Case report - Congenital

Ectopia cordis, a successful single stage thoracoabdominal repair

Khaled Samir*, Olivier Ghez, Dominique Metras, Bernard Kreitmann

Department of Cardio-Thoracic Surgery (See Pr Metras), La Timone, Children’s Hospital, La Timone University Center, 13005 Marseille, France

Received 25 July 2002; received in revised form 6 April 2003; accepted 8 April 2003

Abstract

This is a report of a case of the rare ectopia cordis malformation of the thoracoabdominal type without intracardiac anomalies. The patient had a successful single stage repair with reduction of the herniating heart and reconstruction of a cartilaginous cover to protect the heart. The result was very good and the follow up for 13 months was very satisfactory.

Keywords: Ectopia cordis; Repair; Single stage

1. Introduction

Ectopia cordis (EC) is a rare malformation, occurring in 5.5–7.9 per million live births due to failure of maturation of the midline mesodermal components of the chest and abdomen [1].

It can be defined as a complete or partial displacement of the heart outside the thoracic cavity. It could be total or partial depending on the amount of the cardiac volume presenting outside the thoracic cavity. It could be classified into five types: cervical, cervicothoracic, thoracic, thoracoabdominal, and abdominal [2].

Thoracoabdominal EC (the heart exists in a direct communication between the thoracic and the abdominal cavities) frequently is associated with Cantrell’s pentad [3] (bifid sternum, deficiency of the diaphragm, diaphragmatic pericardium, defects of the anterior abdominal wall, and intracardiac defects) [4]. It can be considered as a very rare anomaly that is usually associated with other both intracardiac and extracardiac anomalies and a low survival rate despite the various techniques reported. With the development of the antenatal diagnosis and the tendency of parents to choose therapeutic abortion as a solution; this malformation is becoming more rare in the clinical practice. Surgical correction of these defects is complex and generally requires a staged closure including: (1) coverage of the ‘naked heart’; (2) placement of the heart into the thoracic cavity; and (3) sternal or thoracic reconstruction and survival past the perioperative period is rare [5].

2. Patient and methods

A female infant born during the 39th week of a non-identical twin pregnancy had an antenatal diagnosis of thoraco-abdominal EC as a part of Cantrell’s syndrome. The conduction of pregnancy was smooth and so was the delivery by a caesarian section for both a normal boy (2.7 kg) and the malformed girl (2.9 kg). The postnatal examination confirmed the diagnosis with an omphalocele. The anterior thoraco-abdominal defect extended from the lower border of a small sternum at the level of the 4th rib to below the umbilicus (15 cm long, 11 cm width), only covered by thin skin that allowed the cardiac impulse and the intestinal movements to be seen. In front of the hemodynamic stability, absence of intracardiac anomalies and normal alimentation the little girl was discharged from the hospital at the age of 11 days and a strict pediatric and cardiologic follow up was applied and apart from some dyspnea during nursing there was no problem. Echocardiographic examination confirmed the diagnosis with an omphalocele. The anterior thoraco-abdominal defect extended from the lower border of a small sternum at the level of the 4th rib to below the umbilicus (15 cm long, 11 cm width), only covered by thin skin that allowed the cardiac impulse and the intestinal movements to be seen. In front of the hemodynamic stability, absence of intracardiac anomalies and normal alimentation the little girl was discharged from the hospital at the age of 11 days and a strict pediatric and cardiologic follow up was applied and apart from some dyspnea during nursing there was no problem. Echocardiographic examination confirmed the diagnosis with an omphalocele. The anterior thoraco-abdominal defect extended from the lower border of a small sternum at the level of the 4th rib to below the umbilicus (15 cm long, 11 cm width), only covered by thin skin that allowed the cardiac impulse and the intestinal movements to be seen. In front of the hemodynamic stability, absence of intracardiac anomalies and normal alimentation the little girl was discharged from the hospital at the age of 11 days and a strict pediatric and cardiologic follow up was applied and apart from some dyspnea during nursing there was no problem. Echocardiographic examination confirmed the diagnosis with an omphalocele.
the omphalocele size. The echocardiography showed an almost closed VSD with a slightly dilated subcutaneous apex with a diverticular aspect. In supine decubitus; after sterilization of the thoraco abdominal field the towels were placed, the skin covering the heart was incised very carefully in the median plane with the incision extended over the deformed sternum. There was no pericardium covering the subcutaneous part of the heart that looked as if herniating through a large pericardial window. The heart was totally dissected after performing a median incision in the short sternum. The heart had a double apex due to the presence of a LV apical diverticulum (Fig. 2). It was impossible to reduce the heart to the pericardial space so we decided to open left pleural space and despite this, the first trial of reduction was accompanied with bradycardia and hypotension. To solve this problem we used a snaring mechanism by a non-absorbable 5/0 monofilament joining the LV apex with the left diaphragmatic copula that helped us doing a gradual well tolerated reduction until the apex with completely reduced to the pleural cavity. A pericardial replacement membrane of 0.1 mm was used to repair the pericardial defect and was fixed in a way so as to keep the heart in place. The lower three costal cartilages attached to the bifid sternum were dislocated from their costochondral junction and were deviated medially and fixed in place to protect the heart. After the complete repair of the EC a usual repair of the omphalocele was done.

3. Result

The postoperative period was uneventful and the patient was discharged after 9 days with a very satisfactory echocardiographic control. Follow up for 10 months showed a very good result with a very well tolerated repair.

4. Discussion

Some reports have described EC being diagnosed in the first trimester of pregnancy. The prenatal diagnosis of isolated EC is easy; counseling the patient, the perinatal management including term, place, and method of delivery, and optimal care of the newborn are more difficult. Ectopia cordis is a malformation that pediatricians rarely encounter, even at pediatric cardiology centers. Much more frequently it is a problem for sonographers and obstetricians; however, pediatric cardiologists should be aware of such cases, especially when additional abnormalities are present [6].

Thoracic EC and other midline defects are rare congenital anomalies that often occur with other intracardiac defects. Despite significant improvements in neonatal and infant cardiac surgeries, operations for thoracoabdominal ectopia cordis carry an extremely high mortality with only a few reported survivors of thoracic ectopia cordis [7].

Studies on chick embryos revealed that mechanical disturbance of the so-called membrana reuniens causes the development of EC. Hydrocortisone is known to produce a high incidence of EC. The incidence of EC after the hydrocortisone intraamniotic application on the 4th embryonic day reached 84.8%. Flow cytometry analysis of the cell cycle revealed that EC induced by hydrocortisone administration was associated with a significantly lowered proliferation activity of the prospective body-wall mesenchyme involved in the closure of the anterior wall of thorax [8].

Due to the rarity of EC there is no fixed or famous technique but only sporadic techniques mentioned in the literature. In our case simple careful reduction of the cardiac hernia to the left pleuron and reconstruction of the thoracic wall was used in a single stage.

5. Conclusion

Single stage repair of thoraco-abdominal ectopia cordis can be achieved with a good result.
References


Appendix A. ICVTS on-line discussion

Author: Dr. Sameh Ibrahim Sersar, Assistant Lecturer of Cardiothoracic Surgery, Mansoura University, Department of Cardiothoracic Surgery, Mansoura University, Mansoura, Egypt.

Date: 14-Oct-2003

Message: Can I ask about the protocol of timing used for repair of different congenital chest wall defects in your centre and what are the different methods to reduce the heart in difficult cases of reduction?