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Case Report

Ectopia Cordis with Double Outlet Right Ventricle: A Rare Case of a Syrian Neonate with Cantrell Pentalogy

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Abstract

Ectopia cordis is a very rare congenital defect, in which the heart is displaced outside the thoracic cavity. It may be associated with other midline defects of the sternum, abdominal wall and diaphragm as in Pentalogy of Cantrell Syndrome, which has a poor prognosis. Herein, we report a case of a 2-hour-old female Syrian newborn with Pentalogy of Cantrell Syndrome and an unusual association of complex heart defect consisting of a double outlet right ventricle (DORV) - TGA type (sub pulmonary VSD) with PDA and mild pulmonary stenosis. Our patient has undergone only the first stage of surgical intervention, and died after surgery due to a cardiogenic shock. As far as we know, this case is the first reported case in Syria.

Keywords: *Ectopia Cordis; Pentalogy of Cantrell; Double Outlet Right Ventricle; Surgical Intervention*

Abbreviations

POC: Pentalogy of Cantrell; EC: Ectopia Cordis; TGA: Transposition of the Great Arteries; VSD: Ventricular Septal Defect; PDA: Patent Ductus Arteriosus; DORV: Double Outlet Right Ventricle

Introduction

Ectopia cordis (EC) is a very rare congenital defect, in which the heart is displaced outside the thoracic cavity. This defect may be associated with other defects on the midline within the so-called "Cantrell syndrome", which is considered a rare condition. Herein, we report a case of a Syrian newborn with Pentalogy of Cantrell Syndrome (thoraco-abdominal syndrome). As far as we know, our case is the first reported case in Syria.

Case Report

A 2-hour-old full-term female neonate was admitted to Children's Damascus University Hospital with a complaint of a heart protrusion outside the chest wall. She was born by a Caesarean delivery to a non-consanguineous parent. There was no maternal history of infections, radiation, smoking, non-prescribed medication, alcohol abuse or taking drugs during pregnancy. There was no family history of any congenial heart diseases. A screening ultrasound was performed at 17-week gestational age, and it showed the fetus heart positioned completely outside its chest wall. However, the mother decided to continue the pregnancy.

On physical examination, there was a good air entry to the lungs, respiratory rate was 60 per minute, heart rate was 140 per minute, and oxygen saturation was 80% on room air. The sternum was absent. The whole heart was pumping outside the thoracic cavity. There was also an upper anterior abdominal wall defect with omphalocele.

Echocardiography revealed a double outlet right ventricle (DORV) - TGA type (sub pulmonary VSD) with PDA and mild pulmonary stenosis.

Initial management included a supportive care (intravenous antibiotics and fluids with supplemental oxygen), then the patient was admitted urgently after 2 hours to perform a surgical intervention. During the operation, the surgeon stated that there was a clear diaphragmatic hernia. There was no possibility to reposition the heart back into the chest cavity and close the chest wall. A palliative intervention was performed by covering the heart with biologic patch. Our patient died directly after surgery due to a cardiogenic shock, despite of adequate resuscitation.

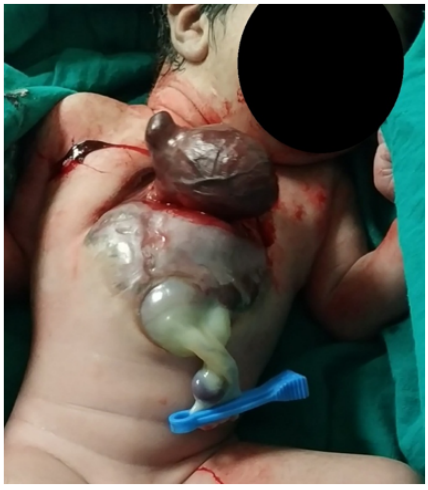

Figure 1

Discussion

Ectopia cordis is one of the rare congenital anomalies, with an estimated prevalence of 5.5 to 7.9 per million live births [1]. The term ectopia cordis was first described by Haller., *et al.* in 1706 [2]. It is characterized by a partial or total protrusion of the heart outside the chest cavity. EC is classified based on where the heart is displaced into subcategories: cervical (5%), cervicothoracic and thoracic (65%),

thoracoabdominal (20%) and abdominal (10%) [2]. Our patient had thoracic type which is the most common one. The cause of this condition is an abnormal development of the midline mesodermal components, and it can be associated with other midline defects such as omphalocele, congenital heart diseases, sternal defects and diaphragmatic hernia [3]. EC is a characteristic abnormality in Pentalogy of Cantrell (POC) which is a rare syndrome consists of upper abdominal wall defect, lower sternal defect, anterior diaphragm defect, epicardium defect, and intracardiac defects [4]. In our report, all of these findings were actually present in the patient. The most common cardiac anomaly associated with EC is ventricular septal defect, then atrial septal defect and tetralogy of Fallot [5]. Our patient had a complex heart defect consisting of a double outlet right ventricle (DORV) - TGA type (sub pulmonary VSD) with PDA and mild pulmonary stenosis. This intracardiac complex anomaly is an unusual association of POC. The severity of the intracardiac anomalies is linked with the prognosis of POC syndrome, as well as other associated malformations do. Gruberg., *et al.* had mentioned in a report that mortality can reach 50% in patients with complex cardiac malformations who undergo surgery during the first days of life [6]. In general, the prognosis of EC is very poor. The optimal management in EC is surgical repair, and it can be done in a single stage or a multistage approach which is more preferable. Multistage approach usually includes -in the first step- providing a soft tissue covering for the heart, then -in the next steps- reduction of the heart into the thoracic cavity, repair of intracardiac anomalies, and thoracic reconstruction[7]. EC is incompatible with life without surgical repair and, in addition, surgical repair usually carries a high risk and a low chance of survival. Prognosis is related to the type of EC, the coexisting congenital heart defects and the associated extracardiac malformations [8]. In our case, the neonate died immediately after the first-stage surgery. Depending on the fact that EC is a poor prognosis condition, early diagnosis during pregnancy and termination of the pregnancy may be advisable. Still, this matter is linked to local culture and medical laws.

Conclusion

Ectopia cordis is an incompatible condition with life. The chance of survival is highly dependent on early detection, associated anomalies, surgical intervention as early as possible after birth and availability of ultimate post-operative care. However, the prognosis of EC remains poor.

Acknowledgements

None declared.

Conflict of Interest

The authors declare that they have no competing interests.

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Ethical Approval

No approval was required.

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