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A Bifid Sternum Case Underwent the Earliest Repair in the Literature

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Incomplete congenital sternal cleft is a rare anomaly. To avoid more complex reconstruction requirement in older children, primary closure of this defect during neonatal period should be recommended. A 5-day-old newborn with bifid sternum who underwent early primary repair was reported in this paper.

Key Words: Sternal cleft, Bifid sternum, Newborn, Primary repair

Literatürde En Erken Onarılmış Bir Bifid Sternum Olgusu

Inkomplet konjenital sternal kleft nadir bir anomalidir. Büyük çocuklarda daha kompleks rekonstrüksiyon ihtiyacından kaçınmak için, yenidoğan periodunda bu defektlerin primer olarak kapatılması önerilmektedir. Bu yazıda mümkün olan en kısa sürede primer onarılan bifid sternumlu beş günlük bir yenidoğan bildirilmiştir.

Anahtar Kelimeler: Sternal kleft, Bifid sternum, Yenidoğan, Primer onarım

Sternal defects constitute a spectrum of rare deformities of the sternum, heart, and upper abdominal wall. It is usually reported as an isolated case presentation. The first surgical correction was reported by Lannelongue in 1888,[1] but the first successful repair was published by Burton in 1947.[2] A considerable variety of procedures for sternal cleft repair have been reported in the literature [2-4] but it is an undisputed premise that sternal clefts should be corrected at the neonatal period within the first month of life. At this stage, the sternal bars can be easily approximated by simple sutures, owing to the chest's flexibility. This also permits approximation of the sternal bars with less danger of cardiac compression.[3,5]

This anomaly appears to represent an isolated developmental field defect with probable multifactorial etiology. Familial occurrence of sternal fusion defect has not been reported. It can be complete (the rarest form) and incomplete (the upper cleft sternum or bifid sternum). The bifid sternum is usually isolated, with orthotopic normal heart and normal skin coverage. It is generally observed at birth and is asymptomatic. In those rare symptomatic cases, cyanosis, dyspnea and pulmonary infections are the main clinical features.[6]

In this paper we present a newborn with bifid sternum who underwent early successful primary closure.

CASE REPORT

A newborn with birth weight 2600 g, admitted to the Department of Pediatric Surgery, with a midline thoracic bulge, evident during expiration and crying. During inspiration, a depression appeared in the same area. On admission the neonate was asymptomatic. Observation and palpation allowed us to diagnose a 4.5-cm-wide U-shaped, sternal cleft (Figure 1). The diagnostic image was consistent with the clinical evaluation: a chest roentgenogram showed the typically widely separated clavicles and absence of manubrium and of ossification centers of the sternum. The electrocardiogram and an echocardiogram of the infant were normal.

At the age of 5 days, primary closure of the defect was performed. Through a midline cevicothoracic incision, the sternal bars were dissected free from the insertion of pectoralis major muscles anteriorly, and from endothoracic fascia. The resection of inferior sternal portion is done to get "V" transformation of the "U" defect. The pectoral

muscles joined in the midline, and the edges of the bars were then sharply freshened, and multiple encircling nonabsorbable sutures were passed through four intercostal space. A penrose dainage was left under the skin flaps and the skin closure was performed with absorbable subcuticular sutures. No per-operative complication occurred. The patient started feeding at 8 hours postoperatively. The antibiotic was stopped after taking out the drain. Patient was discharged 5 days after repair. Now he is 18 month- old and in good condition (Figure 2).

Figure 1.

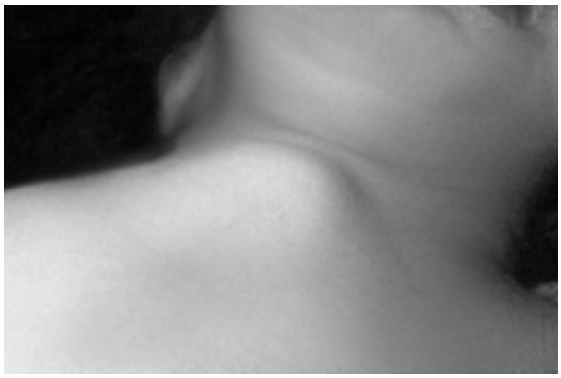
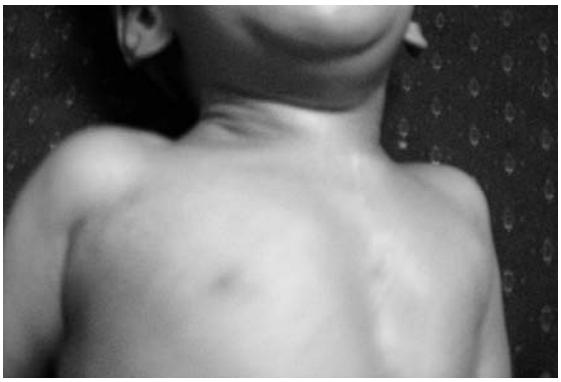


Figure 2.



DISCUSSION

Congenital deformities of the sternum are best considered as four entities with limited overlap among categories that can best be based on the tissue coverage of the heart. The sternum is derived from paired concentrations of thoracic lateral bands that fuse in a craniocaudal direction by the ninth week of gestation. The superior part of the manubrium arises from three small mesenchymal primordiums, while the sternal bands fuse laterally with the costal cartilage. Disturbances of normal ventral midline thoracic fusion can present as a spectrum of abnormalities, including a prominent suprasternal notch, irregularities in shape of the xiphoid, ectopia cordis, superior sternal cleft, or complete sternal cleft.[1,6,7]

Incomplete sternal clefts, usually involving superior segments, are associated with abnormalities of midline fusion, such as ventral hernias, congenital heart defects, and deficiencies of the diaphragmatic pericardium.[6,7] Hemangiomas and midline abdominal raphea are also seen.[8]

The indications for repair of a congenital bifid sternum are to restore bony protection to the mediastinal structures, to provide normal intrathoracic pressure relationships and to eliminate paradoxical motion of the thoracic viscera and the large, visible contour deformity.[7]

Several methods of correction have been described. These include oblique division of the superior cartilage, chondrotomies, insertion of cartilaginous autografts, and utilization of wire mesh or Teflon.[3,8-11] There is a general agreement that the best treatment is primary closure as a neonate. As the infant grows, the cartilage becomes less malleable and the thoracic contents move anteriorly into the defect.[12] A V-shaped defect in the sternum can often be approximated by suture closure with relaxing osteotomy of the edges if needed. The U-shaped defects often require transection of the limbs of the U at the juncture with the normal sternum, followed by suture approximation.[6,12,13] Nevertheless, patients who have not undergone repair during the neonatal period may require reconstruction later on. The increasing rigidity of the chest wall and the physiologic accommodation of the thoracic organs to a certain circumference of the chest prevent simple approximation without seriously compromising the heart and lungs.[14] These features distinguish older patients from newborns. One of the earliest successful procedures for correction in older children was devised by Sabiston in 1958.[3] By using multiple bilateral sliding chondrotomies he was able to bring the sternal halves together while simultaneously increasing chest wall dimensions and flexibility. After that, miscellaneous techniques were developed, e.g., split ribs, costal cartilage grafts, and segments of costal margin; stainless steel wire mesh; segments of cartilage divided laterally and swung medially to cover the defect; autogenous periosteal graft raised from the tibia in the lower limb; and perichondral flap elevation and pectoralis muscle transpotion. In previous reports of repair in older patients, the use of

prosthetic materials such as Marlex mesh, Teflon, silicone elastomer, or perforated acrylic plate have also been adapted by different authors.[8-11]

According to literature and in our opinion, it is always better to avoid the use of prosthetic materials, considering the risks of infection and the inability of these inert materials to grow with the patient.[5,7,15]

To the best of our knowledge, this case is the smallest neonate underwent primary closure in the literature. As a conclusion, since the thorax is highly flexible shortly after birth, we recommend, repair of bifid sternum early in life when primary closure is generally safe and relatively easy.

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