

A case of a rare thoracic
malformation with
paradoxical chest wall
movement

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Sternal cleft (also known as bifid sternum or sternal fissure) is a rare thoracic malformation. Both partial and complete forms have been described, and it may occur as an isolated finding or in association with other congenital anomalies, mostly defects of the heart or the gastrointestinal tract.

We report on a term newborn with an isolated upper sternal cleft and describe his surgical therapy and follow-up.

After an uneventful pregnancy, this male baby was born vaginally after 38 weeks of gestation (birth weight 3190 g) to a healthy G2/P1 mother. Apgar scores were 10, 10, 10 at 1, 5, 10 minutes, respectively, and umbilical artery pH was 7.16.

After birth, he presented with a V-shaped, concave sunken area in the frontal midline of the chestwall, about 6 x 8 cm of size, that moved paradoxically during respiration and crying (Fig. 1 and movie). In addition, heart pulsations could be seen directly underneath the skin.

Upon palpation, an almost complete upper sternal cleft was diagnosed, with a small bridge at the xyphoid process. Pulsations of the heart and major vessels could be palpated just subcutaneously. There were no signs of

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respiratory distress and no heart murmur. A small skin-fold cranial to the umbilical cord was the only additional anomaly (Fig. 2). No further anomalies were detected on physical examination, audiometry, echocardiography, abdominal and cerebral ultrasounds. Chest x-ray (Fig. 3) showed an increased distance between the sternal ends of the clavicles. After 2 days, the baby left the NICU without further problems.

At 2 months of age, the cleft was surgically closed. After drawing the landmarks of the cleft onto the skin (Fig. 4), a median incision was made over the sternal defect, and the subcutaneous tissues were divided. A 5-6 cm long V-shaped defect of the sternum was found cranially and the sternal halves were joined caudally by a cartilaginous bridge. The pectoral muscles were notably hypoplastic. They were dissected off the sternal parts on each side as well as the mediastinum and pleura. Mersilene sutures were used to approximate the two sternal halves and complete closure could be achieved without signs of cardiac or respiratory compromise. The mobilized pectoral muscles were then sutured together over the closed sternum and the skin was closed with an absorbable subcuticular running suture. The midline supraumbilical raphe was not disturbing for the parents and it was decided not to repair it at this point. The post-operative course was uneventful, and the boy was discharged 6 days after surgery. When seen 1 (Fig. 5), 2 and 6 months after surgery, he did well both in



Fig. 1

V-shaped sternal defect which moves paradoxically during crying (A: retracting during inspiration, B: bulging during expiration); note: movie can be accessed at www.neonet.ch/COTM-2008-05.



Fig. 2

Small skinfold cranial to the umbilical cord.



Fig. 3

Chest X-ray showing an increased distance between the sternal ends of the clavicles.



Fig. 4

Landmarks of the cleft prior to surgical closure.

Clinical appearance one month after surgery.

Fig. 5



functional and cosmetic terms. The only remarkable observation was a soft tissue bulge with crying or coughing at the jugular space in the midline.

Sternal cleft is a very rare congenital malformation with unknown incidence. The first case was described by Torres in 1740 (1). At 6 weeks' gestation the sternum appears as two parallel separated mesenchymal bands. Starting at 8 weeks, these bands fuse craniocaudally to form the body and inferior parts of the manubrium until 10 weeks of gestation. Most isolated sternal defects result from a failure of the lateral mesenchymal bands to fuse at approximately eight weeks' gestation. The etiology of sternal cleft deformity is unknown. Hypotheses include chronic nutritional deficiency, deficiency of riboflavin during pregnancy, disruption of the HoxB4 gene (2), persistence and proliferation of midline angioblastic tissue (3), and rupture of the yolk sac (4), with early rupture (< 8 weeks of gestation) interfering with cardiac descent, internal cardiac development, and midline fusion of thoracic structures, and late rupture resulting in a cleft sternum only.

Sternal clefts occur sporadically, although one autosomal recessive familial association was reported in 1984 (5). Prenatal diagnosis is challenging and reports are sparse (6, 7, 8). At birth, the defect may be V-shaped, when the cleft reaches the xiphoid process, or broad and U-shaped, with a bony bridge joining the two

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edges, ending at the third or fourth costal cartilage. While most infants are asymptomatic at birth, major defects destabilize the chest wall, and the changes in intrathoracic pressure resulting from paradoxical chestwall movement may rarely cause displacement of the heart and large vessels and impair hemodynamics. As a result, there may be right ventricular overload, cyanosis, dyspnea, and arrhythmias (9, 10).

According to the degree of fusion, sternal clefts can be classified as complete or partial, of which the latter can be located superiorly, inferiorly or in the middle (sternal foramen) (11). The complete and the inferior type are more frequently associated with other anomalies than the superior type. A third of patients with sternal clefts also have facial hemangiomas and a supraumbilical raphe (12). Further associated malformations are omphalocele, ectopia cordis, pericardial defect, and congenital heart defects. Combinations of these anomalies together are known as Cantrell's pentalogy (13).

There is general agreement that the condition requires surgical correction even in asymptomatic patients to improve respiratory function, prevent paradoxical motion of the mediastinal viscera with respiration and provide mechanical protection from penetrating trauma.

Surgical repair of a congenital sternal cleft was first

attempted in 1888 (14). The first successful repair was published in 1947 (15). Since then several techniques have been described (16, 17, 18, 19).

The ideal timing and technique of the operation is still controversial (20), ranging from 2 days (21) to 3 months of age (22). Later on, the chest wall is less compliant, making the surgical procedure more demanding. Primary approximation can be achieved without cardiorespiratory compromise in most cases operated on early in infancy. It can be reinforced with pectoralis major flaps to protect the primary repair and reduce the lateral distraction forces exerted by anomalous pectoralis insertion (23). Closure with bilateral pectoralis muscle flaps alone has also been reported (21). Repair using autologous tissue is preferred to prosthetic materials, as these are associated with tissue reactions and increased risk of infection. Hazari et al. (17) described reinforcement of the primary sternal approximation with a titanium plate at 11 weeks of age in a baby with associated cardiac abnormalities that had been surgically corrected at 5 weeks of age. At the time of the second cardiac operation at one year of age, the titanium plate was removed and the sternal cartilage was found to be well formed with areas of ossification. In 2004, Abel et al. reported the operative repair in a 9-year-old girl by a free graft from the iliac crest (22). This child was not operated on earlier, as the condition was believed to be inoperable.

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