Complete Sternal Cleft In Adolescence

H Karakaya, N ?ahin, A Arslan, A Trakya

Citation

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Abstract

Introduction: We report a complete sternal cleft associated with pectus excavatum who underwent successsful surgical repair of the sternal anomaly by direct approximation of the freshened sternal remnants to the midline. Case Report: A 13-year-old girl presented with a complete sternal cleft since birth, causing a cosmetic concern and constant fear of trauma. Physical examination was measured 4 cm wide at proximal and 1 cm wide at distal sternum, and pectus excavatum. Anaesthesia was induced with fentanyl 2 microg/kg, thiopenthone 6mg/kg and vecuronium 0,1 mg/kg and maintained with 50 %O₂IN₂O and sevoflurane 1,5-2.0 %. Heart rate, ECG, arterial blood pressure, central venous pressure, SaO₂ and arterial blood gases were monitored. After an uncomplicated postoperative period, the patient was discharged. Conclusion: Our effort was to preserve the normal anatomic and mechanical function of the thoracic cage as well as to achieve proper closure of the defect while protecting the heart and great vessels without using any synthetic material, with good cosmetic results.

INTRODUCTION

Isolated complete congenital sternal fissure or cleft (absent sternum) in adolescence is very rare anomaly, and very few cases have been reported so far ($_1$). Although the embryology of the thoracic wall is well known, the etiology of cleft sternum remains unclear. Failure of fusion of the lateral sternal bands by an early disturbance affecting the midline mesodermal structures between the sixth and ninth gestational week is a possible mechanism but no familial, teratogenic or nutritional factors have been identified as a potential cause ($_{2,3}$).

Besides the isolated forms with good prognosis, cleft sternum can present in association with other life-threatening disorders as ectopia cordis or as a part of Cantreell's pentalogy, which comprises a lower sternal defect, a deficiency of the anterior diaphragm and pericardium, a midline supraumblical abdominal wall defect and an intracardiyac disorders ($_{4,5,6}$).

We report successful repair of the complete sternal cleft and anterior pericardial defect associated with pectus excavatum.

CASE REPORT

A 13-year-old girl presented with a complete sternal cleft associated with pectus excavatum since birth, causing a cosmetic concern and constant fear of trauma. Physical examination was measured 4 cm wide at proximal and 1 cm wide at distal sternum, and pectus excavatum. Pulsations of the heart and great vessels could be easily seen through the defect, which was covered by a thin layer of skin. During valsalva maneuver the sternal cleft became wider and the heart bulge became dramatically more prominent. No other abnormalities were present, and the results of chest roentgenogram, echocardiography and spiral computed tomographic scan comfirmed the findings of the physical examinations (fig 1-2).

Figure 1

Figure 1-2: Spiral computed tomographic scan of the chest with three-dimensional reconstruction demonstrating the complete sternal cleft.



Figure 2



For premedication, midozolam (0,05mg/kg) was given intravenously before the patient was taken to the operating room. Peripheral venous and radial arterial cannulations were performed under local anaesthesia before induction. Anaesthesia was induced with fentanyl.

2 microg/kg, thiopenthone 6mg/kg and vecuronium 0,1 mg/kg and maintained with 50% O_2 , N_2O and Sevoflurane 1,5-2.0 %. Heart rate, ECG, arterial blood pressure, central venous pressure, SaO₂, arterial blood gases and urine output were monitored continuously.

The skin overlying the sternal defect was incised in the midline. The pericardium was found to be absent anteriorly

along the length of the defect. The heart and great vessels were found to be normal. Refashioned pericardium was directly closed in the midline.

The pectoralis muscles were only incised over the place of the cartilage excision. The fragments of the abnormal 5 cartilages were excised subperichondrially and bilaterally. After pectus excavatum deformity was corrected, the periosteum of each bar was incised on its lateral border. The two sternal bar were then approximated with five interrupted steel wires. The closure was completed with the two major pectoralis muscles reapproximated for the midline.

Because of the extensive resection of deformed cartilage, the two sternal bars could be easily approximated without interposition of costal cartilage. Finally the internal or external fixation of the sternum prevents parodoxical respiration. Closed suction fasciocutaneous flaps were advanced to achieve primary skin closure.

After the operation, the patient admitted to the ICU and extubated third hour. After an uncomplicated postoperative period expect for tachycardia at first hour. The patient was discharge from the hospital after 10 day chest roentgenogram and computed tomographic scanning done on the 6^{th} month postoperatively showed a healthy closure of the defect.

DISCUSSION

Cleft sternum is usually not associated with cardiac defects, is usually not fatal, and comprises a rare spectrum of ventral fusion abnormalities, which may be classified as complete or incomplete. Complete clefts sternum is the least likely form ($_7$, $_8$). Its clinical significance is that it leaves the heart and great vessels unprotected ($_9$).

Sternal cleft association with pectus excavatum is rare anomaly. The currently accepted cause of pectus excavatum that misdirected growth of the lower costal cartilages, which form congenitally in a concave manner because of rapid growth and create a depressed sternum ($_{10}$). When these anomalies are occurred at the same time that surgical treatment doesn't make any difficulties. Numerous methods of surgical correction of the sternal cleft have been reported, including direct approximation of the sternal bands, sliding or rotating chondrotomies, interposition of bone, cartilage, or prosthetic grafts, and closure by pectoralis muscle flap advancement ($_{11,12, 13, 14}$). However the determining factor in justifying the appropriate technique is the age of the patient. Presently, general consensus advocates early correction during the neonatal period, because the bony structures of the thorax are still compliant and permit direct sternal closure (11, 15).

Whether dealing with older children or young adults, the rigidity of the chest wall and the lack of space available to the intrathoracic organs must be considered, especially when the defect is too large to be closed directly. The heart is least tolerant organ to reduced space inside the thoracic cage (1, 8, 10).

In our case, because of the extensive resection of deformed cartilage, the two sternal bars could be easily approximated. We didn't need to get the other methods.

According to the literature, 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 (8).

Because our patient was 13 years girl, our effort was to preserve the normal anatomic and mechanical function of the thoracic cage as well as to achieve proper closure of the defect while protecting the heart and great vessels without using any synthetic material, with a good cosmetic results.

CORRESPONDENCE TO

DR. NURSEL SAHIN Akdeniz Üniversitesi Tip Fakültesi Anesteziyoloji ve Reanimasyon AD. Dumlupinar Kampüsü ANTALYA- TURKEY Telephone: + 90 242 227 43 43 (55 318) Fax : + 90 242 247 00 45 E-Mail Address: sahinnursel@hotmail.com

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Author Information

Hanife Karakaya, MD

Department of Anesthesiology and Reanimation, Akdeniz University School of Medicine

Nursel ?ahin, MD

Assistant. Professor, Department of Anesthesiology and Reanimation, Akdeniz University School of Medicine

Asuman Arslan, MD

Department of Anesthesiology and Reanimation, Akdeniz University School of Medicine

Ayten Trakya, MD

Professor, Department of Anesthesiology and Reanimation, Akdeniz University School of Medicine