

THORACO-ABDOMINAL ECTOPIA CORDIS: CASE REPORT

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Ectopia cordis is a rare and impressive congenital abnormality, occurring in 5.5 to 7.9 per 1 million live births.¹ The defect is characterized by partial or complete displacement of the heart out of the thoracic cavity. This anomaly is classified into five types: cervical, cervicothoracic, thoracic, abdominal, and thoraco-abdominal.² The two most common forms of ectopia cordis are the thoracic and thoracoabdominal type.^{3,4} The latter is frequently associated with Cantrell's pentalogy, which include bifid sternum, deficiency of the diaphragm, defect of diaphragmatic pericardium, defect of the anterior abdominal wall, and intracardiac defects.^{3,5}

Previous reports suggest a poor prognosis for patients with ectopia cordis, particularly in the presence of thoracic ectopia cordis and significant heart defects.¹ More recently, with the advances in the medical field and surgical techniques, more patients born with this medical condition have been successfully treated and have survived. In general, the goal of the initial management is directed at providing coverage of the bare heart with skin or synthetic material without causing hemodynamic embarrassment. Later, subsequent operations to repair the intracardiac defects and to reconstruct the chest wall can be done.⁶ In this report, we present a case of thoracoabdominal ectopia cordis and an overview of its management in the medical literature.

Case Report

A 2250-g male neonate of normal delivery presented with an anterior thoracoabdominal defect with extrathoracic heart, a cleft sternum and omphalocele which were recognized at birth. The newborn was intubated and transferred to our medical center at day one of life. The midline defect extended from the lower margin of the neck

FIGURE 1A. Anterior view of the thoraco-abdominal ectopia cordis with omphalocele in day 1 of life before surgical intervention.

FIGURE 1B. Lateral view of the thoraco-abdominal ectopia cordis.

FIGURE 2. The patient at 2 months of age after first-stage surgical repair of ectopia cordis.

to the umbilicus. The sternum was completely bifid, with an inter-ridge distance of 6 cm, through which the heart was protruding for 4-5 cm and the apex pointing anteriorly. There was a major exomphalos as well, with a thin membrane covering the omphalocele.

Initial management included covering of the heart and omphalocele with sterile-soaked dressing and systemic antibiotics coverage (Figure 1). Direct echocardiography showed a moderate size atrial septal defect (ASD) with no other intracardiac abnormalities. Surgical repair of thoracoabdominal ectopia cordis was planned thereafter. Intraoperatively, the sternum was observed to be completely split, and present in the form of a 1 cm ridge with a gap of 5 cm in between. Mobilization of the skin and muscles were performed over the rib cage on both side. A "Z" incision advancement was done at all costochondral cartilages, and an attempt was made to approximate the sternal ridges. The thymus gland was small and was not removed. The gap was closed up to the 4th costal cartilage without any untoward effects. The heart could not be accommodated in the chest, and any further approximation of the lower half of the sternum led to severe compression of the heart resulting in hypotension with bradycardia. Since the heart could not be accommodated in the thoracic cavity, a Dacron graft (Impra Cardiovascular patch, Bard) designed to complete the deficient part of the sternum was attached to the split sternum, and sutured to the anterior part of the diaphragm. The abdominal cavity was stretched and the skin of the chest was mobilized and approximated to the midline to allow closure of the thoracoabdominal defect over the Dacron graft. A complete skin closure was achieved without hemodynamic embarrassment. The newborn was transferred subsequently to the cardiac intensive care unit.

The infant remained stable in the postoperative period. He required assisted ventilation for 21 days post surgery. Nutritional support was provided with total parenteral

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nutrition and nasogastric tube feeding. He was discharged after 2 months post surgery in good condition (Figure 2). A follow-up examination at three months of age showed good weight gain. The heart was covered by skin and protruded 1 to 2 cm above the thorax. An echocardiography showed a small ASD but otherwise normal heart.

Discussion

Ectopia cordis is a rare and striking congenital heart defect, which was first observed 5000 years ago.⁷ The defect is described as malposition of the heart, partially or completely outside the thorax.⁸ According to the position of the misplaced heart, ectopia cordis can be classified into five types: 1) cervical, in which the heart is located in the neck with sternum that is usually intact; 2) thoracic-cervical, in which the heart is partially in the cervical region but the upper portion of the sternum is split; 3) thoracic, in which the sternum is completely split or absent, and the heart lies partially or completely outside the thorax; 4) thoraco-abdominal, which usually accompanies Cantrell's syndrome; 5) abdominal, in which the heart passes through a defect in the diaphragm to enter the abdominal cavity.^{2,6}

Our case seems to belong to the thoracoabdominal type with a predominant thoracic component. The following characteristics were observed in our patient: completely bifid sternum, anterior extrathoracic heart with ASD, absence of parietal pericardium, and an omphalocele with supraumbilical abdominal wall defect. As such, our patient has all the classical components of Cantrell pentalogy, but the diaphragmatic defect.

The majority of ectopia cordis patients have associated intracardiac defects. Ventricular septal defect, atrial septal defect, tetralogy of Fallot, and diverticulum of the ventricle are the most commonly encountered heart lesions.^{3,9} The severity and the complexity of the intracardiac defect contribute largely to the poor prognosis associated with this malformation.³ Ectopia cordis has also been reported with other congenital anomalies such as abdominal wall defects, cranial and facial malformations, cleft lip and palate, anencephaly, hydrocephaly, neural tube defects, pulmonary hypoplasia, genitourinary malformation, gastrointestinal defect, and chromosomal abnormalities.^{1,4,10}

If the diagnosis of ectopia cordis is confirmed during the pregnancy, an early plan should be made for elective atraumatic cesarean delivery.¹⁰ Immediately after birth, the newborn should be stabilized and the lesion should be covered with saline-soaked gauze pads and wrapping to prevent desiccation and heat loss of the exposed viscera.⁴ After completing the preoperative evaluation, the patient should be taken promptly to the operation room for surgical repair of the defects.

The overall objectives of ectopia cordis management are: closure of the chest wall defect, including the sternal defect, repair of the associated omphalocele, placement of the heart into the thorax, and repair of the intracardiac

defect.^{6,9} Unfortunately, in most of the cases the thoracic cavity is small, and the mediastinum offers too little space for the heart. Attempts to close the chest wall after replacing the heart into the thoracic cavity often result in intolerable hemodynamic embarrassment secondary to kinking of the great vessels or compression of the heart muscle.^{1,6} Therefore, a staged repair oftentimes is a necessary approach to correct this anomaly. The first priority is to obtain coverage of the exposed heart. This can be accomplished in some cases by mobilizing the skin over the chest wall and directly closing the skin. If this maneuver seems to cause hemodynamic instability, then either a skin grafting or prosthetic patch should be considered.^{6,11} In our case, a Dacron patch was first placed to close the sternum defect, and then the skin flaps were approximated and closed. The Dacron patch offered good support to the thoracic cage and more protection to the heart into the thorax. This technique decreased the protrusion of the heart outside the thorax without compression. Some other techniques to repair the chest defect have been described in the medical literature. In one report, the authors illustrated the use of bilateral pectoralis major and rectus abdominis mucocutaneous flaps to repair thoracoabdominal ectopia cordis.⁴ Another report described an attempt to do a one-stage repair of ectopia cordis immediately after birth using transected ribs to reconstruct the thorax.¹³ The potential advantages of staged repair are to minimize the compression of the heart and the big vessel, and to allow the thoracic cavity to expand gradually.

After a successful first-stage operation to provide coverage of the heart, the subsequent operations aim to repair the intracardiac defect and to reconstruct the chest wall. In most of the reported cases, the intracardiac defect repair was performed after the first stage operation.¹ Nevertheless, if the intracardiac defect is "simple" and amenable to immediate surgical correction, it might be legitimate to repair it during the initial stage operation. In a case published in 1973, a repair of thoracoabdominal ectopia cordis and ventricular diverticulum was achieved successfully in a one-stage operation with the use of cardiopulmonary bypass.¹² More recently, an attempt to repair a double-outlet right ventricle and complete thoracic ectopia cordis was done in a single-stage operation immediately after birth. The newborn survived 12 days after surgery and died from sepsis.¹³

As the child grows, it becomes indicated to reconstruct the chest wall for protective and cosmetic reasons. Autologous rib grafts have been used with success to reform a bony thorax.⁶ This method is safe and reliable particularly if the amount of the native ribs is sufficient enough for graft donation without compromising the integrity of the patient's chest wall.² Another alternative is to use alloplastic materials to close the sternal defect and to reconstruct the chest wall. Unlike the initial first-stage repair of the chest that needs to be done soon after birth, the

second-stage surgery to reconstruct the chest wall is an elective procedure usually done after the first year of life.^{2,6} In our case, we expect that with the growth of the chest wall, the Dacron patch will gradually flatten and the protrusion of the heart will decrease. As the patch stiffens, it will provide a strong protection cover to the heart.

In conclusion, ectopia cordis is a rare congenital malformation which may require a staged procedure to achieve a complete repair. Historically, the prognosis of this condition is poor. However, with the advances in all aspects of medicine, the number of infants who undergo successful surgical repair and survive is steadily increasing.

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