ABSTRACT

Isolated cleft sternum is a rare congenital defect of the anterior chest wall and is the result of failed ventral midline fusion of sternal bands. We present two cases operated 18 and 24 days after birth with satisfactory results. Surgery is indicated to protect the heart and mediastinal contents.

KEY WORDS: Cantrell’s pentology, cleft sternum, neonatal repair

INTRODUCTION

Congenital midline cleft of sternum is a rare developmental anomaly, usually occurring as part of a defined syndrome, e.g., Cantrell’s Pentology. Isolated sternal cleft without any associated anomaly is very uncommon. It results from failed ventral midline fusion of the sternal bands, which normally occurs during the first three months of embryonic life. Isolated sternal clefts present in early infancy because of the obvious cosmetic deformity. The anomaly is more common in females and the association with midline supraumbilical raphe and facial hemangiomas is well known. Surgical treatment in the newborn period is preferred as the chest wall is pliable and the thoracic viscera accommodate easily to the closure of the chest wall. We present our experience with neonatal surgical repair of subtotal isolated cleft sternum.

CASE REPORTS

Case 1

A 20 days-old, full-term male baby weighing three kilograms and with an uneventful birth history presented with a palpable gap in the upper midline of the chest since birth. Crying resulted in a distinct bulge of precordium. A ‘U’-shaped subtotal sternal cleft up to the xiphoid process was obvious. Visible pulsations of the heart could be seen when the baby was asleep. Umbilicus was normal and abdominal examination was unremarkable. Echocardiography was normal. There was no other obvious congenital anomaly present. The patient was operated on day 24 after birth through a midline chest incision. The sternal bars were exposed and the pericardium and pleura separated from the undersurface [Figure 1]. The edges of the sternal bars were mobilized and approximation was achieved by prolene sutures inserted through the intercostal spaces. The patient’s hemodynamic status was monitored for five minutes before tying the knots. The patient had a hemodynamically uneventful postoperative recovery. At a three month follow-up visit, the child was seen to develop extensive facial hemangiomas [Figure 2]. A computerized tomography (CT) scan did not show any intracranial lesions. Oral Betamethasone was begun. Patient has completed one year of follow-up and is doing well.

Case 2

Our second patient was a 15 day-old, full-term male baby weighing 2.5 kg, who had normal Apgar scores at birth. The parents had a similar complaint of midline chest bulge on crying. He had a midline ‘U’-shaped sternal cleft. There was no other obvious congenital anomaly present. The Doppler echocardiography was
normal. The patient underwent sternal repair on the 18th day of life. Mobilization of the sternal bars was easily accomplished. The pericardium and pleura were intact. Satisfactory midline approximation of the two halves was possible without hemodynamic compromise. The baby was discharged on the 5th postoperative day. He remains well on follow-up at eight months of age.

**DISCUSSION**

The first case of sternal cleft was reported in 1740.[9] The first successful surgical repair of the anomaly was done by Burten in an 11 week-old child in 1943.[4] Since these reports, very few cases have been reported. Successful surgical repair in neonates has been reported by some authors. The earliest age at which repair was done was by Marcello et al.[5] in two patients 14 and 28 days after birth. Developmentally, the sternum is formed by two longitudinal mesenchymal bands independent of the ribs. These bands fuse in the craniocaudal direction by the 9th week of gestation. The ossification of this fused sternum starts at 5-6 months of embryonic life. Failed ventral fusion of these bands due to unknown reasons, results in cleft or bifid sternum.

The implicated etiologies include Riboflavin deficiency[6] and HOXB4 gene disruption.[7] The resultant anatomical defect produces a concave defect in the sternum covered by skin. The skin moves paradoxically with intrathoracic pressure changes. The shape of the defect can vary from a narrow ‘V’ to a wider ‘U’-shaped cleft. If the anomaly occurs as a part of upper midline syndrome, e.g., Cantrell’s Pentalogy, then associated diaphragmatic and pericardial defects are also present.

One of our patients had developed hemifacial and hemisternal hemangiomas which is a well known association. The timing of appearance of hemangiomas is variable and parents should be informed before surgery. Other associated conditions are supramullibal raphe, omphalocele and umbilical hernia. Intracardiac anomalies should always be anticipated, the common ones are ventricular septal defect (VSD) and Fallot’s tetralogy, which should always be ruled out by Doppler echocardiography.

The clinical presentation due to the defect is bulging of the skin with crying as seen in both of our patients. Some babies can have a low Apgar score at birth, with cyanosis and respiratory distress, all due to significant displacement in the position of heart or secondary ectopia cordis, which impairs the venous return. An electrocardiogram can show ventricular conduction disorders and extra systoles. Recurrent respiratory tract infection can occur due to reduced aeration of lungs.

Pre-natal diagnosis has been reported by USG. The appreciation of the defect should prompt the physicians to look for a cardiac anomaly. The spectrum of the defect varies from a small superior or inferior sternal cleft to a total cleft.[64] sometimes involving the cleft of the mandible also. There is a general agreement that the earliest possible primary direct closure is the best treatment.[6-8] The flexibility of the thoracic cage is inversely proportional to the age of the patient. Repairs done after the age of three months have always required more supportive postoperative care with higher incidence of cardiac complication. Beyond one year of age, autologous rib / coastal cartilage grafts / prosthetic materials are required. Good mobilization of the two sternal edges is the key to a secure approximation.

Wedge excision of the xiphoid process is required to approximate the lower edges. All the peristernal sutures are placed before tying the knots. Hemodynamic stability is reassessed by the anesthetist and after approximation, the knots are tied after waiting for a brief period. The patient is to be watched in the early postoperative period. The risk of complication decreases with the early age of repair. Facial hemangiomas developed in one of our patients and in the absence of intracranial lesions, were successfully treated with oral steroids. Both our patients remained well during follow-up. We conclude that early neonatal repair of cleft sternum is feasible and offers the best functional and cosmetic results.

Source of Support: Nil, Conflict of Interest: None declared.