

Thoracic ectopia cordis with anatomically normal heart

Ectopia cordis torácica com coração anatomicamente normal

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Abstract

Ectopia cordis is a rare congenital malformation, which is commonly associated with other intracardiac defects. A two-day-old full-term baby girl was admitted to Santa Casa de Misericórdia Hospital Montes Claros, NG, Brazil, with thoracic ectopia cordis. A transthoracic echocardiographic study did not identify any associated congenital heart diseases. The infant underwent surgical treatment using a rib graft to create a neo-sternum. She was discharged after presenting a good outcome on the 20th postoperative day.

Descriptors: Heart defects, congenital. Ectopia cordis. Thoracic wall, abnormalities. Infant, newborn.

Resumo

A ectopia cardíaca é uma má formação congênita rara, normalmente associada a outras más formações intracardíacas. Uma criança do sexo feminino com dois dias de vida, nascida a termo de uma primeira gestação sem intercorrências (G1P1A0), por parto cesariano, foi admitida na Santa Casa de Montes Claros, em Minas Gerais, apresentando ectopia cardíaca na forma torácica. O estudo ecocardiográfico transtorácico não evidenciou cardiopatia congênita associada. A paciente foi submetida a tratamento cirúrgico, utilizando enxerto de costela. Apresentou boa evolução, recebendo alta hospitalar no vigésimo dia de pós-operatório.

Descritores: Cardiopatias congênitas. Ectopia cardíaca. Parede torácica, anormalidades. Recém-nascido.

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INTRODUCTION

Ectopia cordis is an extremely rare congenital heart malformation occurring in 5.5 to 7.9 per 1 million native populations [1]. It is characterized by the anomalous position of the heart, outside of the thoracic cavity. The thoracic presentation is the most common form (59%). There are only a few cases published in the literature related to the survivors of this disease [2].

We report one case of ectopia cordis, with thoracic presentation, without associated congenital heart defect, with a favorable disease outcome after surgical procedure. This case report is justifiable due to its rare form of occurrence because it is related to the absence of associated congenital heart defect.

CASE REPORT

A two-day old full-term baby girl from a primipara mother without clinical events (G1P1A0), born from Cesarean section, was admitted at Santa Casa de Misericórdia Hospital, Montes Claros, MG, Brazil. On clinical examination it was observed the exposition of the heart on the anterior surface of thorax covered only by the pericardium, which have already presented signs of necrosis (Figure 1). A transthoracic echocardiography did not identify any associated intracardiac congenital cardiopathy.

The patient underwent a surgical procedure with midline incision and resection of the necrotic tissue. A pleurotomy was performed and the thoracic cavity was transformed into a single cavity in order to accommodate the heart. A resection of a rib segment was performed to create a neo-sternum. A bilateral drainage of the thorax and a layer synthesis were performed up to the skin (Figure 2).

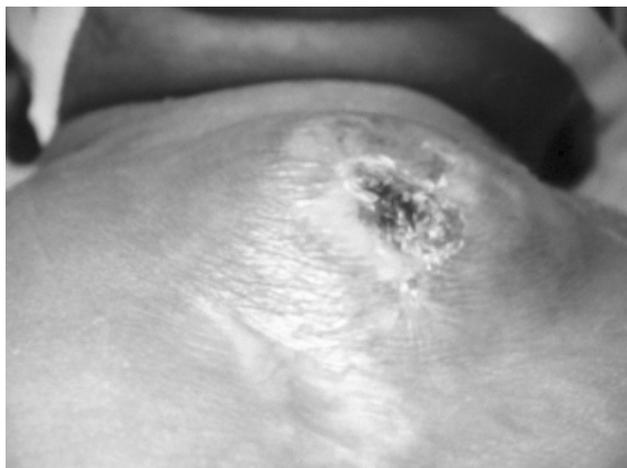


Fig. 1 - Heart in the anterior face of the thorax, covered only by pericardium with signs of necrosis



Fig. 2 - Patient after surgical intervention using median incision and resection of necrotic tissue

In the postoperative, the patient required mechanical ventilation for 72 hours and she did not present any other morbidity.

DISCUSSION

Ectopia cordis is a rare congenital heart defect (5.5 to 7.9 per 1 million newborn children). Usually, it is associated to other intracardiac congenital heart defects, and the troncoconal malformations are the most common ones [1, 3]. It is characterized by the anomalous position of the heart, with the thoracic form having a better prognosis making possible a long-term survival [1, 2].

The prenatal diagnosis has greatly improved and can be performed at about the 10th week [2].

Surgery is the only therapeutic option what associated with the improvement of myocardial protection and per- and postoperative supports have afforded a better survival rate [4]. Several surgical techniques have been described, thus the disease can be approached by one or two surgical stages. The first surgical stage is performed in patients requiring immediate treatment and aims to collect skin and soft tissue to recover the heart. Nevertheless, it is not always possible, and due to the low cardiac output the use of prosthesis is required. The purpose of the second surgical stage is to correct the associated congenital heart defects and to reconstruct the sternum [1].

The advantage of the autologous graft (autograft) over the prosthesis is due to the fact that the graft can grow with the patient and can develop more resistance to infections [1, 3].

In this case report, because the associated congenital heart defect was not present, as reported by Serrano et al. [5], the reconstruction in the neonatal period was chosen due to the greater flexibility of the thorax [3]. It was used a resected rib segment, after the thoracic cavity had been

opened, forming bars in the course of the sternum fixed with stitches (sutures) of ethibond 2.0. In this way, it was avoided the application of synthetic material as well as a procedure with a further greater rotation of muscular flaps.

Concluding, the early surgical approach has provided the repositioning of the heart into the thoracic cavity due to the greater elasticity of the thoracic wall. The use of a rib segment graft has been proved to possible because it is easily accessible and manageable causing the neo-sternum to be stabilized.

REFERENCES

1. Morales JM, Patel SG, Duff JA, Villareal RL, Simpson JW. Ectopia cordis and other midline defects. *Ann Thorac Surg.* 2000;70(1):111-4.
2. Escudero Ruiz de Lacanal L, Maese Heredia R, Cuenca Peiro V, Conejo Munoz, Medina Soto A, Garcial del Rio M. Thoracic ectopia cordis. *An Pediatr (Barc).* 2004;60(2):190-2.
3. Campos JR, Filomeno LT, Fernández A, Ruiz RL, Minamoto H, Werebe EC, et al. Repair of congenital sternal cleft in infants and adolescents. *Ann Thorac Surg.* 1998;66(4):1151-4.
4. Ben Khalfallah A, Annabi N, Ousji M, Hadrich M, Najai A. Thoracic ectopia cordis with tetralogy of Fallot. *Tunis Med.* 2003;81(suppl 8):661-5.
5. Serrano Gomez SG, Bermluez Sosa MT, Palma Hernandez E, del Castillo Salceda LF, Pinzon Muslera O, Hernandez Cortez B, et al. Ectopia cordis multidisciplinary approach with successful result. *Ginecol Obstet Mex.* 2006;74(8):439-43.