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Primary closure of thoraco-abdominal ectopia cordis

A. K. Singal, P. Balamourougane, V. Bhatnagar
Department of Paediatric Surgery, All India Institute of Medical Sciences, New Delhi, India

Correspondence: Prof. V. Bhatnagar, Department of Paediatric Surgery, All India Institute of Medical Sciences, New Delhi - 110029, India. E-mail: veereshwarb@hotmail.com

ABSTRACT

Thoraco-abdominal ectopia cordis is an unusual congenital anomaly. The case of a 45-days-old female child, who presented with a defect in the chest and upper abdomen and visible cardiac pulsations and bulge during crying, is being reported. There was an inferior sternal cleft with divarication of the recti up to the umbilicus. The cleft was covered with a thin translucent membrane. Primary repair was accomplished successfully without causing mediastinal compression.

KEY WORDS: Divarication of recti, primary closure, sternal cleft, thoraco-abdominal ectopia cordis

INTRODUCTION

The spectrum of defects in thoraco-abdominal ectopia cordis includes an inferior sternal cleft and diastasis recti. The heart maybe located in the thorax, but is covered by a membrane or thin skin.[1] These anomalies are rare and the challenge lies in closure of the thoraco-abdominal wall defect without causing compression of the heart. Various techniques are available for the repairing of these anomalies but primary closure, if feasible, is the technique of choice.[2-4] Though easy and less complicated in neonatal age, it often becomes impossible in older children due to loss of compliance of the chest cage. We report a child with thoraco-abdominal ectopia cordis who was successfully managed with primary repair.

CASE REPORT

A 45-days-old female child presented with defect in the chest and upper abdomen with visible cardiac pulsations and bulge during crying. There was a cleft of the sternum from manubrium downwards with involvement of the upper abdominal wall to just above the umbilicus. The cleft was covered with a thin translucent membrane in the chest and a somewhat thicker membrane lower down in the abdomen [Figure 1]. Associated anomalies included left-sided macrochelia, polydactyly in the hand and syndactyly in the foot. Echocardiogram showed a small PDA. During surgical repair, the membrane was found to be densely adherent to the pericardium and peritoneum. A small rent in the pericardium was repaired with fine absorbable sutures. The sternal halves were approximated in midline with 1-0 prolene sutures without much tension. The overlying soft tissues and muscles were closed over the repair with absorbable sutures [Figure 2]. A suction drain was placed underneath the flaps. Post-operatively the child required ventilatory support for 2 days. Suction drain was removed at 72 hours and the child was discharged uneventfully on day 6 [Figure 3].

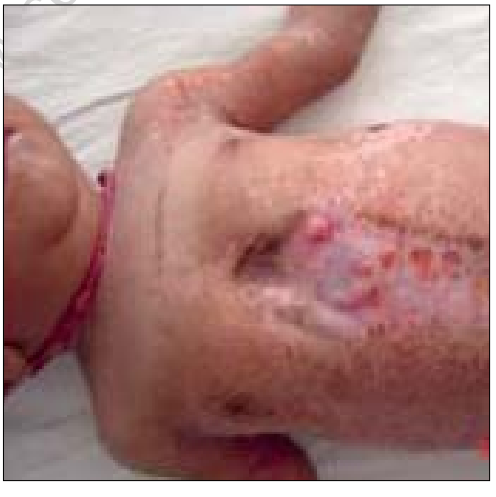

Figure 1: Thoraco-abdominal ectopia cordis showing an inferior sternal cleft with divarication of the rectus muscles. The defect is covered with a thin membrane through which the heart pulsations can be readily seen

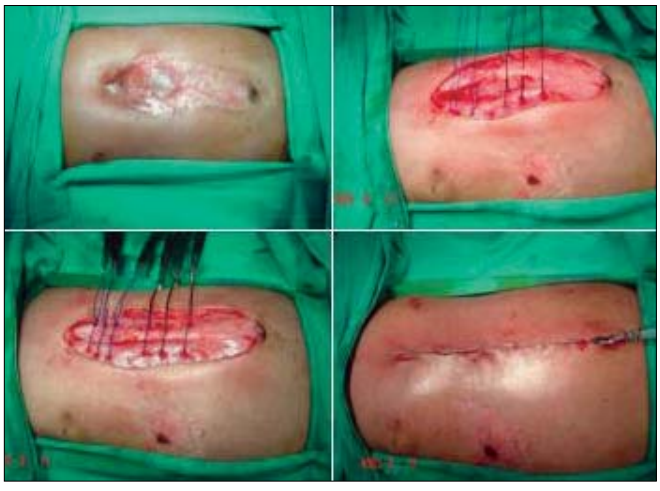

Figure 2: Steps in the primary closure of the thoraco-abdominal ectopia cordis

Figure 3: Post-operative appearance following primary closure

DISCUSSION

The sternum originates from two longitudinal mesenchymal bands, which appears at 6 weeks in the anterior thoracic wall of the embryo, independently of the ribs. These start fusing in a cephalocaudal direction from 7 weeks onwards and this fusion is complete by the 10th week. Similarly ossification starts at 5-6 months of embryonic life and proceeds in the cephalocaudal direction. Thus, it is believed that sternal defects result from the failure of the mesenchymal plate fusion process during the 8th week of gestation.[3,5]

In 1990 Shamberger and Welch suggested a classification for these patients dividing them into four types: (a) thoracic ectopia cordis, (b) cervical ectopia cordis, (c) thoracoabdominal ectopia cordis, and (d) sternal cleft or bifid sternum.[1,2] The simple sternal cleft should have an orthotopic heart, an intact pericardium and normal skin coverage. Isolated sternal clefts can also be classified as partial or total.[5] The partial forms can be superior, medium, or inferior. Pure inferior sternal clefts are extremely rare and are generally found in conjunction with thoracoabdominal ectopia cordis. In the present case the heart was in an orthotopic location and there was no major intracardiac anomaly except patent ductus. This association has been reported in literature albeit with a superior sternal cleft.[6,7] The membrane bridging the defect was thin and was adherent to the pericardium and peritoneum. Another interesting feature was divarication of midline abdominal musculature, which required correction.

Sternal defects should be corrected as lack of bony cage makes the mediastinal structures vulnerable to trauma and unsightly appearance. Later, the correction is difficult and there is a disturbance in respiratory and cardiac functions.[3] Many methods for repair of cleft sternum have been described ranging from sliding chondrotomies, repair of the defect with autologous grafts (costal cartilage, ribs, parietal bone) and prosthetic materials (Marlex mesh, teflon, silicone prosthesis). Direct approximation of the sternal halves or primary repair is preferable, because this technique reduces the problems with the prosthetic material and the difficulties in the osteocartilaginous cicatrisation, which occurs in the chondrocostal division procedures.[2-4,6-8] However, the age of the patients is the prime determinant for the feasibility of this procedure. During the neonatal period, the chest wall is more flexible and this allows an easy approximation of the sternal halves without mediastinal compression. After the neonatal period and certainly after one year of life, primary repair is difficult. In the present case the primary repair was accomplished successfully in a 45 days old child with a rare type of sternal defect.

Thoraco-abdominal ectopia cordis is an unusual congenital anomaly. Optimal correction can be obtained by primary closure of the defect in young patients, especially during the neonatal period, with an easy technique. In children above that age, more extensive techniques may be applied, although the results are satisfactory from a functional and aesthetic point of view.

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