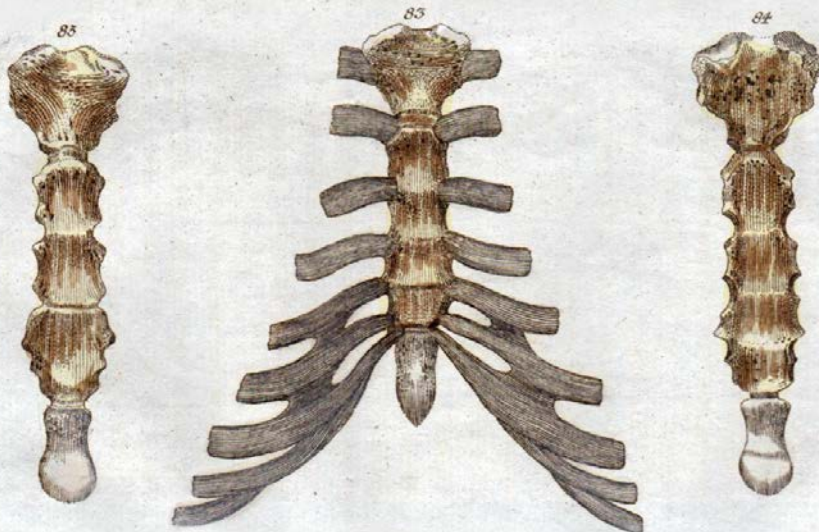


Sternal cleft – a rare
congenital malformation

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Title figure:

Sternum: manubrium, body and xyphoid process

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Sternal cleft is a rare congenital chest wall defect resulting from a partial or total failure of sternal fusion at an early stage of embryological development. Its exact incidence is unknown. Only a few cases have been described in the literature. Many different etiological factors have been suggested but none of them has been clearly proven.

The defect is often asymptomatic, although in most cases, the anomaly is associated with a wide spectrum of other midline defects. Diagnosis can be made clinically after birth by inspection and palpation. Prenatal diagnosis is difficult. The malformation can expose mediastinal viscera and vessels to life-threatening injuries. Depending on the size of the defect, sternal clefts may be treated or not. The optimal choice of treatment is surgical repair in early life to achieve primary closure.

We will present the case of a near term baby with a partial superior form of sternal cleft without any other associated anomalies.

CASE REPORT

This female infant was born to a healthy 38-year-old G1/P1 at 35 5/7 weeks by normal vaginal delivery following premature rupture of membranes. The pregnancy had been unremarkable, and screening ultrasound examination at 22 weeks of gestation did not show any fetal abnormalities. The premature baby's birth weight was 2445 g (P30). Apgar scores were 8, 8, and 8 at 1, 5, and 10 minutes, respectively, and the umbilical artery pH was 7.0 with a base excess of -13 mmol/l.

On the 3rd day of life, when the baby was crying, a V-shaped defect of the superior part of the sternum (measuring about 3 × 2 cm) was noted. Throughout the respiratory cycle (Movie), there was notable recession during inspiration and bulging during expiration (Fig. 1). In addition, pulsations of the heart and of major vessels could be seen and palpated underneath the skin.

**Fig. 1**

Throughout the respiratory cycle marked recessions (during inspiration) and bulging (during expiration) over the upper part of the sternum can be noted.

No other abnormalities could be detected on physical examination. In particular, there was no heart murmur, normal pulses and normal pre- and postductal oxygen saturations (98 % and 97 %, respectively). Echocardiography showed normal heart anatomy and function.

During the first days after delivery, the baby showed mild signs of respiratory distress on exertion (for example when drinking or crying), such as nasal flaring, chest retractions and intermittent grunting. Oxygen saturations remained above 90 % on room air at all times. Due to hypoglycemia, the baby was fed by a nasogastric tube for a total of two days, and hyperbilirubinemia was treated with phototherapy. On day of life 6, the baby was discharged home without any signs of respiratory distress, stable blood sugar levels and normal serum bilirubin levels.

At the age of seven months, surgery was performed to close the sternal cleft. Preoperatively, the nipples, the upper sternal cleft, the lower borders of the thoracic cage and the xyphoid process were outlined on the skin as landmarks (Fig. 2A). After a median incision over the sternum, skin flaps were raised on either side and the upper sternal cleft (3 × 2 cm) was identified (Fig. 2B).

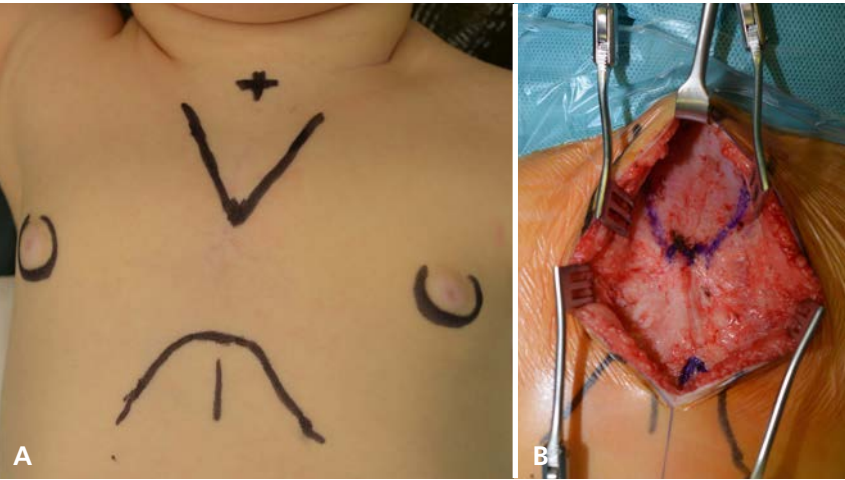
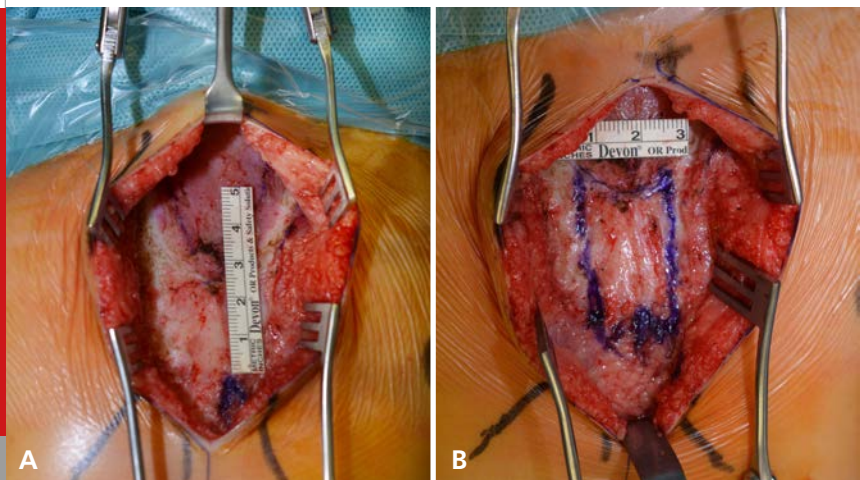


Fig. 2

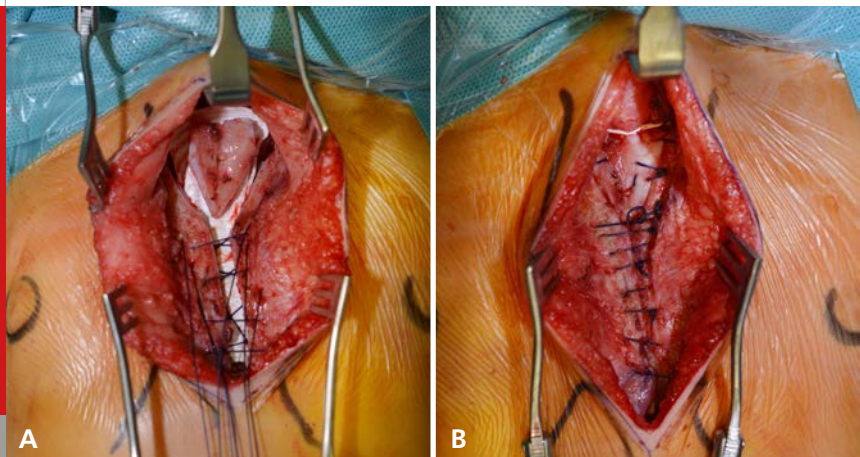
A: Preoperatively, landmarks are outlined: mammillae, upper sternal cleft, lower border of the thoracic cage and the xyphoid process; B: appearance after surgical preparation.

Fig. 3



Dimensions of the upper sternal cleft (USC): distance from xiphoid process to USC 3 cm (A), width of USC 2 cm (B).

Fig. 4



*A: Osteoplasty: part of the lower sternum is resected and transplanted to the upper defect (note that a dual-mesh non-absorbable membrane was inserted for protection underneath the sternum);
B: appearance prior to skin closure.*

The upper sternal cleft measured around 3×2 cm and the intact sternum from the xyphoid process to the lower end of the cleft measured 3 cm (Fig. 3).

Part of the lower sternum was resected and transplanted to the upper defect. A dual-mesh non-absorbable membrane was inserted for protection underneath the sternum (Fig. 4A). The two sternal halves were then approximated by sutures (Fig. 4B). After suture of the pectoral muscles, the skin was closed with an absorbable subcuticular running suture.

The postoperative course was unremarkable, and the baby was discharged on the 5th postoperative day. On follow-up 4 months later, the free graft had healed well, practically restoring a normal sternum for the child (Fig. 5).



Fig. 5

Clinical appearance at follow-up (age of 11 months)

Sternal cleft is a rare idiopathic congenital chest wall malformation with unknown etiology and a reported incidence of 1 case per 100'000 live births (1). Only sporadic cases have been described in the literature (2). Sternal cleft accounts for approximately 0.15 % of all chest wall malformations (3). The first case was described by Torres in 1740 (4). Females are affected more frequently than males (61 % versus 39 %, respectively) (5).

The malformation can be classified into two major forms. In complete sternal clefts, there is no bone present between the hyoid and the pubis in contrast to partial forms with a defect in the superior or inferior part of the sternum (3, 6). Isolated central bony defects as a fusion failure of the sternum in its middle portion are extremely rare (2). The most common form, though still rare, is the partial superior type, accounting for 67 % of all patients, followed by the complete form (19.5 %), the partial inferior form (11 %) and the sternal foramen (2.5 %) (5). The space between the costal ridges may be V-shaped when the cleft reaches the xiphoid process, or broad and U-shaped with a bony bridge joining the two edges, ending at the third or fourth costal cartilage (7).

The superior sternal cleft is often an isolated defect but can be also associated with other malformations like cervicofacial hemangiomas or abdominal raphe (8, 9). It is important to inform the parents that the

timing of the appearance of hemangiomas is variable. They can also appear after surgery (10).

Inferior incomplete clefts are usually associated with other defects of the anterior chest wall such as ectopia cordis or the pentalogy of Cantrell (ectopia cordis, intracardiac defects, sternal cleft, omphalocele, pericardial defect allowing communication with cavity) (11). Other rare associated malformations such as gastroschisis, VACTERL syndrome, Dandy-Walker-syndrome, pectus excavatum or chest wall hamartomas have also been described (2).

Sternal clefts result from defective embryologic fusion of paired mesodermal bands in the ventral midline. In regular development, two mesenchymal bars fuse between the seventh and tenth week of gestation, starting cranially at the manubrium and finishing distally at the xiphoid process (2). Disturbances of normal ventral midline thoracic fusion can present as a spectrum of abnormalities, including a prominent suprasternal notch, irregularities in the shape of the xiphoid, ectopia cordis, superior, inferior or complete sternal clefts (10).

Many etiological factors had been linked to this malformation (2), including alcohol consumption early in pregnancy (10) and nutritional deficiencies, especially lack of methylcobalamin (vitamin B12) or riboflavin (vitamin B2) (1, 2). Disruption of the HOX b gene might

be involved in the development of sternal clefts (1, 5, 10). The malformation occurs sporadically, although autosomal recessive forms have also been described. Notably, many cases have been reported from the Middle East (2).

Sternal clefts are often asymptomatic (74 %), and diagnosis can be easily made clinically at birth by palpation and inspection due paradoxical respiratory movements (Fig. 1, 2, Movie). Therefore, diagnosis is usually possible in the neonatal period (64 %), however, several authors have reported patients diagnosed at older ages, (i.e., in adolescence or even adulthood). Imaging studies, including chest X-ray, computerized tomography, and careful cardiologic evaluation (electrocardiogram and echocardiography) can help to identify any associated anomalies. Prenatal diagnosis is difficult and sternal clefts seem to be more easily identified when associated with cardiac anomalies. The second trimester is the best period for prenatal sternal study (2, 5, 10).

Early recognition of the malformation is important since it is associated with an increased risk of life-threatening events involving mediastinal viscera and vessels (10). Prior to surgery, associated defects and syndromes should be excluded (5).

Surgical treatment is generally recommended, if possible in the neonatal period to take advantage of

the elasticity of the thoracic cage and to achieve primary closure. Surgical approaches include primary closure with autogenous tissue (73 %), bone graft interposition (10 %), prosthetic closure (7 %) or muscle flap interposition (3 %). During the first months of life, the flexibility of the chest wall is maximal, and compression of underlying structures is minimal. Patients undergoing surgical repair after the age of three months have required more postoperative supportive care and shown a higher incidence of cardiac complications due to the reduction of the thoracic volume (10). Sometimes, partial or total removal of the thymus is necessary (2, 5). Intraoperative complications have been reported due to pericardial or pleural tears. Postoperative complications include retrosternal seromas or pneumothoraces (5).

See also: Case of the Month May 2008: A case of a rare thoracic malformation with paradoxical chest wall movement.

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