

Complete sternal cleft treatment in a low birth weight patient

Düşük doğum ağırlıklı hastada komplet sternal kleft tedavisi

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ABSTRACT

A complete sternal cleft is a very rare congenital anomaly causing severe respiratory compromise. Surgical reconstruction options are limited, particularly in low birth weight newborns. Herein, we report a case of low birth weight premature newborn with a complete sternal cleft and its surgical treatment.

Keywords: Congenital heart defect, pectoralis muscles, premature birth, sternum.

ÖZ

Komplet sternal kleft, ciddi solunum sıkıntısına neden olan, çok nadir görülen bir doğumsal anomalidir. Cerrahi rekonstrüksiyon seçenekleri özellikle düşük doğum ağırlıklı yenidoğanlarda sınırlıdır. Bu yazıda, komplet sternal kleft olan düşük doğum ağırlıklı prematür bir yenidoğan olgusu ve cerrahi tedavisi sunuldu.

Anahtar sözcükler: Doğumsal kalp defekti, pektoral kaslar, prematür doğum, sternum.

Sternal clefts are malformations which occurs as a fusion failure of sternal elements with an incidence of 0.15% among all chest wall deformities.^[1] It can be classified in two types as complete or partial.^[2] Complete sternal clefts can be a part of Cantrell syndrome and can be life-threatening due to intracardiac anomalies and respiratory compromise. Herein, we describe the successful surgical repair of a newborn case with absent sternum with prolene mesh and pectoralis major muscle flaps.

CASE REPORT

A two-day-old newborn baby with total sternal cleft was transferred to our neonatal intensive care unit (ICU). The patient was born at the 34th gestational week with a weight of 1,400 g. She was a twin sister. There was no family history of congenital anomalies. She had respiratory distress syndrome of newborn and was intubated. She had also nutritional

intolerance. A midline sternum and skin defect were noted on the anterior chest wall (Figure 1). There was a membranous skin covering the sternal defect and cardiac structures were almost visible through it. After further examination and tests, she was diagnosed with esophageal atresia and abdominal ultrasonography showed hydronephrosis in the right kidney and bilateral urolithiasis. Echocardiography examination revealed no intracardiac anomaly. Computed tomography (CT) showed an absent sternum and bilateral lung atelectasis (Figure 2). When the patient was five days old, she underwent surgical repair of esophageal atresia via a right posterolateral thoracotomy. At 24 days of age, she was extubated with the aid of non-invasive ventilatory support. The plastic surgery team consulted the patient and we decided to wait for surgical reconstruction, until she reaches a body weight of 3,000 g. The patient was taken care of in the neonatal ICU until surgery. During neonatal ICU stay, she fed orally, and the course was uneventful with respiratory support. When the patient

Received: April 10, 2020 Accepted: May 18, 2020 Published online: October 21, 2020

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Cite this article as:

Temur B, Mete S, Beken S, Önalın MA, Ereğ E. Complete sternal cleft treatment in a low birth weight patient. Turk Gogus Kalp Dama 2020;28(4):684-687

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Figure 1. A preoperative image of sternal cleft.

was five months old and 3,085 g, the operation was performed. A written informed consent was obtained from each parent.

During surgery, a midline skin incision was done carefully and the subcutaneous tissue was passed through. The pericardium was found deficient inferiorly, and a part of the heart was lying directly under the membranous skin. Primary closure with direct approximation of the costal cartilages was not feasible, due to the risk of mediastinal compression. The defect was large, and the chest cavity was small. The pericardium was closed primarily with prolene

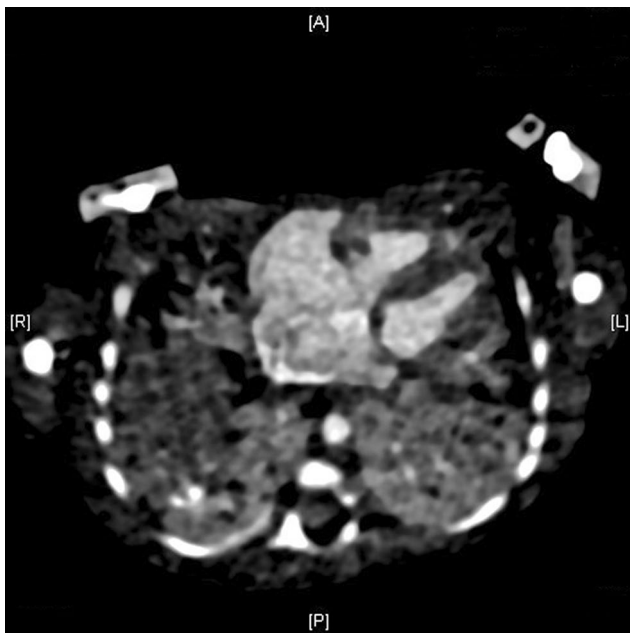


Figure 2. A computed tomography showing bulging of mediastinal viscera and a complete sternal cleft.

sutures. Both pectoralis major muscles and fascia were released. A prolene mesh (Prolene™, Ethicon Inc., NJ, USA) was measured and placed above the pericardium and attached to the ribs with prolene sutures (Figure 3). The pectoral muscle flaps were advanced to the midline over the mesh and sutured to each other. The fasciocutaneous flaps were advanced over the muscles flap and sutured in midline. A Hemovac® (Hemovac®, Zimmer Biomet, USA) drain was placed in the mediastinum. Primary skin closure was done. There was no complication after surgery and the wound healed without any problem. The patient was extubated postoperatively on Day 2 and discharged on Day 21. Six months after surgery, her chest wall was stable and she was growing up without any respiratory problems (Figure 4).

DISCUSSION

Sternal cleft anomaly results from failure of development and fusion of two lateral mesodermal lateral plates by the eight weeks of gestation.^[2,3] Antenatal diagnosis is possible. Complete clefts are uncommon. Ramirez-Solis *et al.*^[4] reported that, in mice, alcohol intake, disruption of the Hoxb4 gene and methylcobalamine deficiency were associated with sternal defects. It may be related to omphalocele



Figure 3. An intraoperative image of a complete sternal cleft. A prolene mesh was measured and placed above pericardium and attached to ribs with prolene sutures.



Figure 4. A postoperative image of sternum at six months.

and craniofacial hemangiomas. Our case had associated esophageal atresia and hydronephrosis. Sternal cleft, supraumbilical abdominal anterior wall defects, anterior diaphragm defect, pericardial defects, and intracardiac anomalies constitute the Cantrell pentalogy. Also, sternal clefts can be a part of posterior fossa brain malformations, hemangioma, arterial lesions, cardiac abnormalities, and eye abnormalities (PHACE) syndrome or Poland's syndrome consisting of hand anomalies, nipple anomalies, dextrocardia, renal agenesis, and various tumors.^[5,6] Physical examination, echocardiography, and CT are helpful to define the anomalies and decide the surgical repair strategy. Newborn babies with a complete sternal cleft need to be hospitalized, since the membranous skin covering the defect leaves the heart unprotected and there is a risk for respiratory compromise.

The methods for repair of complete sternal defects are primary closure, sliding, or rotating chondrotomies, closure with prosthetic grafts, flaps of bone, cartilage, autogenous tissue, or muscles.^[5,7,8] In our case, prolene mesh and pectoral muscle flaps provided good and solid coverage of the defect. This technique enabled to obtain enough mediastinal space and prevent cardiac compression. Resection of costal cartilages and disruption of sternoclavicular junction for mobilization of the sternal bars may be performed in older age.^[3] Padolina et al.^[9] reported incomplete cleft repair

in two patients with the modified Sabiston's sliding chondrotomies technique by which chondrotomies were performed after dissecting the clavicles and costal periosteum free. As the cartilage tissue is more elastic and expandable in early infancy, it can be easier to close the sternum.^[9] Synthetic patches such as polypropylene, polyester, and polytetrafluoroethylene (PTFE) patches can be sutured to the ribs and used to cover the defect in patients with congenital anomalies as well as in patients with trauma and other destructing bone diseases.^[10] We used the polypropylene mesh above the pericardium as a barrier. The infection rate is estimated as 10 to 25% for synthetic meshes and removing the mesh may be needed in some of the cases.^[10,11] Biological materials such as bovine or porcine pericardial patches can be also used to prevent infection; however, they have a risk of calcification.^[9] In our case, the defect was large and the cleft was total; therefore, we decided to use a more firm and strong material (i.e., a polypropylene mesh instead of a pericardial patch). The use of muscle flaps for chest wall and sternum reconstruction is a common method.^[8,12] Although latissimus dorsi and rectus abdominis flaps, even the omentum flaps, are good options for reconstruction, pectoralis major seems to be the principal flap.^[12,13]

In conclusion, sternal reconstruction of a complete sternal cleft with pectoralis major muscle flaps and prolene mesh is a feasible, safe, and effective procedure in newborns with favorable outcomes. We believe that the timing of the intervention in newborns depends on the clinical status and body weight of the patient, and presence of intracardiac anomalies. Waiting for reconstruction until the patient reaches a body weight of 3,000 g is recommended in premature babies.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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