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*Case Reports*

# Combined Repair of Upper Sternal Cleft and Pectus Excavatum in a Child

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## Abstract

Sternal cleft accompanied by pectus excavatum is a rare type of congenital anomaly of the chest wall. Surgical correction is a suitable approach to restore the heart, large vessels, and respiratory dynamics early. This is a report of the successful surgical correction of upper sternal cleft anomaly accompanied by pectus excavatum in a child. The pectus excavatum was corrected without the use of any prosthesis. The cleft was closed by primary approximation with enough dissected pectoralis major muscle and partial thymectomy, mobility, and flexibility ensured by pectus correction. The integrity of the sternum and the chest wall was normal at the end of the 12-month follow-up period.

**Keywords:** Chest wall deformity; sternal cleft; funnel chest; surgical correction

## Introduction

Sternal cleft (SC) is a rare congenital chest wall malformation that results from defective embryologic sternal fusion of mesenchymal cells in the ventral midline.[1] Recognition of a partial SC may be difficult if there is no apparent associated anomaly. Surgical correction is ra suitable approach to restore the heart, large vessels, and respiratory dynamics early in the course of disease.

Coexistence of SC and pectus excavatum (PE) is very rare. Therefore, available information and results related to the simultaneous correction of these 2 anomalies by simultaneous surgery are limited.

The case presented in this article is an 8-year-old girl with a superior U-shaped SC associated with PE which was successfully corrected using the Ravitch procedure without a metal bar and direct approximation of the sternal bars.

## Case Report

In December 2019, an 8-year-old girl was brought to the Avicenna Hospital thoracic surgery clinic, reporting a deformity in the anterior chest, heart palpitations, and the feeling of her heartbeat in the upper anterior chest. During the physical examination, together with PE pulsation of heartbeats, approximately 4 cm of cleft in the upper part of the sternum was detected. In addition, there were band-shaped scars extending from the lower end of the sternal defect to the umbilicus (Fig. 1). An asymmetrical PE and thymus gland in the anterosuperior mediastinum extending under the skin were present on computed tomography (CT) scan (Fig. 2). The distance between the sternal bars was measured on chest CT to be 5.31 cm.

Thoracic tomography confirmed the preliminary findings. Intrathoracic organs were reported as normal. Cranial magnetic resonance imaging, abdominal ultrasound, and echocardiography findings were within normal limits.

A written informed consent was obtained from the legal guardians of the patient. A vertical midline incision along the defect was made. A U-shaped superior SC was clearly seen.

The sternoclavicular joint was separated on each side to mobilize the sternal bars. Costal cartilage of the first and second ribs was not divided because adequate mobilization was provided. This procedure was possible because the patient was 8 years old and the thorax was flexible. Bilateral sternal bars were released from the underlying tis-

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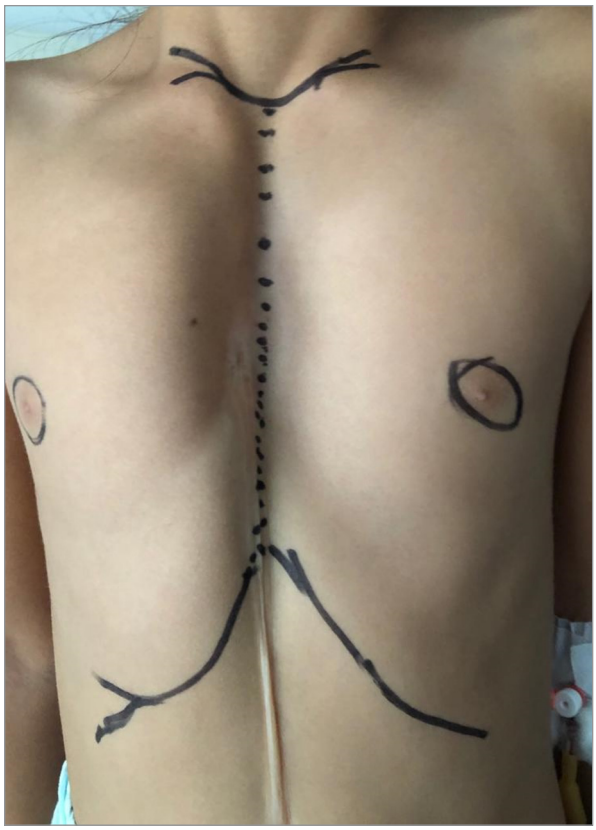

**Fig. 1** Patient before surgery, age 8 years.

sues, and the intact pericardium was freed from each sternoclavicular joint. The fused remaining portion of the sternum was separated via a midline osteotomy. A partial thymectomy was performed to create more space for primary closure. An extensive dissection and liberation of the pectoralis major muscle was performed, and then the third, fourth, fifth, and sixth costal cartilages on both sides of the thorax were resected. After, 2 sternal bars were easily approximated with 2 steel wires (manubrium and corpus) plus 4 nonabsorbable sutures at the level of the manubrium, corpus, and xiphoid process without interposing the costal cartilage (Fig. 3). The detached pectoralis major muscle was sutured along the midline. The xiphoid was sutured to the rectus abdominis muscle, and no prosthetic material was needed to support the sternum. Closed suction drains were placed in the mediastinum and above the pectoralis major muscle flaps. The wound was then closed in double layers using absorbable sutures. The patient had an uncomplicated postoperative recovery and was discharged to home 5 days after the surgery.

One year later, the patient remained asymptomatic with good cosmetic results (Fig. 4A). Chest radiography and CT scans showed a healthy closure of the cleft (Fig. 4B).

### Abbreviations and Acronyms

| | |
|---|---|
| CT | computed tomography |
| PE | pectus excavatum |
| SC | sternal cleft |

## Discussion

Depending on the degree of the separation, SC is classified as complete or incomplete. Incomplete defects are subdivided into superior and inferior, with the most common form being the superior SC, as seen with the patient in this case study.[2]

This rare entity may be associated with other congenital anomalies such as cardiac anomalies, PE, midline defect anomalies including connective tissue nevi,

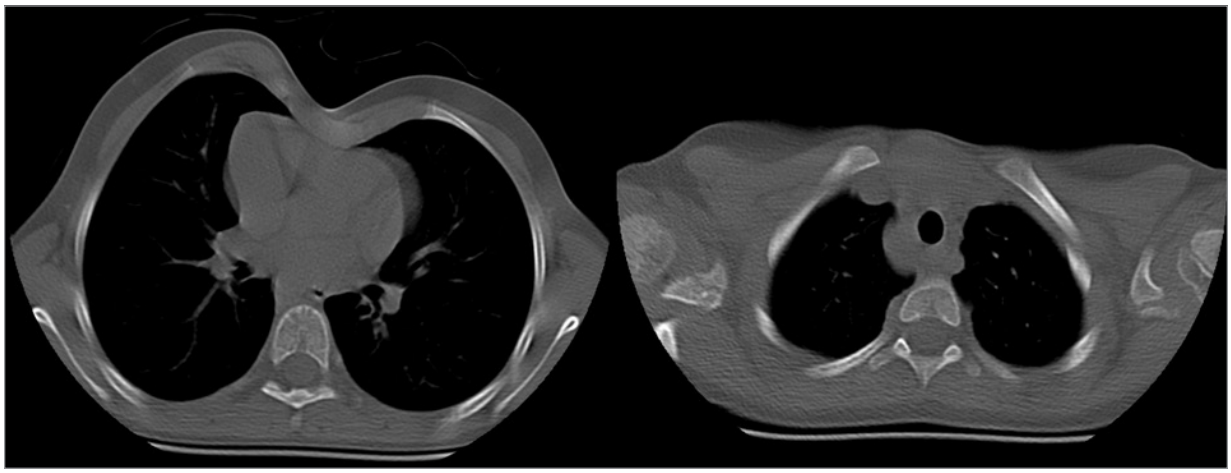

**Fig. 2** Preoperative chest computed tomography images demonstrate severe asymmetric pectus excavatum with significant sternal rotation and cleft. In addition, the thymus gland is seen in the anterior part of the mediastinum.

PHACES (Posterior fossa; Hemangiomas of the face, neck, and/or scalp; anatomical anomalies of the cerebral or cervical Arteries; Cardiac anomalies/Coarctation of the aorta; Eye abnormalities; and Sternal anomalies) syndrome, or pentalogy of Cantrell—which may or may not be compatible with life.[3]

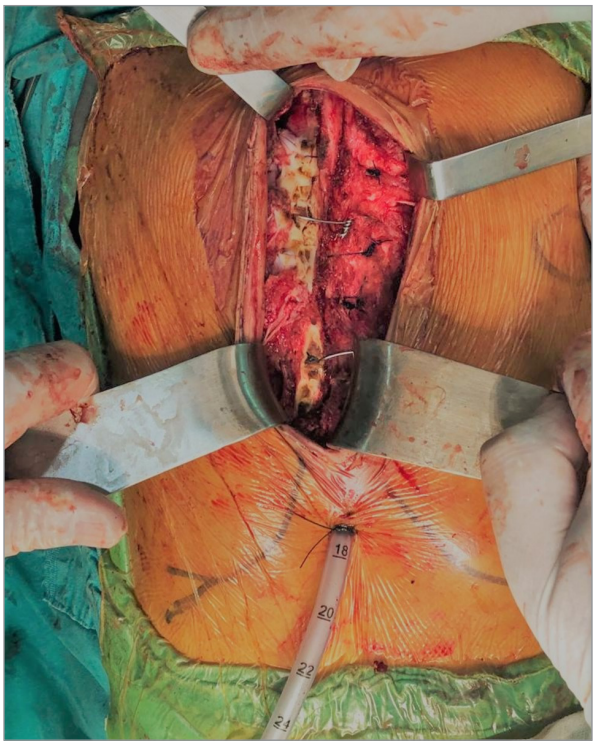


**Fig. 3** Two sternal bars were easily approximated with 2 steel wires plus 4 nonabsorbable sutures at the level of the manubrium, corpus, and xiphoid process.

Pectus excavatum is a deformity of the sternum and costal cartilage with depression. The cause of PE may stem from unbalanced overgrowth in the costochondral regions. Patients may feel palpitations and chest pain during exercise and almost always report anxiety and body perception disorder.[4] Pectus excavatum can be symmetrical or asymmetrical and associated with scoliosis and connective tissue disorders. The PE in this case was type 4 according to the Willital classification, so it was asymmetrical PE.[5] The surgical repair of PE in childhood has been a well-established procedure with open repair and minimally invasive repair.

Pectus excavatum and SC are mostly seen alone; their coexistence is extremely rare. In a literature review by Torre et al[2] on SC cases from 2010 onwards, it was found that only 8 patients (10%) were also affected with PE (diagnosed preoperatively or at follow-up); in 2 of them, PE and SC were treated simultaneously, whereas in the other 6, PE could not be repaired.

Surgery should maintain the growth of the chest wall, protect the mediastinal structures, and improve cosmetic appearance. Several surgical techniques have been described in the literature for simultaneous correction of the 2 anomalies, including the Ravitch technique with or without a steel bar, the Nuss procedure, primary approximation, sliding chondroplasty, periosteal and chondral flaps, prosthetic repair, use of a muscle flap, and autologous graft.[6]

Sarper et al[7] reported the case of a 13-year-old girl with complete SC accompanied by PE. The pectus deformity was corrected using the Ravitch technique

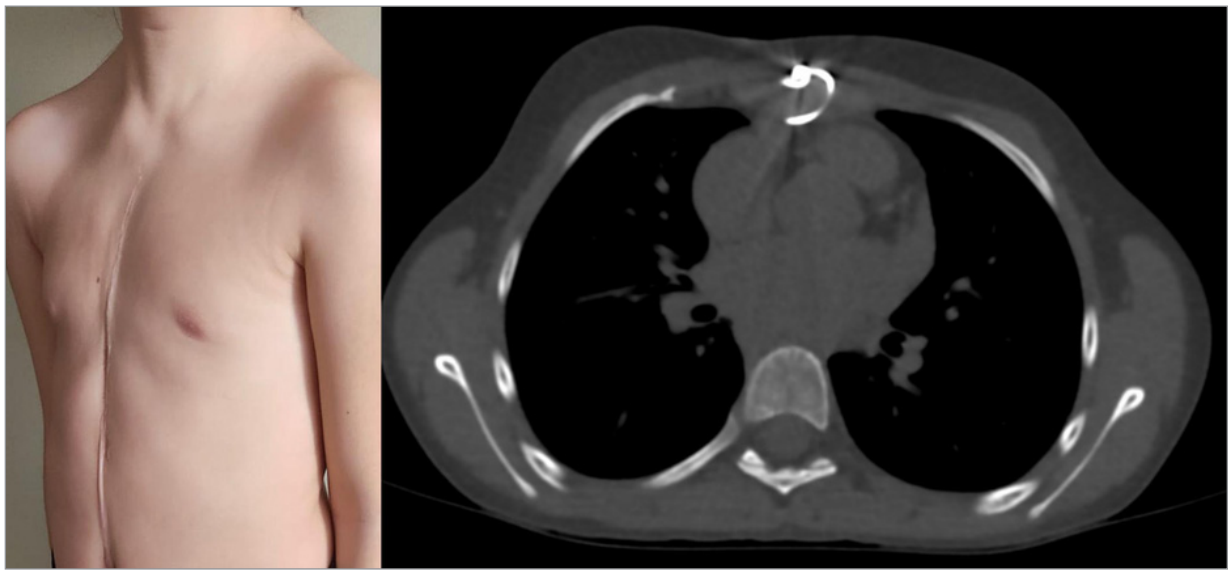

**Fig. 4** **A)** Patient 1 year after surgery. **B)** Postoperative computed tomography scan shows 12 months following surgery.

without a steel bar, and the SC was repaired by periosteal flap. It was not necessary to operate on the sternal flap because of sufficient mobility of the thorax and incomplete SC.

In the British literature, the number of reported SC cases accompanied by PE was 5 since 2010. Three of them had simultaneous surgical correction.[8-10]

Kabiri et al[8] operated on a 25-year-old woman with direct sternal approximation using the Ravitch procedure and steel wires, but no metal bars were used.

Tocchioni et al[9] report a case in which they sutured the SC of an 18-year-old woman, along with release of the pectoralis major muscle and using a periosteal flap, Prolene mesh, and autologous cartilage graft; they successfully used the Nuss procedure for the PE.

Smith et al[10] reported on the 13-year-old girl for whom they successfully applied the Ravitch procedure, Nuss bar, and direct closure with steel wires for the sternum simultaneously.

Like with the choice of surgical method, timing is also important. The ideal correction age for the SC is in the neonatal period.[11] However, in the case of PE, the ideal correction age is generally accepted as before puberty.[12]

Considering the size of the cleft, and the condition and development of the pectus deformity, a simultaneous surgical correction can be carried out before puberty. However, severity of the cleft and other accompanying non-PE anomalies should also be taken into account in the decision-making process to successfully perform an operation on the 2 deformities.

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