Journal of Pediatric Surgery (2012) 47, E1–E4

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Repair of ectopia cordis using a resorbable poly-L-lactic-polyglycolic acid plate in a patient with pentalogy of Cantrell☆,☆☆

Hui-Ling Chia[a,*], Mohamed Zulfikar Rasheed[a], Kim-Kiat Ong[b], Vincent Kok-Leng Yeow[a]

[a]*Department of Plastic, Reconstructive and Aesthetic Surgery, KK Women's and Children's Hospital, Singapore 228 899*
[b]*Department of Cardiothoracic Surgery, KK Women's and Children's Hospital, Singapore 228 899*

Received 24 June 2011; revised 11 January 2012; accepted 11 January 2012

Key words:
Sternal reconstruction;
Ectopia cordis;
Pentalogy of Cantrell;
Resorbable plate;
Poly-L-lactic acid;
Polyglycolic acid

Abstract We present a case of a 10-month-old male infant with thoracoabdominal ectopia cordis, as part of Cantrell pentad, repaired using a poly-L-lactic-polyglycolic acid plate, a resorbable plating system widely used in craniomaxillofacial reconstruction. This is the first reported case of sternal reconstruction using a poly-L-lactic-polyglycolic acid plate. The repair was successfully carried out without cardiopulmonary compromise and good aesthetic outcome was achieved.

Ectopia cordis is a rare congenital anomaly where there is malposition of the heart, partially or completely outside the thorax. We report the first reported case of congenital sternal reconstruction using a resorbable poly-L-lactic-polyglycolic (PLLA-PGA) plate (Lactosorb; Walter Lorenz Surgical, Jacksonville, FL) in a patient with ectopia cordis, as part of Cantrell pentad [1].

1. Patient and method

An Indonesian-Chinese baby boy was born at full term to nonconsanguineous parents after an uncomplicated pregnancy and spontaneous vaginal delivery. Postnatal examination revealed an anterior thoracoabdominal defect with ectopia cordis and 2-dimensional echocardiography shortly after birth showed mesocardia, with moderate perimembranous ventricular septal defect (pmVSD) and atrial septal defect/patent foramen ovale.

The child was referred to Singapore for management at 10 months of age because of worsening cardiopulmonary function. On examination, there was partial herniation of the heart through a lower sternal defect, which was only covered by thin skin, allowing the cardiac impulse to be seen. An epigastric hernia was also present (Fig. 1). He was diagnosed with pentalogy of Cantrell, with features of congenital intracardiac defects and defects in the lower sternum, midline abdominal wall, anterior diaphragm, and diaphragmatic pericardium [1].

A repeat 2-dimensional echocardiography revealed a large pmVSD and patent ductus arteriosus, both with left-to-right flow and significant pulmonary hypertension. In

☆ Conflict of Interest Statement: The authors did not receive any funding and declare no conflict of interest in this present work.
☆☆ Implant: Lactosorb (Walter Lorenz Surgical, Jacksonville, FL).
* Corresponding author. Tel.: +65 9139 0179; fax: +65 6394 1524.
E-mail address: chiahuiling@gmail.com (H.-L. Chia).

0022-3468/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2012.01.072

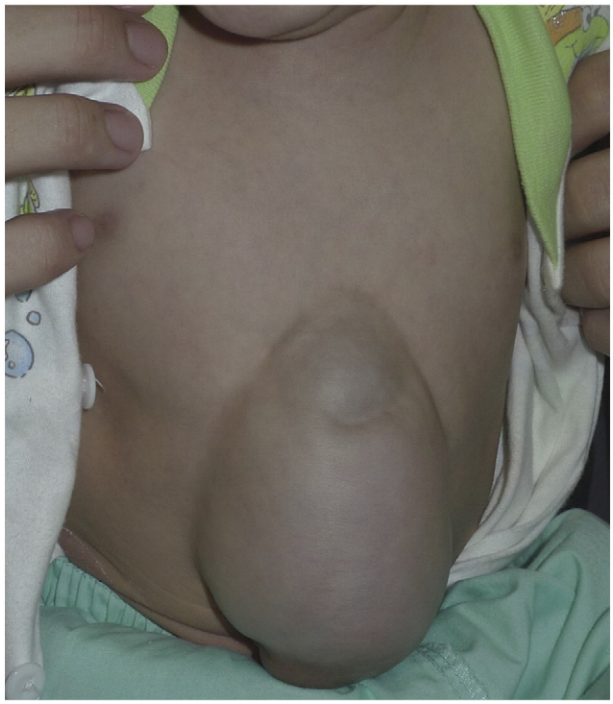

Fig. 1 Preoperative photo. Ten-month-old patient with abdominothoracic ectopia cordis, presenting with partial herniation of the heart through a lower sternal defect. This was covered by very thin skin alone, allowing the cardiac impulse to be seen. An epigastric hernia was also present.

view of the deteriorating cardiopulmonary function, correction of the intracardiac defects was planned. The child was referred to our unit for reconstruction of the sternal defect at the same setting.

Preoperative computed tomographic thorax showed deficiency of the lower sternum and pericardium, with heart protruding through the defect. There were defects in the anterior aspect of the diaphragm and abdominal wall, with an associated midline upper abdominal hernia.

During the operation, patch closure of the pmVSD, patent ductus arteriosus ligation, and patent foramen ovale closure was carried out by cardiothoracic surgeons, and the pericardium was closed with an expanded polytetrafluoroethylene (ePTFE) membrane. Apart from a large lower sternal defect, exposure of the underlying musculature showed underdeveloped pectoralis muscles and widely separated upper rectus muscles, which were attached laterally onto the hypoplastic lower ribs, leaving a deep V-shaped defect (Fig. 2).

Bilateral pectoral muscle and skin flaps were raised, and the superior edges of the rectus abdominis were detached from the costal margins. A resorbable PLLA-PGA plate was cut to shape, molded and inlaid into the lower sternal defect and free edge of the anterior diaphragm, and secured with 1/0 polypropylene sutures. Closure was achieved by approximation of the upper rectus muscles and pectoral muscle and skin flaps (Fig. 3).

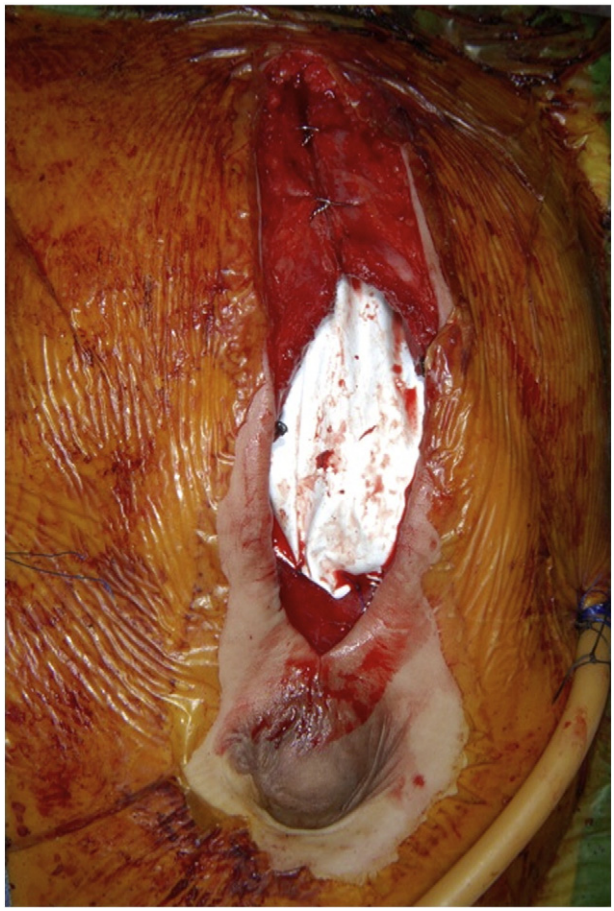

Fig. 2 Intraoperative photo. The pericardium was closed with an ePTFE membrane. Apart from a large lower sternal defect, the upper rectus muscles were also widely separated, leaving a deep V-shaped defect.

2. Results

The patient had a smooth postoperative course and was discharged on postoperative day 7. At 3-month, 15-month, and 3-year follow-up, the child was active and asymptomatic, and the lower sternum was firm to palpation (Fig. 4).

2.1. Comment

The principles for repair of ectopia cordis include the following:

1. Repositioning of the heart into the thorax without undue cardiopulmonary compromise.
2. Delay of reconstruction to approximately 2 years of age if there are minimal intracardiac defects (when the child gains more cardiovascular reserve and to allow growth of the thorax) [2].
3. Chest wall reconstruction to provide solid protection of the heart.
4. Preferable use of autologous tissue for reconstruction.