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Case Report

Pentalogy of Cantrell With Total Ectopia Cordis and A Major Omphalocele. A Case Report

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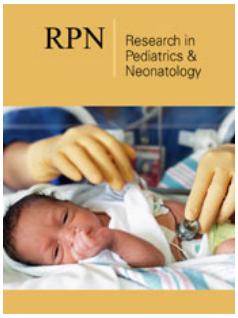

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Abstract

Ectopia cordis (EC) is a rare malformation due to failure of maturation of the midline mesodermal components of the chest and abdomen. It can be defined as a 0.1% of congenital heart diseases, it could present isolated or belonging to the spectrum of the Pentalogy of Cantrell (PoC), this is a rare congenital disorder first described in 1958 by Cantrell. We are reporting a rare case of total Ectopia cordis, associated to a major omphalocele, total agenesis of the sternum, anterior diaphragmatic deficiency, absence of pericardium, and Persistence of the Ductus arteriosus, making therefore these features compatible with a full spectrum of the Pentalogy of Cantrell, encouraging us to report this case.

Keywords: Ectopia cordis; Major Omphalocele; Sternum agenesis; Anterior diaphragmatic deficiency; Absence of pericardium; Persistence of the Ductus arteriosus

Introduction

Ectopia cordis (EC) is a rare malformation due to failure of maturation of the midline mesodermal components of the chest and abdomen. It can be defined as a 0.1% of congenital heart diseases, it could present isolated or belonging to the spectrum of the Pentalogy of Cantrell (PoC). Pentalogy of Cantrell (PoC) is a rare congenital disorder first described in 1958 by Cantrell. The full spectrum of this syndrome consists of midline supraumbilical abdominal wall defect, defect of the lower sternum, deficiency of the anterior diaphragm, defect in diaphragmatic pericardium and congenital heart disease [1]. The incidence of ectopia cordis (Ec) is sporadic with only 5.5 to 7.9 per 1 million live births [2]. PoC has its origin in embryologic development, resulting from defective formation and differentiation of the ventral mesoderm at about 14 to 18 days of embryonic life [1]. The aetiology is probably sporadic and multicausal transmission, the exact pathogenesis remains unknown. Outcome of PoC with ectopia cordis and congenital heart defect is disastrous [3].

Case Report

We present a neonate 3 hours after birth, through C section, Apgar 7/10, 6/1, weigh 3,2 kg. Crying at delivery but central cyanoses, respiratory distress, intubated and ventilated. No sternum, heart outside the chest, no pericardium covering. Intact Omphalocele about 10 x 10 cm. Rest of physical exam unremarkable, was done diagnoses of a PoC and complete thoracic ectopia cordis with cephalic orientated cardiac apex, deficiency of the anterior diaphragm, agenesis of the sternum, persistence of the arteriosus duct, left superior cava vein and a major omphalocele, Echocardiogram PDA. INR 2,67 other blood test normal (Figure 1). After stabilisation was taken to theatre. In theatre was produce a chest cavity to receive the heart, removing three left costal cartilage and dissected the left superior vena cava reflecting it more to the left. The heart was located with the apex up; after that was coverage the ectopia cordis by mesh and cutaneous pedicled flap ; the Omphalocele , was covered by silver sulfadiazine and located a mesh for a conservative treatment due to the big size of it made impossible the surgical correction of the omphalocele (Figure 2). After operation patient keeping on mechanical ventilation till 12 hours later when the child suffered a cardiac arrest and didn't respond to the resuscitation manoeuvres.

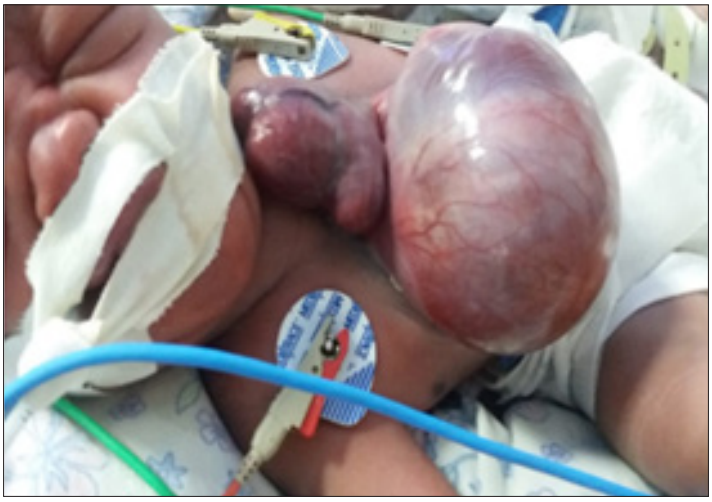

Figure 1

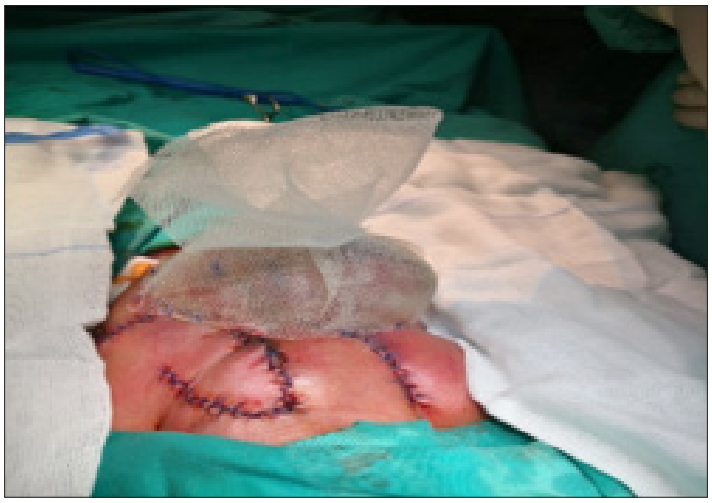

Figure 2

Discussion

Pentalogy of Cantrell including complete thoracic ectopia cordis is a rare congenital disorder of unknown pathogenesis with sporadic incidence [2,4]. Especially, complete thoracic ectopia cordis in addition to complex heart disease, are known to have poor prognosis [4,5]. Surgical internalization of the heart into the thoracic cavity is associated with high risk of cardiovascular compromise and ischemia [5] The paucity of survivors with this entity demonstrates the need for extreme caution in preserving hemodynamic stability when the heart is covered, whether the repair is performed as a primary or a staged repair.

References

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