CAS CLINIQUE / CASE REPORT

THE CLEFT STERNUM
A POSSIBLE ROLE FOR PLIABLE PROSTHETIC RECONSTRUCTION

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ABSTRACT • **Background** : Faced with our intraoperative inability to primarily close a very wide isolated cleft sternum of a pediatric patient without causing cardiovascular decompensation, we describe our use of a synthetic material for partial approximation allowing muscular coverage. **Method** : We report an infant who was born with an isolated large complete sternal cleft where a trial of primary surgical repair had to be abandoned because of cardiovascular compromise. A similar difficulty was encountered in approximating the origins of both pectoralis major muscle flaps at the midline. Thus, a pliable synthetic patch was helpful in partially obliterating the widened sternal defect allowing successful muscular coverage. **Result** : Closure of a wide congenital sternal cleft using a synthetic material that partially obliterated an otherwise widely separated cleft sternum which was neither amenable for primary repair, nor by bilateral pectoralis major advancement alone, was possible. **Conclusion** : Strong prosthetic pliable material may offer a simple and useful procedure allowing obliteration of the widely separated cleft sternum not amenable for primary repair.

Keywords : sternum, thoracoplasty, prosthesis

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RÉSUMÉ • **Contexte** : Face à notre incapacité préopératoire de fermer un très large sternum creux chez un patient pédiatrique sans provoquer une décompensation cardiovasculaire, nous décrivons notre utilisation d'un matériel synthétique pour rapprochement partiel permettant une couverture musculaire. **Méthode** : Nous présentons dans ce rapport un enfant né avec une fente sternale complète isolée où un essai de réparation chirurgicale primaire a dû être abandonné en raison de troubles cardiovasculaires. Une difficulté similaire a été rencontrée dans le rapprochement des origines des deux grands lambeaux pectoraux musculaires à la ligne médiane. Ainsi, un patch synthétique souple a donc été utilisé pour effacer partiellement le défaut sternal élargi permettant une couverture musculaire réussie. **Résultat** : Fermeture d'une fente sternale congénitale large en utilisant un matériel synthétique qui a partiellement effacé un sternum de fente contraire largement séparé qui ne pouvait être réalisé ni par une réparation primaire, ni par un avancement bilatéral pectoral majeur. **Conclusion** : Le matériel souple prothétique fort peut offrir une technique simple et efficace permettant l'effacement d'une fente sternale largement séparée dont la réparation primaire s'avère impossible.

INTRODUCTION

Failure of ventral midline thoracic fusion can present itself as a spectrum of abnormalities including a prominent suprasternal notch, irregularities in shape of the xiphoid, ectopia cordis, superior sternal cleft and the rarest of all, the isolated complete sternal cleft which appears to represent a separate developmental field defect with probable multifactorial etiology [1]. The classical condition is not associated with extrinsic positioning of the heart outside the anterior chest wall nor with intrinsic malformations. Primary closure with or without pectoralis muscle is the golden standard for management, however, in very large defects pliable prosthetic reconstruction might provide the surgeon with a viable option for reducing the size of the defect allowing tension-free pectoralis muscle closure.

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MATERIALS AND METHODS

Two months following birth, a full term female infant was transferred to our institution with the diagnosis of a large congenital anterior chest wall defect.

Upon admission, the infant weighed 3.450 kg, had 96% oxygen saturation with stable vital signs. Patient had a complete clefting of the sternum 8 cm large with a pulsating heart covered with a thinned out caudal area of skin. A midline umbilical raphe was connecting the midsternal area down to the umbilical stump.

There was no associated abdominal or neck abnormalities. A large right facial hemangioma was evident (Figure 1).

Positive findings by echocardiography included isolated dextrocardia, a small patent ductus arteriosus and patent foramen ovale. Brain MRI revealed hypoplasia of the right cerebellum, absence of the inferior aspect of the vermis.

Patient being stable, the surgical team decided to delay the procedure; however, both parents were alarmed and expressed their concerns in taking care of their baby with a pulsating heart under a very thinned out skin.

RESULTS

At the age of nine weeks surgical intervention was performed and included excision of the ulcerated skin to reveal an absent pericardium, intact diaphragm, exposed pulsating myocardium. Bilateral pericardial flaps were then successfully raised, advanced and opposed under minimal tension with absorbable material to cover the heart. An attempt of primary repair to approximate the sternal ends resulted in a drop of oxygen saturation, blood pressure and severe decrease of venous return. Both pectoralis muscles were completely released from their origins and insertions followed by a trial of approximating them together in the midline which again failed due to the width of the intervening defect.

Thus, under close cardiovascular monitoring, a thin walled 0.8 mm, 3 x 6 cm GORE-TEX® graft (W.L. Gore & Associates, Flagstaff, AZ) was tolerated to narrow the defect to 2 cm and sutured to both edges of the sternum with 3.0 PROLENE® (Ethicon Inc. Johnson & Johnson Company, New Brunswick, NJ) (Figure 2). This allowed both pectoralis muscles to be sutured together in the midline covering the graft by using absorbable 3.0 MONOCRYL® sutures (Ethicon Inc. Johnson & Johnson Company, New Brunswick, NJ). Minimal tension was required to close the skin over a pericardial drain. The postoperative course was uneventful. Upon follow-up eight months later, the patient had a stable anterior chest wall, however, with progression of her right hemi-facial hemagioma (Figure 3).

DISCUSSION

The body of the sternum is developed from the lateral plate mesoderm where cells at the sixth intra-uterine week start migrating ventrally to form two parallel mesenchymal bands fusing by the tenth week. Between 1888 and 1977 only 44 cases by Ravitch [1] were reported. This was followed by Longino *et al*. [2] who, in 1955, gave a very good description of the entity which he found to be different from other partial i.e. upper cervical or/and abdominal entities which are associated with ectopia cordis.

In 1958, David Sabiston Jr [3] described costal cartilage cuts in the first, second and third ribs making use of periosteal flaps to aid in successful closure of large defects not amenable for primary closure. In 1975, Verska used division of alternating coastal cartilages with interposition at different levels for the same reason [4]. Bone (cranial, iliac, ribs) and cartilage grafts were also used for filling the gap and attaining stability when the defects were too large to close primarily [4-5].

With the advent of pectoralis muscle flaps in the eighties, autologous repair for further padding and coverage was possible as described by Snyder *et al*. [6] who believed that *"secondary osteocartilagenous reconstruction, if required, should be facilitated by the medial traction produced with this muscle closure."*

Although we agree with the experience of others that

Figure 1. Patient immediately preoperatively with the exaggerated mid sternal cleft, the midline umbilical raphe and barely visible facial hemangioma.

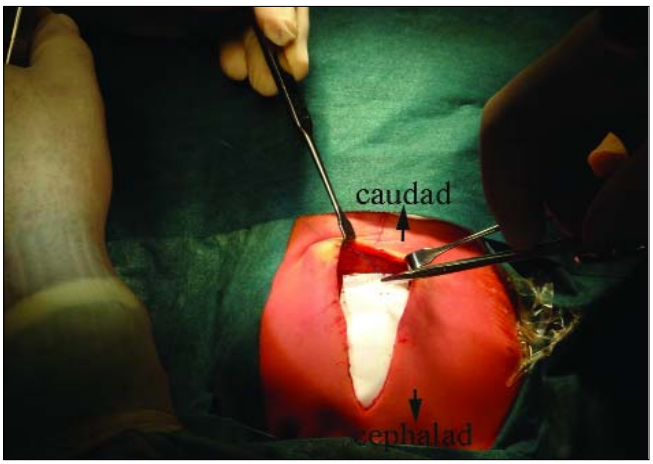

Figure 2. Prosthetic graft being sutured under tension narrowing the defect to 2 cm without cardiovascular compromise.

Figure 3. Patient 8 months postoperatively with the chest appearance. Notice the associated increasingly apparent right facial hemangioma.

Figure 4. Patient one year and a half postoperatively with the sternal appearance. Notice the improvement of the facial hemangioma after Propranolol® therapy.

primary closure, when feasible, is the simplest and best solution to restore bony integrity to the mediastinal structures, prevent paradoxical motion of the thoracic viscera and to eliminate visible deformities [7-8], yet, provision of normal intrathoracic pressure might not be possible. Thus, the above-mentioned circumstances might not be attainable in all patients [9-11]. Previous authors have used synthetic material such as stainless steel mesh, silicon sheets, TEFLON® [1] (DuPont, Wilmington, DE), acrylic [12], MARLEX® (C.R. Bard, Inc., Murray Hill, NJ), PROLENE® mesh, yet synthetic materials do result in tissue reactions and increased risk of infection [1]. Recently, PROLENE® has been used for reconstructions that resulted from radical resection of hemangiomas of the sternum [13], similar to the previous use in reconstruction of sternal defects resulting from primary malignant tumors [14]. The strength, pliability and the minimal amount of the material that we used was found to be advantageous in this particular situation that presented itself with a large defect.

We have found that the controlled use of pliable material such as GORE-TEX® which is used in vascular surgery, due to its strength, smooth nonabrasive surface and low incidence of infection is a viable option in this extremely rare condition when primary closure with bilateral pectoralis major muscles is not possible making it a relatively simple, atraumatic, first stage procedure where a gradual approximation of the sternal cleft can be safely achieved to allow muscle coverage. It was our observation at one year and a half that the defect was barely visualized or palpable due to normal chest expansion (Figure 4).

DISCLOSURE: "The authors have no financial interests in any of the products, devices, or drugs mentioned in this manuscript."

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