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Sternal Cleft: About Three Cases

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Abstract

The sternal cleft or the bifid sternum is due to a defect of fusion of the sternum and its incidence is estimated to be 0.15% of all malformations of the thoracic cavity. The etiology of sternal cleft remains unknown. Primary closure in the neonatal period remains the unanimous recommendation. However, surgeons are faced with neglected slits that sometimes extend into adolescence. In this work, we discuss our management of sternal cleft in three patients compared to that reported in the literature. The first patient is a child aged 2 years and 6 months, admitted for a higher sternal cleft. Both clavicles were 54 mm apart with no other associated abnormalities. It benefited from a primary closure with good evolution. The second patient is a child aged 2 years and 8 months, admitted with an upper sternal cleft associated with vascular malformations and labial angioma. It benefited from a thymectomy plus primary closure with good evolution. The last patient is a child aged 2 years with a complete sternal cleft. She had a primary laparoscopic closure. The aftermath was marked by a partial recurrence of the suprasternal part which was resumed by classical surgery with a good evolution. Whether the sternal cleft is symptomatic or not, surgical treatment is recommended to protect the heart and large vessels from trauma on the one hand and to improve respiratory dynamics on the other. Primary closure is best during the neonatal period. The imminent risk of compression of the underlying organs, against indicates direct closure. Several processes are then proposed and all have the same objective: to gain in length and thus obtain a cover with the least risk of compression. In our patients, despite their ages, the defect lent itself to a primary closure without tension, which justified our therapeutic choice. It was possible by conventional surgery and laparoscopy. We admit that this primary closure can always be preferred when the residual compliance of the rib cage allows a closure without risk of compression of the underlying organs, whatever the age.

Subject Areas

Pediatrics, Surgery & Surgical Specialties

Keywords

Sternum, Child, Sternal Slit, Surgery

1. Introduction

Sternal cleft or bifid sternum is a rare congenital malformation due to a defect in fusion of the sternum. It is the rarest of thoracic malformations where its incidence is estimated at 0.15% [1] [2] [3].

Sternum development begins in the sixth week of gestation as two parallel mesenchymal condensation bands. Cells migrate from both sides of these bands to fuse on the midline around week 10. At the same time, the chondification of the bands begins and appears a cranial rudiment called pre sternum. Ossification begins at the manubrium and upper sternal body in the sixth month, then at the mid-sternal body in the seventh month, and the lower body in the first year of life. Finally, that of xyphoide continues gradually between five and eighteen years [1].

The sternal cleft is therefore the result of a failure to fuse the mesenchymal bands. Although its first description dates from 1740, the etiology of sternal cleft remains unknown to this day. Studies suggest the expression of the Hoxb gene as a possible contributing factor but no family predisposition has been reported. However, alcohol intake and methylcobal-amine deficiency were associated with female predominance [1] [3].

The diagnosis is obvious and the cross-sectional imaging allows confirming the diagnosis, assess the extent of the abnormality and look for the associated malformations. If no treatment protocol is established, primary closure in the neonatal period is strongly recommended. However, all surgeons are still faced with neglected slits up to adolescence [3] [4].

2. Observation

We discuss in this work our management of sternal cleft in three girls compared to that reported in the literature.

Observation 1: A girl aged 2 years and 6 months with no special family history; admitted for sternal cleft observed since birth. According to the parents she had a good adaptation to the extra uterine life without cyanosis or respiratory distress and she remained asymptomatic. The aesthetic harm motivated the consultation for a takeover. The clinical examination found a patient in good general stable condition, a U-shaped depression in the upper sternal region during inspiration (**Figure 1**) and at the cries a mass occupies this extended depression on 8 × 6 cm (**Figure 2**). This mass is soft and painless with perception of heartbeat. The rest of the clinical examination was without particularity. At the x-ray of the chest, there was an absence of ossification of the sternal body; 3D reconstruction CT confirms the upper sternal cleft extended 47.7 mm long by 46

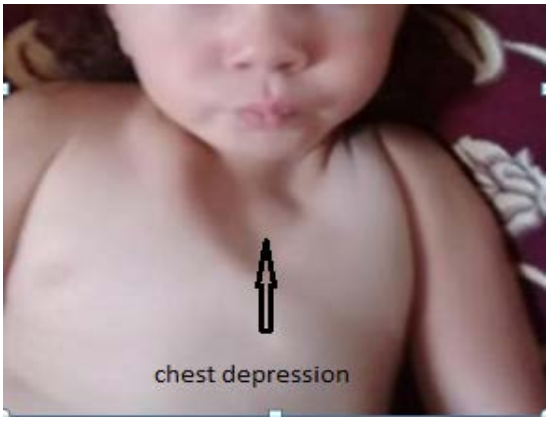

Figure 1. Clinical image of an upper sternal cleft.

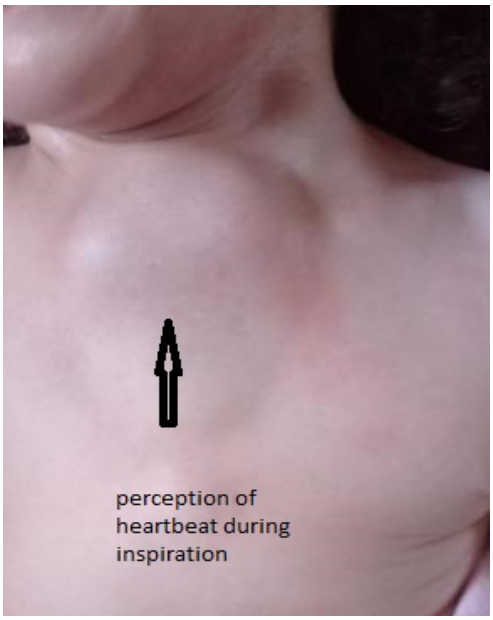

Figure 2. Clinical image of the crack with swelling during calls.

mm wide. The two collarbones were 54 mm apart (**Figure 3**). *No other scannographic abnormalities were found and the echocardiography was free of abnormalities*.

Under general anesthesia after a median longitudinal incision, we performed a dissection of the two sternal bars, release of the pectorals from their sternal insertion and dissection of the endothoracic fascia relative to the sternal bars (**Figure 4**). A cartilaginous wedge was resected to transform this malformation into U in the form of V. After analysis of the thoracic compliance, the primary closure seemed possible. The two sternal bars were reconciled by points separated at vicryl 0 after avivement of the banks (**Figure 5**), suture of the pectorals on the median line and closure on the sub-sternal drain.

The post-operative follow-up was simple with no signs of chest compression, drains removed after 48 hours and exited on the fifth post-operative day. At the last clinical and radiological check-up the patient was in good condition and

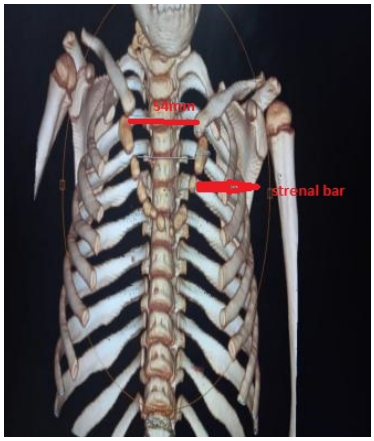

Figure 3. Chest CT with 3D reconstruction 47.7 mm long by 46 mm wide. The two clavicles were 54 mm apart.

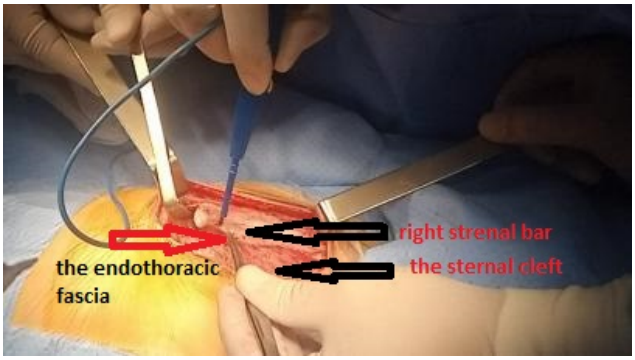

Figure 4. Operative image dissection of the thoracic endo fascia relative to the sternal bars.

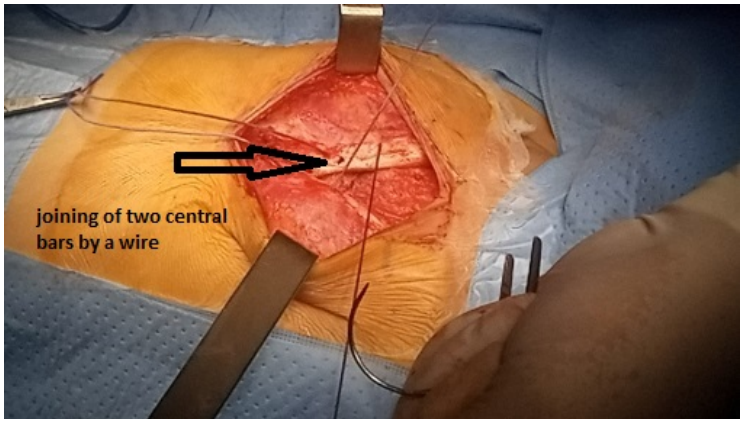

Figure 5. Operative image shows primary closure by stitches separated from the two sternal bars.

asymptomatic.

Observation 2: A girl aged 2 years and 8 months, admitted for sternal cleft diagnosed since the age of one month, no other clinical signs associated. At the CT, there was an incomplete upper sternal V-fissure and an abserent birth of the right subclavian artery on the descending portion of the aortic arch and goes in-

to the esophageal retro. She also has a labial angioma. It benefited from a thymectomy set up a bovine patch in pre-cardiac then primary closure of the sternal cleft by separate points to vicryl 0. The post-operative follow-up were simple with good clinical evolution.

Observation 3: A 2-year-old girl with complete sternal cleft with associated supra sternal cleft diagnosed at birth. It benefited from a primary closure by coelioscpie by steel wire according to the same principle as conventional surgery. The evolution was marked by a recurrence of the supra sternal part which was repeated twice without success. The evolution of sternal cleft was good without recurrence.

3. Discussion

The sternal cleft is classified as a partial or complete slot. Partial slits may sit on the lower, middle, or upper part. The top slots are the most common and are almost always isolated. Occasionally the slit is found in a syndromic form like PHACES syndrome for the higher variety (acronym for cervical or facial hemangioma, posterior fossa malformations, vascular malformations, aortic coarctation and ocular abnormalities) and Cantrell pentalogy for the lower variety.

Our second observation is an upper sternal cleft embedded in PHACES syndrome.

Depending on the appearance, the slot may be minor with a V-shape or have a U-shape. It may extend from the manubrium to the third or fourth intercostal space [1] [2].

Indeed, all surgeons are faced with neglected sternal slits with a delay of consultation that sometimes extends to adolescence. Extreme ages of 19 years are reported in Brazil and 23 years in Morocco [3] [4]. However, it should be noted that all these slots were diagnosed either from the neonatal period or before the first year of life. This implies a common responsibility for caregivers and parents in this delay in taking charge. The scarcity of the condition and the lack of information on the pathology would contribute to this delay in management.

Symptomatic forms often appear from birth with cyanosis attacks, dyspnea of varying severity, recurrent lung infections [1]. The diagnosis is often evident at birth through clinical examination. The pre-operative check-up should include echocardiography for associated cardiac malformation, a chest x-ray supplemented with CT with 3D reconstruction to better assess the extent of the abnormality as well as other associated malformations. The injected scanner will eliminate undetected vascular malformations on echocardiography [1].

Even for asymptomatic forms, surgical treatment is advised to protect the heart and large vessels from trauma on the one hand and to improve respiratory dynamics on the other. However, no therapeutic protocol is consensually established.

However, the studies are unanimous that the primary closure is the best during the neonatal period where we have the maximum elasticity of the rib cage with the least risk of compression of the underlying organs. It was performed for the first

time in 1949 by Maier and Bortone on a 6-week-old baby [1] [2] [3] [4] [5]. The cure will then be done by direct approximation of the banks by simple points after their avivement and a resection transforming the forms in U into V [1].

The imminent risk of compression of the underlying organs, against indicates direct closure. Several processes are then proposed. All have the same objective: gain in length and thus obtain coverage with the least risk of compression. These include lateral chondrotomies, oblique fracture of the clavicles, pectoral flap, periosteal flap of the sternal bars, bone grafts (iliac crest, tibial periosteum). They are preferable to synthetic prosthesis (titanium, silicone, etc.) [1] [3]. Eduardo Acastello *et al.* [2] suggested indications based on patient age. For children under one year of age a primary closure, a chondrotomy associated with a rupture of the bilateral sternoclavicular joint for children one to three years of age and flaps and/or autografting for children over three years of age.

Primary closures were reported in older patients, 1 years in the series of H Karakaya *et al.* [6] reported a successful primary closure in a 13-year-old patient. In our patient, the defect was suitable for a primary closure without tension, which is why we opted for the latter.

Gautam Sengupta *et al.* [7] used a thymectomy as we did. This allowed us to obtain more space and minimize the risk of compression. Turan *et al.* [8] and Madhok *et al.* [9] demonstrated no effect on immunity in children when partial thmectomy was performed.

In syndromic forms, the severity of heart defects that may be associated may lead to therapeutic abstention [10].

The evolution is favorable with a good aesthetic result good psychomotor development. The rare complications reported are a subcutaneous collection [1] [3].

4. Conclusion

Sternal cleft is a rare malformation of the rib cage. Although surgical repair in the neonatal period by primary closure is unanimously advised and accepted, the surgeon is often confronted with neglected forms. We admit that this primary closure can always be preferred when the residual compliance of the rib cage allows a closure without a pre-eminent risk of compression of the underlying organs, regardless of age.

Conflicts of Interest

The authors declare no conflicts of interest.

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