Outcomes of primary repair of sternal cleft defects: Providing a "bony cover"

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ABSTRACT

Background : **Sternal clefts are rare congenital anterior chest wall defects created by a lack of midline thoracic fusion. Various surgical repairs have been proposed to provide protection to underlying viscera in these defects.**

Aim : **This study aims to perform primary sternal cleft repair using techniques, leading to the provision of a complete bony cover and to assess their outcomes on follow-ups.**

Materials and Methods : **During 2009–2020, seven patients were referred to our unit with sternal defects. Out of them, four infants with sternal clefts underwent primary repair using bilateral perichondrial flap creation of the sternal bars and sliding costal chondrotomy at our institute. In one of them with a wider defect, bilateral "intraperiosteal" sliding clavicular osteotomy was additionally performed to achieve tension-free closure.**

Results : **Satisfactory surgical outcomes were achieved with an uneventful postoperative period. On follow-up, all four patients are thriving well and have a stable anterior chest wall. Those with follow-ups longer than 5 years showed evidence of bone formation.**

Conclusion : **Bony cover to the heart can be provided in all varieties of sternal cleft defects using primary surgical repair early in infancy. The delay in surgical correction increases the complexity of the procedure and may require the use of prosthetic material which has its own disadvantages.**

Keywords : Inferior cleft, intraperiosteal osteotomy, perichondrial flap, sliding chondrotomy, Superior cleft

INTRODUCTION

Sternal cleft is a type of anterior chest wall deformity diagnosed clinically with paradoxical movement of the chest wall in the newborn. Patients may be asymptomatic or can have recurrent respiratory infections. Defects can be corrected with primary repair when presenting early or may require techniques involving the use of autologous or prosthetic materials in late presentation.[1]

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This study pertains to the discussion on the repair of sternal cleft defects and their outcomes.

MATERIALS AND METHODS

During 2009–2020, seven patients were referred to our unit with anterior midline parietal defects. One

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patient had abdominal ectopia cordis associated with intracardiac lesions, two patients had Cantrell's pentalogy, and four patients had sternal cleft defects with one of them having minor intracardiac lesion. Table 1 represents the demographics and clinical details of the patients.

On the evaluation of sternal clefts, paradoxical chest wall movements in the sternal region and pulsatile movement of the heart through the defect were noted. An inferior sternal cleft in a patient was associated with an anterior abdominal wall defect which was covered with a darkly pigmented parchment skin. The one with complete cleft and the remaining two superior clefts were noted to have a darkly pigmented midline raphe up to the umbilicus. Two-dimensional echocardiography was done in all cases to rule out associated cardiac defects. Thoracic computed tomography (CT) with 3D reconstruction done before presenting to us, confirmed the type of the sternal cleft. Primary surgical correction was performed in all four patients by the technique described here.

All procedures were performed after due informed consent. Under general anesthesia and orotracheal intubation, right internal jugular venous and invasive arterial pressure monitoring lines were placed. The surgical site, exposing the anterior neck, thorax, and supraumbilical abdomen, was prepared and draped. The midline incision was given on the skin well across the sternal defect. Utmost care was taken to avoid damage to underlying cardiac structures while separating the skin from the underlying pericardium. The thymus was excised. Bilateral skin flaps were raised to visualize the sternal heads of the pectoralis major muscle. Bilateral pectoral flaps based on the clavicular and acromial attachments were raised off from the anterior chest wall and lateralized. This exposes the whole length of bilateral sternal bars. In case 1, no connecting portion was present between sternal bars, whereas, in cases 2 and 3, the connected caudal ends formed a wide U-shaped cleft. A V-shaped wedge from the xiphoid connecting bridge was excised [Figure 1a]. In case 4 with an inferior cleft, a similar wedge is excised from the manubrial connecting bridge. To facilitate mobility,

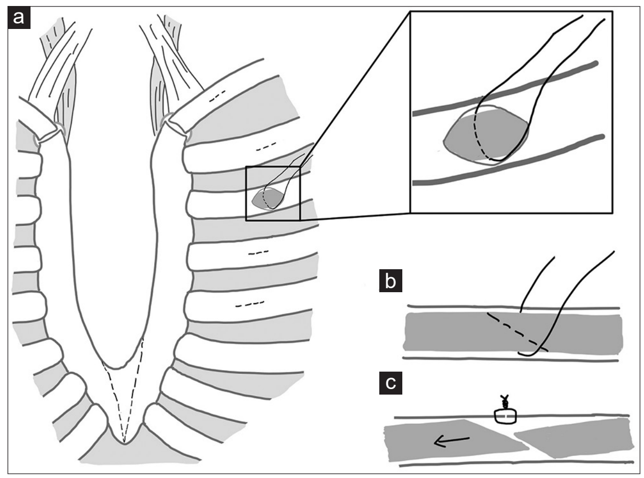

Figure 1: Schematic representation to perform sliding costal chondrotomy and clavicular osteotomy. (a) 2-0 silk suture is looped around the rib after opening the perichondrium and then creating space in between the rib and perichondrium (inset box). This maneuver is additionally repeated for periosteum over clavicles for wider defects. V-shaped wedge tissue excised from the xiphoid region (dotted lines). (b) Sliding chondrotomy and osteotomy were performed on both sides in an oblique manner (dashed line), in the second to fifth ribs and clavicle, respectively. (c) The arrow shows the direction of the rib being pulled medially without completely losing contact with its lateral counterpart during the medial approximation of sternal bars

Table 1: Patient clinical and demographic details

Age/gender	Weight (kg)	Associated defects	Procedure
		Abdominal ectopia cordis	
7 days/male	2.9	DORV, hypoplastic LV, PS	Abdominal relocation of the heart, PTFE patch cover over the heart, anterior abdominal wall defect
		Cantrell's pentalogy	
3 months/female	3.5	mesocardia, DORV, LV diverticulum herniating into omphalocele	Pulmonary artery banding, duomesh repair of the pericardium, diaphragmatic, and anterior abdominal wall defect
2.5 months/male	8	VSD, subaortic membrane	Subaortic resection, VSD patch closure, and anterior abdominal wall defect repair
1.5 months	3.4	Cantrell's pentalogy	Bilateral sternal bar perichondrial flap, and sliding costal chondrotomy

Sternal cleft defects

Case	Age/gender	Weight (kg)	Associated defects	Type of defect	Procedure	Length of hospital stay (days)	Follow-up duration (years)
1	16 days/female	2.1	None	Complete	Bilateral sternal bar perichondrial	6	11
2	2 days/male	2.75	None	Superior	flap, sliding costal chondrotomy	10	8
3	5 months/male	4.8	Small atrial septal defect, right undescended testis	Superior	Bilateral sternal bar perichondrial flap, sliding costal chondrotomy, and bilateral "intraperiosteal" sliding clavicular osteotomy	10	1

DORV: Double outlet right ventricle, LV: Left ventricle, PS: Pulmonary stenosis, PTFE: Polytetrafluoroethylene, VSD: Ventricular septal defect endothoracic fascia is separated from either side of the posterior aspect of the sternal bars. Perichondrial flaps are raised from the anterior aspect of the sternal bars bilaterally using endarterectomy dissectors and are brought medially [Figure 2a]. Small buttonholing encountered due to incorrect plane of dissection while raising flaps is of no major concern as this layer does not contribute to the immediate strength of the repair. Its integrity is of importance for new bone formation. For immediate strength, tension-free approximation of sternal bars is achieved by bilateral sliding chondrotomies as explained. To perform sliding chondrotomy, the second to fifth ribs on both sides are exposed. The perichondrium over these ribs is opened longitudinally for a length of 3–4 mm using #15 surgical knife [Figure 1a]. The chondrotomy is performed beginning from the anterior surface of the exposed rib in a posterolateral direction, obliquely, to facilitate a sliding motion when pulled apart without completely losing approximation [Figure 1b and c]. To aid the placement of oblique incisions while preventing inadvertent opening of the underneath pleura, a 2-0 silk suture is looped around the rib inside the perichondrium and pulled anteriorly during chondrotomy [Figure 1a inset]. In a patient who was 5 months old with a wider defect, this approximation could not come together easily. To facilitate this, the clavicular periosteum at the junction of its medial third and lateral two-thirds was opened longitudinally for 5 mm, and intraperiosteal

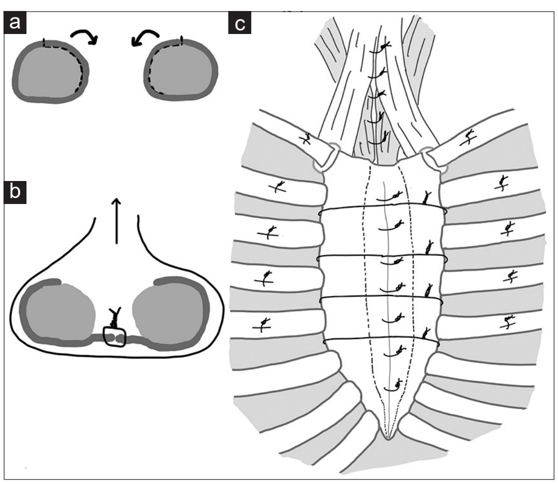

Figure 2: Schematic representation to perform sliding costal chondrotomy and clavicular osteotomy (continued) (a) Transverse cut sections of the sternal bars show perichondrial flaps being raised using endarterectomy dissectors along the dashed line and are mobilized medially as shown by curved arrows. (b) Perichondrial flaps, hence, raised are approximated in the midline using interrupted 5-0 polyglactin sutures. This is assisted by anterior traction applied in the direction of the arrow on the 2-0 polyethylene sutures placed around both sternal bars. (c) All four 2-0 polyethylene peristernal sutures are tied to facilitate sternal bar approximation. The perichondritis and periosteotomy on both sides are closed using interrupted 5-0 polydioxanone sutures. Strap muscles are also approximated in the midline

sliding osteotomy was performed bilaterally. The previously raised perichondrial flaps from the sternal bars were approximated in the midline using multiple interrupted 5-0 polyglactin sutures on a round-body needle [Figure 2b]. A row of four 2-0 polyethylene sutures is placed around both sternal bars and tied to facilitate sternal approximation. Heart rate, blood pressure, central venous pressures, and tidal volume were carefully monitored to look for any evidence of mechanical tamponade. The perichondritis and periosteotomy were closed using interrupted 5-0 polydioxanone sutures. Subxiphoid fascia was approximated in the midline and pectoralis muscle flaps approximated over the sternum using interrupted 3-0 polyglactin sutures. Strap muscles attached to the superior surface of sternal bars were also mobilized and approximated in the midline using 3-0 polyglactin sutures [Figure 2c]. A retrosternal and a sub-pectoral closed suction drain was placed. Simple interrupted sutures were placed for skin approximation using 3-0 nylon suture. Intraoperative images depicting various stages of the procedure have been shown in Supplementary Figure 1.

RESULTS

Postoperatively, intravenous paracetamol (15 mg/kg/dose, 6 hourly) and infusion of fentanyl (1–2 mcg/kg/min) were prescribed for adequate analgesia and sedation. Elective ventilation was provided for 36–48 h due to the possibility of a flail chest physiology. All operated cases were extubated on the 2nd postoperative day. The postoperative period was uneventful. Serous drainage occurs for 5–8 days. Drains were removed only after their yield became insignificant (≤1 ml/kg/day). Patients were discharged in a clinically stable condition on the 6th to 10th postoperative days (mean of 8.6 days).

On respective follow-ups [Table 1], stable covering over the sternal area and completely healed wounds in all patients were noted [Figure 3a]. Chest X-ray (CXR) with a modified projection (posteroanterior-right anterior oblique view with 15° off-center) was done on children beyond 8 years of age. In cases 1 and 2, bony margins were noted on both sides of the newly formed sternal body [Figure 3b]. [Figure 3d].

DISCUSSION

Congenital midline sternal defect is a rare anterior chest wall deformity with an incidence of 0.15% of all chest wall malformations.[1] The sternum is a mesodermal structure and starts developing as a pair of mesenchymal bars at 6 weeks of gestational age. Cells from both sides migrate and fuse in the midline by the 10th week of gestation and failure of this fusion causes the sternal cleft defect.[2] Sternal bone ossification centers start

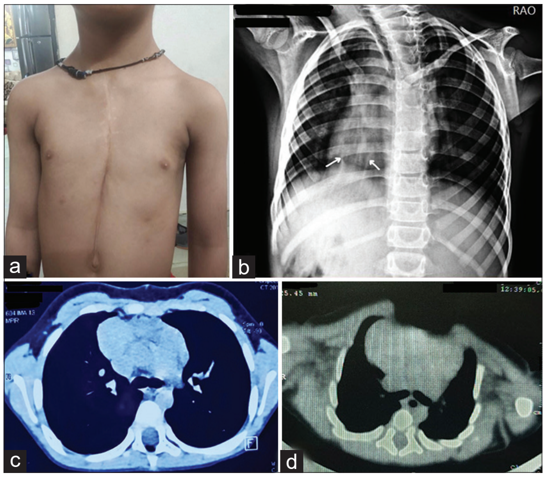

Figure 3: Follow-up of case 2 after 8 years postoperatively. (a) Well-healed midline scar. (b) CXR with modified PA-RAO view showing calcified margins (white arrows) of the newly formed sternum. (c) CT image at T4 level showing a sternal body growth, covering the defect as compared to preoperative CT image showing the sternal cleft defect at the same level (d). CXR: Chest X-ray, CT: Computed tomography, PA-RAO: Posteroanterior-right anterior oblique

coalescing at 6 years and are complete by 12 years, and calcification is achieved by 25 years of age.[3]

The first anatomic classification of sternal defects was given by Weese in 1818.[4] In 1958, Cantrell reported a congenital syndrome known as Cantrell's pentalogy with five anomalies involving midline supraumbilical abdominal wall defects, lower sternal defect, anterior diaphragm defect, pericardial defect, and intracardiac defects.[5] Later, Ravitch further classified sternal defects as sternal cleft without associated anomalies, true ectopia cordis with varying degrees of sternal cleft, and Cantrell's pentalogy.[6] Ectopia cordis is the defect with complete (where, the heart without a pericardial covering, is displaced outside the thoracic cavity) or partial displacement (pulsating heart can be seen through the overlying skin) of the heart outside the thoracic cavity.[2] Shamberger, in 1990, classified sternal defects into four types: thoracic ectopia cordis, cervical ectopia cordis, thoracoabdominal ectopia cordis, and sternal cleft defects.[4] Sternal clefts are classified as complete or partial, based on the absence or presence of a connecting portion between the two sternal bars. Partial defects can be superior or inferior, with the superior type being the most common and usually an isolated abnormality. This defect can be associated with PHACES syndrome which includes posterior fossa malformations, hemangiomas, arterial anomalies, cardiac lesions, eye abnormalities, sternal cleft, and supraumbilical raphe. Inferior cleft defects can be associated with Cantrell's pentalogy.[2]

A defect is clinically evident making the need for additional radiological testing redundant. An echocardiogram must be performed to rule out associated cardiac defects. CXR and CT scans are not mandatory for diagnosing *per se* but may shed light on associated defects. Surgery is recommended to provide protection to the heart and prevent recurrent respiratory infections due to paradoxical respiratory movements.

The first primary surgical closure was done in 1949 by Maier and Bortone on a 6-week-old infant.[3] Sabiston, in 1958, described sliding chondrotomies through bilateral costal cartilages which increases their effective lengths, thus helping in a medial approximation of the defect.[7] Another method, by Meissner, reported dividing the costal cartilages laterally and rotating them medially to cover the defect.[8] Mathai recommended fracturing the clavicles just lateral to the protuberant medial third to help in the approximation of the upper sternum which is otherwise prevented by sternoclavicular joint (SCJ).[9] Disarticulating SCJ or incising anterior sternoclavicular ligament may also assist in upper sternal approximation. Primary closure of the defect has been singularly reported by aggressive chest wall mobilization utilizing diaphragmatic detachment and wide opening of the mediastinal pleura. Although the approximation is feasible, may be limited in application for wider defects owing to ensuing mechanical tamponade.[10] In partial sternal clefts, a direct approximation can be facilitated by converting them into a complete defect by excising a sagittal wedge from the connected part of the sternum and then approximating the freshened edges. Monitoring the hemodynamic parameters during the juxtaposition of the sternal bars must be done before the final apposition. The use of various prosthetic materials such as stainless steel mesh, Marlex, acrylic, silicone, titanium plate, and Teflon have been reported.[1,8] Biological repair materials such as porcine dermal matrix tissue and autologous tissues such as costal cartilage, rib, iliac crest, and muscle graft interposition have also been used for these repairs.[1] Primary surgical correction has the direct advantage of reducing the problems with the prosthetic materials and the difficulties in the osteocartilaginous cicatrization during chondrocostal division process.[2] Table 2 enlists various surgical techniques available for sternal cleft repair.

CONCLUSIONS

Primary surgical repair when done preferably in early infancy provides the best results due to pliable bones and cartilages of the chest wall. With advancing age, procedural complexity increases due to reduced pliability of chondral cartilages, an increase in the gap of the sternal defect to be bridged, and reduced mobility of the rib cage and clavicle. Primary repairs also provide for normal sternal growth and development with good functional and cosmetic outcomes along with a reduced risk of tissue reactions and infections.[2,14] The described technique of intraperiosteal sliding clavicular osteotomy

Table 2: Armamentarium of surgical techniques

Primary surgical correction
Direct surgical approximation of sternal bars[3]
Sliding costal chondrotomy[7]
Division of costal cartilages laterally and rotating them medially[8]
Fracturing the clavicles[9]
Sternoclavicular joint disarticulation[1]
Extensive chest wall mobilization[10]
Use of prosthetic materials[1,11]
Stainless steel mesh
Marlex
Acrylic
Silicone
Titanium plate
Teflon
Use of autologous materials[1,12]
Costal cartilage
Ribs
Iliac crest
Parietal bone of the cranium
Tibia
Muscle graft interposition
Use of biological materials
Porcine dermal matrix tissue[13]

and the selection of the site for periosteotomy at the junction of medial third and lateral two-third provides better healing and minimizes chances of nonunion, while also preserving long-term functionality of pectoral girdle and integrity of SCJ vis-à-vis disarticulation of SCJ or fracturing the clavicle. CT evidence of the ossification further endorses our objective of providing bony cover to the heart. Our results suggest that using these techniques, a safe, lasting bony cover can be obtained in all types of sternal defects preferably performed in early infancy.

Informed consent

Informed consent was obtained from the participants.

Ethical statement

This study is an original work of the authors and has not been submitted to any other journal for simultaneous consideration. All procedures performed in this study were in accordance with ethical guidelines including informed consent obtained from the participants.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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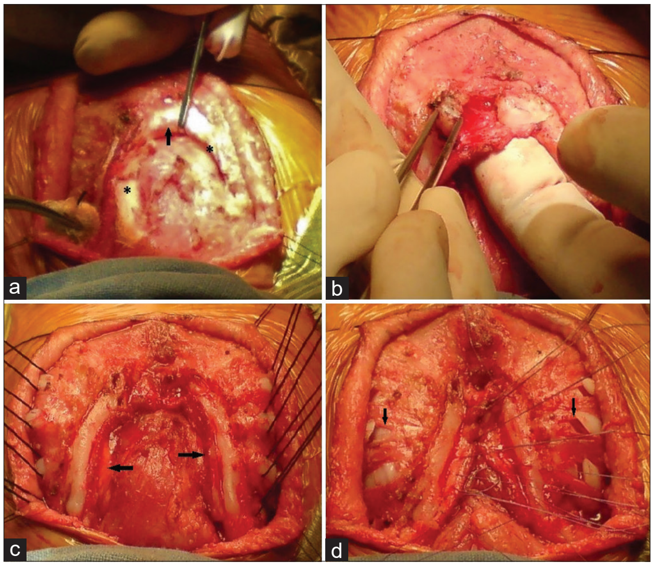

Supplementary Figure 1: Intraoperative images. (a) Raised skin and pectoral flaps exposing the sternal bars (asterisk) with a U-shaped cleft (arrow). (b) A V-shaped wedge from the xiphoid connecting bridge was excised. (c) Perichondrial flaps (arrows) are raised from the anterior aspect of the sternal bars bilaterally and are brought medially. 2-0 silk sutures are looped around the second to fifth ribs through the rib perichondrium and pulled anteriorly to aid in performing sliding chondrotomy. (d) Oblique sliding chondrotomies (arrows) are performed bilaterally and perichondrial flaps from the sternal bars were approximated in the midline