when a pulmonary artery homograft can easily make connection to the native pulmonary artery bifurcation on either side of the aorta.

Hisashi Nikaidoh, MD
Children's Medical Center
1935 Motor St
Dallas, TX 75235

Single-Stage Repair of Thoracic Ectopia Cordis
Joseph J. Amato, MD, Jonathan Zelen, MD, and Nirupama G. Talwalkar, FRCS
Division of Pediatric Cardiothoracic Surgery, Long Island Jewish Medical Center, Schneider Children's Hospital New Hyde Park, New York

Thoracic ectopia cordis is a rare congenital defect most often seen in association with sternal and congenital heart defects. 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. Survival past the perioperative period is rare, with only 2 reported cases in the English-language literature. As with our case, neither had any discernable intracardiac defect. We present a case report of a patient with thoracic ectopia cordis repaired in a single stage using polytetrafluoroethylene membrane and skin for coverage and closure of the heart and thoracic defect.

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Address reprint requests to Dr Amato, Long Island Jewish Medical Center, The Schneider Children's Hospital, New Hyde Park, NY 11042.

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Thoracic ectopia cordis is a rare congenital malformation that presents a difficult surgical challenge. Ectopia cordis is classified into 5 types: cervical, cervicothoracic, thoracic, thoracoabdominal, and abdominal [1]. The two most common forms of ectopia cordis are thoracoabdominal and thoracic. Thoracoabdominal ectopia cordis frequently is associated with Cantrell's pentad [2] (bifid sternum, deficiency of the diaphragm, diaphragmatic pericardium, defects of the anterior abdominal wall, and intracardiac defects). Thoracic ectopia cordis is the most difficult to repair and maintains the highest mortality rate, with only 2 reported survivors in 29 attempted repairs [3, 4]. The orientation of the heart, associated intracardiac defects, small intrathoracic space, and lack of surrounding somatic tissue for coverage of the heart are the primary determinants of survival with the thoracic type [7]. A case of thoracic ectopia cordis repaired in a

A 2.6-kg female neonate was born with a complete thoracic ectopia cordis (heart without pericardial coverage) and omphalocele. The heart was covered with moist saline packs and the child was transferred to our facility within 2 hours of birth. On arrival, an echocardiogram was obtained, which revealed no intracardiac defect. The patient was brought to the operating room shortly after arrival. A mediastinal exploration was performed. The heart was totally extrathoracic (Fig 1). The manubrium was intact but the sternum was absent. An anterior diaphragmatic hernia and small omphalocele also were present. There was a patent ductus arteriosus and a large persistent left superior vena cava. The pleural cavities were entered and a large thymus gland was partially removed to make space in the mediastinum for replacement of the heart. An initial attempt at placing the heart in the thoracic cavity resulted in profound hypotension. The heart and great vessels were completely mobilized. A second attempt at placing the heart in the thoracic cavity also failed secondary to unacceptable hypotension and bradycardia. Inspection of the diaphragm revealed an anterior diaphragmatic hernia as well as a deficiency in the fascia of the anterior abdominal wall. The umbilicus was dissected with ligation of vessels and removed. The diaphragm and anterior fascial defect were approximated and closed. The left phrenic nerve was transected during the mobilization and repair of the diaphragmatic defect. The left costal cartilages were divided, creating a larger thoracic space. The heart then was placed into the left thoracic cavity. Hemodynamic stability was maintained without evidence of circulatory compromise. Gore-Tex surgical membrane and skin for coverage and closure of the heart and defect is presented.

Fig 1. The heart is totally extrathoracic with no pericardial covering. The apex of the heart points in an anterior and slightly caudal direction.
Thoracic ectopia cordis is a rare congenital malformation characterized by the heart being located outside the thoracic cavity, often involving the thoracoabdominal region. It is usually associated with a lack of pericardium, a condition known as the "naked heart." The heart may be partially or totally extrathoracic, with great vessels present within the thoracic cavity. Ectopia cordis is most commonly associated with Cantrell's pentad, a group of associated anomalies which include omphalocele, diaphragmatic hernia, craniofacial anomalies, and other peripheral defects.

The initial management of ectopia cordis is concerned with covering the heart and repairing the associated omphalocele. The heart is manipulated into the thoracic cavity without compromising hemodynamic stability through a staged approach, which may involve the use of polytetrafluoroethylene membrane and skin flaps. The procedure is performed on postoperative day 35, and the patient tolerated the operation well and subsequently was removed from the ventilator 2 days after the plication. The patient was discharged home on the 12th day after plication.

Comment

Ectopia cordis remains a rare congenital malformation involving the midline mesodermal components of the chest and abdomen. It is most often associated with defects in mesodermal maturation of the heart, sternum, rectus abdominis, diaphragm, and endocardium [6, 7]. The most common variety of ectopia cordis is the thoracoabdominal type, commonly associated with Cantrell's pentad. The most common intracardiac defects are ventricular septal defect, atrial septal defect, and diverticulum of the ventricle. The most common defects of the anterior abdominal wall are omphalocele, diaphragmatic hernia, diaphragmatic defect, and pericardial defect [2, 6, 7].

Thoracic ectopia cordis is distinguished from the thoracoabdominal type by its characteristic absent pericardium or "naked heart" emerging through the chest wall above the level of the diaphragm. The heart may be partially or totally extrathoracic with only the great vessels within the thoracic cavity. Frequently, the apex of the heart points in a cephalad direction, making reduction of the heart into the thoracic cavity without compromising hemodynamic stability a true surgical challenge [6, 7].

Survival of ectopia cordis patients is dependent on the presence or extent of associated intracardiac defects. All reports of survival in true thoracic ectopia cordis thus far have been in patients without concomitant intracardiac defects [7].

The initial management of ectopia cordis is concerned with covering the heart and repairing the associated omphalocele. Replacing the heart into the thoracic cavity may not be possible without hemodynamic compromise, and it usually requires a staged approach to repair. To manipulate the heart after several failed attempts to push the heart into the thoracic cavity, we placed traction sutures in the apex of the heart, which was brought out through the left chest wall. We subsequently were able to pull the heart into the left chest without hemodynamic compromise. Had we been unable to achieve hemodynamic stability by this approach, we would have elected to perform a staged repair with primary coverage as the first stage.

Primary coverage can be accomplished either by mobilizing skin over the chest wall or by using a polytetrafluoroethylene membrane and skin flaps, but not without great difficulty, as stated above. It continues to be our opinion that, if the patient is hemodynamically stable without a metabolic acidosis or functional compromise, an attempt at placing the heart within the thoracic cavity is a worthwhile endeavor. If this can be accomplished one may prevent long-term consequences of a scarified mediastinum with its inherent morbidity on subsequent procedures, in addition to the obvious physical deformity compared with our patient (see Figs 1 and 2).

Our desire to create a larger intrathoracic space during the repair of the diaphragmatic defect and omphalocele necessitated the transection of the left phrenic nerve. Thoracic ectopia cordis, with its absent pericardium, resulted in the phrenic nerve being confined to a fore-shortened mediastinum. In this case, the phrenic nerve could not be preserved if a single-stage repair was to be accomplished. This decision subsequently resulted in a prolonged period of mechanical ventilation and two additional operations for diaphragmatic plication. However, if faced with a similar decision, we would preserve the phrenic nerve only if it did not prevent the repair. The eventual aspiration of an oral feeding at 10 months of age was not related to the phrenic nerve injury. The phrenic nerve only if it did not prevent the repair.

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Our experience with this patient illustrates the complexities and controversies encountered with the repair.
of thoracic ectopia cordis. The paucity of survivors with this entity demonstrate 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

Systemic–Pulmonary Shunt With a Right Retroesophageal Subclavian Artery
Benoit Legault, MD, Lionel Camilleri, MD, Patrick Bailly, MD, Isabelle Brazzalotto, MD, Jean-René Lusson, MD, and Charles de Riberolles, MD
Department of Cardiovascular Surgery, Gabriel Montpied Hospital, Clermont-Ferrand, France

A 19-day-old child suffering from cyanosis due to tetralogy of Fallot was palliated by using his right retroesophageal subclavian artery. It was anastomosed side-to-side onto the ascending aorta and end-to-side onto the right pulmonary artery. The palliation obtained with this systemic-pulmonary shunt was satisfying. The right brachial vascular flow was normal.


Right retroesophageal subclavian artery is a frequent defect of the aortic arches, occurring in about 0.3% of living infants [1]. Occasionally, it is associated with other malformations such as tetralogy of Fallot and coarctation [2]. Rising from the descending thoracic aorta distal to the left subclavian artery, it passes through the posterior mediastinum behind the esophagus to join again its normal course toward the right arm.

Address reprint requests to Dr Camilleri, Service de Chirurgie Cardiovasculaire, Hopital Gabriel Montpied, Place Henri Dunant, 63003 Clermont-Ferrand Cedex, France.