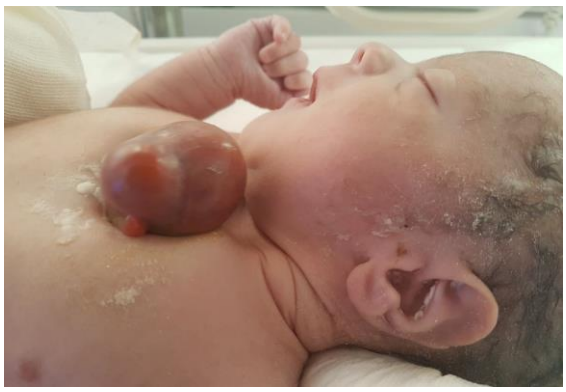


Figure 2. Chest radiography ectopia cordis



Bedside echocardiography was technically difficult, requiring the operator to "hold the heart." Although there were normal atrioventricular and ventriculoarterial connections, mild to moderate tricuspid regurgitation, Large Inlet to PM VSD, severe PS with very small Pulmonary artery Valve & Rt & Lt Pulmonary arteries (TOF Anatomy) were present.

Small PDA, Mod size ASD2 and good cardiac function was seen in bedside TTE.

Discussion

The pentalogy of Cantrell is a rare syndrome with an estimated incidence of 5.5 per 1 million live births (4). It is described as a deficiency of the anterior diaphragm, a midline supraumbilical abdominal wall defect, a defect in the diaphragmatic pericardium, various congenital intracardiac abnormalities, and a defect of the lower sternum. The pathogenesis of pentalogy of Cantrell has not been fully elucidated. Cantrell et al. (5) suggested an embryologic developmental failure of a segment of the lateral mesoderm around gestational age 14–18 days.

Organs may eviscerate through the resulting sternal and abdominal wall defects. EC itself is characterized by complete or partial displacement of the heart outside the body. Cervical, cervicothoracic, thoracic, and thoracoabdominal types of EC have been described (6).

Various other associated anomalies have been reported and include craniofacial and central nervous system anomalies such as cleft lip and/or palate, encephalocele, hydrocephalus, and craniorachischisis (7-8); limb defects such as club foot, absence of tibia or radius, and hypodactyly (9, 10); and abdominal organ defects such as gallbladder agenesis and polysplenia (11).

Often the spectrum of the original pentalogy of Cantrell is not complete. Toyama (12) suggested the following classification of the pentalogy of

Cantrell: class 1, definite diagnosis, with all five defects present; class 2, probable diagnosis, with four defects present, including intracardiac and ventral wall abnormalities; and class 3, incomplete expression, with various combinations of defects present, including a sternal abnormality.

Many different types of congenital heart disease have been reported with thoracoabdominal ectopia cordis including:

Tetralogy of Failor, double-outlet RV, VSD, ASD, tricuspid atresia, Ebstein anomaly, common atrium, common AV septal defect, anomalous pulmonary venous connections, single-ventricle ,pulmonary stenosis/ atresia, aortic stenosis, coarctation of the aorta, TGA, and diverticulum of the LV or both ventricles.

In our patient, the heart protruded through an incomplete sternal defect so that it lay outside the mediastinum, and There were intracardiac anomalies (Extrim TOF Anatomy).

After birth, echocardiography is essential for diagnosis of associated cardiac anomalies. Other features of the pentalogy of Cantrell and known associated anomalies can be diagnosed by conventional radiography or sonography.

The treatment of the pentalogy of Cantrell consists of corrective or palliative cardiovascular surgery, correction of ventral hernia and diaphragmatic defects and correction of associated anomalies. However, most surgical reports have demonstrated poor results for repair of this defect.

Nevertheless, due to the complex nature of these malformations and the risk of infection, most have advocated immediate surgery to correct their congenital heart disease and the anterior chest wall defect.

The best treatment strategy depends on the size of the abdominal wall defect, the associated heart anomalies, and the type of EC.

Conclusion

The prognosis of EC seems to be poorer in patients with the complete form of pentalogy of Cantrell, EC, and patients with associated anomalies.

Intracardial defects do not seem to be a prognostic factor. When the diagnosis pentalogy of Cantrell is suspected, a multidi0sciplinary approach is essential.

A prenatal diagnosis and a medical team working consisting of a gynecologist, a neonatologist, a pediatric cardiologist, a geneticist, and a pediatric surgeon should beuse their expertise in choosing the best approach to this severe disorder.

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Conflicts of interests

The authors declare that there are no conflicts of interest.

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