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Ectopia cordis coexisting with hypoplastic left heart structures

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Abstract: Herein we present the fatal case of premature baby with coincidence of extreme form of ectopia cordis and hypoplastic left heart structures.

Key words: ectopia cordis, hypoplastic left heart syndrome.

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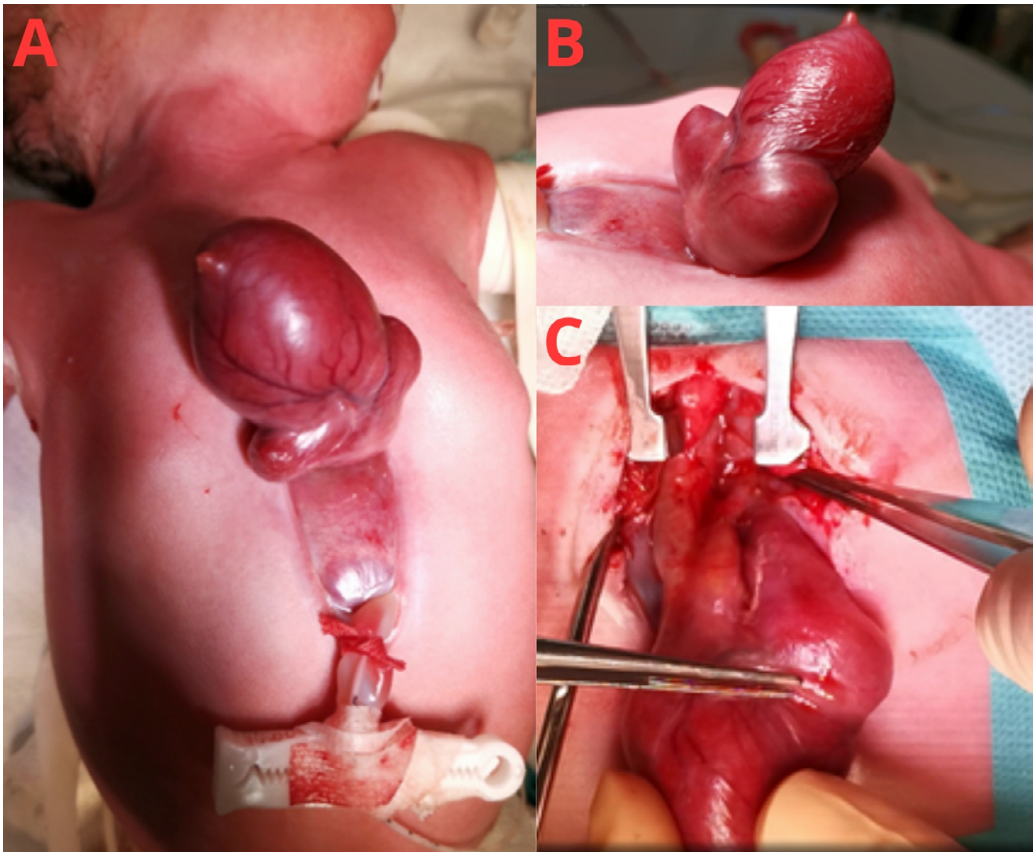
Ectopia cordis is extremely rare congenital anomaly, with a prevalence estimated between 5.5 and 7.9 cases per million live births [1, 2]. It is characterized by the partial or complete extra-thoracic location of the heart. The prognosis is often fatal, particularly when associated with other cardiac abnormalities [3, 4]. A variant with the heart's apex oriented cephalad (upward) is susceptible to any manipulations, thus the postnatal diagnostic process of the heart defect may be complicated by the extreme hemodynamic instability [5]. The congenital heart defects coincidence with ectopia cordis may occur in 7–35%. The conotruncal malformations are predominant. The hypoplasia of left ventricle is reported extremely rare [6, 7]. This report details a case of ectopia cordis coinciding with hypoplastic left ventricle in a premature baby-girl.

The neonate, delivered via cesarean section due to impending intrauterine death at 34 weeks of gestation with a body weight of 2.1 kg. The ectopia cordis has been diagnosed at 20 weeks of gestation, with suggestion of heart defect, without clarification of the precise nature of it. After delivery, physical examination revealed a thoracic ectopia cordis, with the heart protruding from the lower part of sternum and apex directed cephalad as well as lack of the skin in the area of

abdomen between the ectopic heart and the umbilicus. The rapid attempts of performing precise, postnatal, epicardial echocardiography was complicated by severe hemodynamic instability. Thus, the decision was made about immediate (30 min. after delivery), surgical isolation of the protruding heart from the environment to reduce the risk of mediastinitis. The heart was covered with double-layer xenograft reaching the level of umbilicus, reinforced from inside with artificial construction preventing any compression to heart. Additionally, partial sternotomy was done to create landing zone in the mediastinum for part of the heart. However, the hemodynamic instability with rapidly progressive hypoxemia was observed. The intense care included fluid administration, refine mechanical ventilation techniques with nitric oxide administration, PGE_1 infusion and inotropic support did not prevent gradual deterioration of patient status. The extreme form of hypoxemia has led to hypotension, severe metabolic acidosis, anuria, subsequently the shock and death 30 hours after delivery. The autopsy revealed well developed right ventricle, hypoplastic left ventricle, intact interatrial septum, ventricular septal defect, hypoplastic pulmonary branches measuring 1.5 and 2 mm in diameter, and partial anomalous pulmonary venous return with the left pulmonary veins connecting to an anomalous left superior vena cava. The ductus arteriosus was not identified.



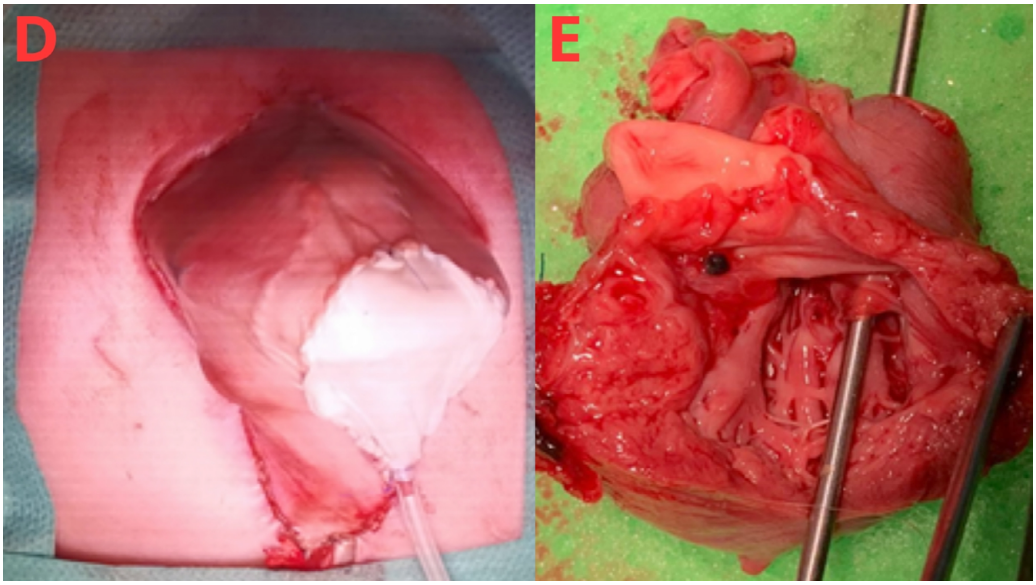


Fig. 1. Detailed Photographic Series on Ectopia Cordis in a Preterm Neonate.

A. Superior Perspective of Ectopia Cordis: This image presents an overhead view of ectopia cordis, where the heart is abnormally located at the lower part of the sternum. The heart's apex points towards the head, and a notable absence of skin between the heart and the umbilicus is evident. The subject is a 1-hour-old, preterm neonate with a body weight of 2.1 kg.
B. Lateral View of Ectopia Cordis: Captured from the left lateral side, this image clearly shows the ectopia cordis, with both atria and the ventricle distinctly visible. This perspective emphasizes the extent of cardiac displacement and structural details.
C. Ectopia Cordis with Visible Major Vessels: Following the repositioning of the heart's apex towards the abdomen and a partial lower sternotomy, this image highlights the ectopia cordis with two main vessels, the aorta and pulmonary trunk, being visible.
D. Isolated Ectopia Cordis with Xenograft Patches: The ectopic heart is isolated from the external environment using xenograft patches. The covering is extended to the a fragment of the abdomen until the umbilicus and is reinforced with an internal construction to prevent any compression of the heart.
E. Autopsy Findings: The autopsy image reveals a well-developed right ventricle with a small ventricular septal defect, indicated by a probe going to a hypoplastic left ventricle.

In conclusion, the precise prenatal diagnosis of ectopia cordis and coexisting heart defect is crucial in planning the immediate postoperative management and counseling the patients. The postnatal diagnostic and management tools are limited in patients with extreme form of ectopia cordis. Epicardial echocardiography may provoke life threating hemodynamic instability. Other diagnostic tools (CT, NMR or angiography) are less effective, especially in premature babies with low or very low body weight, which is observed in majority of neonates with ectopia cordis. Managing of the patients with form of single ventricle type is extremely demanding in premature neonates, especially when pulmonary vascular resistance is still elevated and coexisting with hypoplasia of pulmonary arteries. Coincidence of extreme form of ectopia cordis and congenital heart defect (especially single ventricle form) has the poor prognosis.

Conflict of interest

None declared.

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