

Complete cleft sternum

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ABSTRACT A case of complete cleft sternum is presented along with the nomenclature of sternal defects. It is recommended that the term *ectopia cordis* should be applied only to cases in which the heart and thoracic viscera are genuinely ectopic. Surgical correction of complete cleft sternum should be performed in the neonatal period whether the infant is symptomatic or not. Simple closure of the defect, as for a median sternotomy, is possible during the first month of life and this avoids the more complex reconstructions necessary in older children.

Major congenital abnormalities of the sternum are rare. When present they may be associated with other congenital malformations, particularly abdominal wall defects and *ectopia cordis*. Many of these defects, which have been called bifid sternum, cleft sternum, or sternal fissure, can be corrected surgically. We have recently seen again a case of complete cleft sternum which was successfully operated on in 1974. This case forms the basis of our report in which the classification of cleft sternum and the surgical management of complete clefts are discussed.

Case report

A 10-day-old infant from Saudi Arabia was admitted to the Brompton Hospital in January 1974. She was the product of a full-term, uncomplicated pregnancy and normal delivery. The parents and three siblings were well and without congenital abnormality.

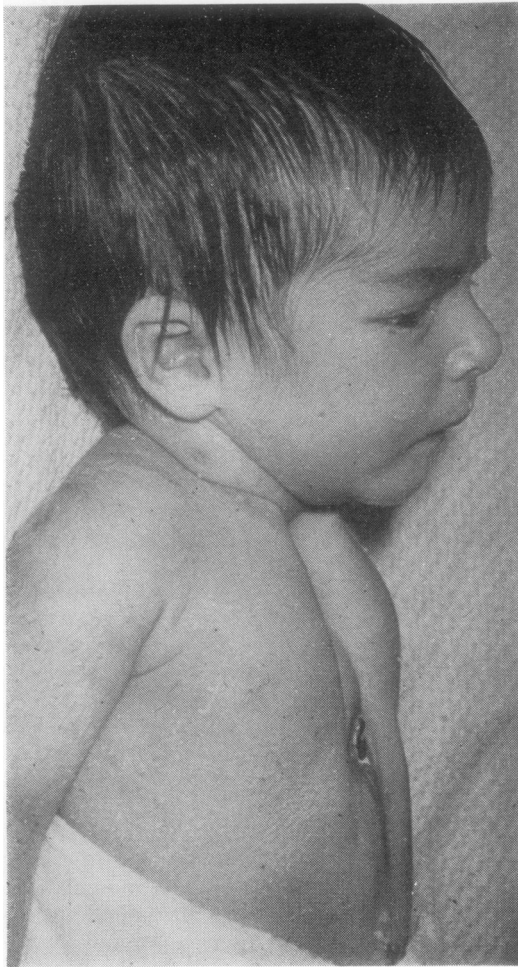
On examination the infant was well nourished and weighed 3.4 kg. There was an ellipse of abnormal, partly ulcerated pink skin on the anterior chest with no apparent sternum deep to it (fig 1a). This left a defect between the ribs, which was up to 5 cm across. The heart and thoracic viscera could be seen and felt to move paradoxically in this defect during respiration, and protrude through it alarmingly when the child cried (fig 1b). It was possible to approximate the ribs in the midline by applying gentle lateral pressure. This manoeuvre did not embarrass respiration or affect blood pressure. The heart sounds, which were normal, and the cardiac impulse were most easily detected to the right

of the midline. Physical examination was otherwise unremarkable and no other congenital abnormality was found. A chest radiograph taken while the child was crying indicated a cleft sternum with the thoracic viscera projecting anteriorly. A clinical diagnosis of complete cleft sternum and possible dextrocardia was made. Surgical correction was recommended and performed on the day after admission.

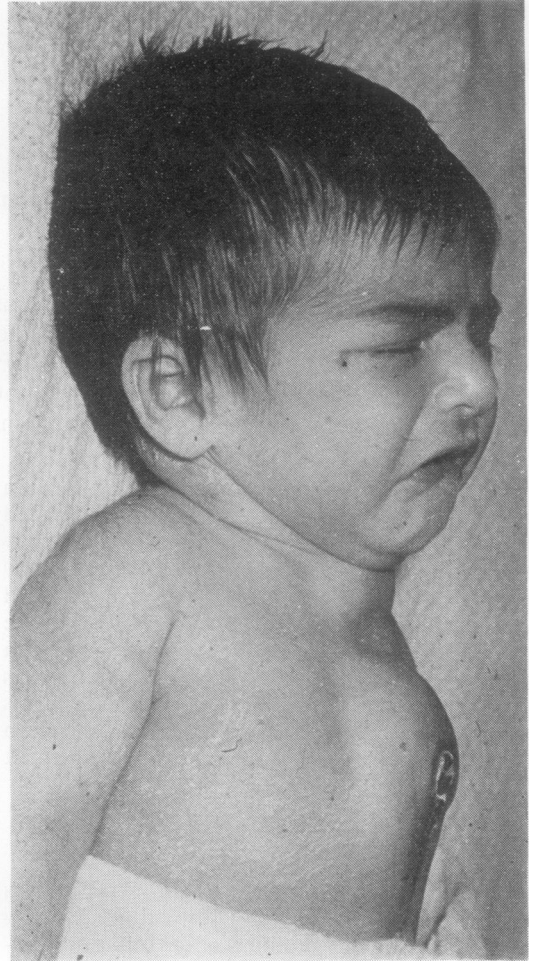
At operation a vertical elliptical incision was made excising the abnormal skin. The pericardium, which was adherent to the skin, was opened inadvertently and immediately closed by suture. A complete cleft sternum was found; the sternal bands were well formed and converged inferiorly to the linea alba of the abdominal musculature. There was no deficiency of the abdominal musculature or diaphragm, but the pectoral and sternomastoid muscles were separated by the span of the sternal defect. The sternal bands were dissected free of the pleura and their medial borders freshened. They were then approximated with interrupted nylon sutures without difficulty. Cardiorespiratory embarrassment did not occur, and the closure was completed with subcutaneous and skin layers. No muscular layer was required as the pectoral and sternomastoid muscles assumed a normal position with closure of the sternum.

The child made an uneventful recovery and was discharged from hospital on the eleventh day after operation.

We have recently reviewed this child who is now a healthy 5-year-old. She has developed normally and is asymptomatic. Physical examination is unremarkable apart from an inconspicuous midline scar and an apex beat which is still displaced to the right. The thoracic cage is well-developed, symmetrical, and without deformity.



(a)



(b)

Fig 1 *The infant lying (a) at rest and (b) while crying. The thoracic viscera can be seen to protrude when the infant cries.*

A chest radiograph shows situs solitus with dextrocardia and a left aortic arch.

Discussion

The development of the sternum has been studied by several authors.¹⁻⁴ It is thought that the sternum has a common origin with the pectoral musculature and largely develops from the lateral plate mesoderm. Cells from the lateral plate mesoderm migrate ventrally in the sixth intrauterine week to form two parallel mesenchymal bands or bars one on each side. These bands fuse

craniocaudally in the midline by the tenth intrauterine week to become the body of the sternum and part of the manubrium. Three other small mesenchymal primordia, which arise between the developing clavicles, complete the cranial part of the manubrium. The sternum chondrifies and then ossifies from multiple ossification centres which appear in sequence from cranial to caudal, beginning at the sixth intrauterine month. At birth the sternum is mainly cartilage.

Although the embryology of the sternum has been carefully studied, the aetiology of sternal clefts is obscure. It is thought that most isolated sternal defects arise from a failure in development of, or a failure in fusion of the mesenchymal elements from which the sternum is derived. In cases where ectopia cordis is present, the

failure of sternal fusion is probably secondary to the malposition of the thoracic viscera.

Cleft sternum comprises a group of rare congenital anomalies which can be classified as superior, inferior, or complete clefts. A superior cleft is either a U-shaped defect with the cleft ending at the level of the fourth costal cartilage,⁵ or a V-shaped defect if the sternum is cleft to the xiphoid process.^{6,7} Inferior clefts are usually associated with other abnormalities of midline fusion. Some of these cases have a characteristic group of defects which have been called the pentalogy syndrome (ventral hernia, cleft distal sternum, deficiencies of the diaphragm and diaphragmatic pericardium, and a congenital heart defect).⁸ Complete cleft sternum has only been reported on a few occasions.⁹⁻¹⁴ In addition there is a case initially reported as absence of the sternum in which sternal elements have been demonstrated on subsequent radiographs.^{5,15} With the exception of this last case, the defect was an isolated complete cleft of the sternum as illustrated in fig 2. In the case reported by Ravitch there were also abdominal wall, diaphragmatic, and pericardial defects more reminiscent of an inferior cleft sternum.

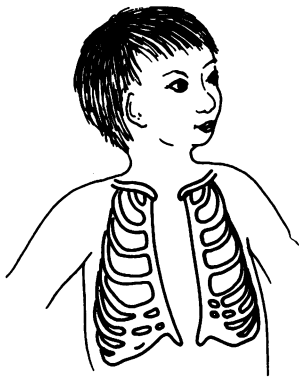


Fig 2 *Extent of the skeletal abnormality in complete clefts of the sternum.*

The term *ectopia cordis* has given rise to confusion in published accounts of these sternal anomalies. In most cases of partial or complete cleft sternum, the thoracic viscera are prominent and mobile, being covered only by soft tissues. They are, however, correctly sited anatomically and do not require reduction when the sternal defect is closed. Although some authors would describe these cases as having partial *ectopia cordis*,^{14,16} we prefer to use the term cleft sternum alone for these defects and reserve the term *ectopia cordis* for those rare cases in which the

heart is ectopic.¹⁷ We feel that this is a valid distinction for the surgeon to make as the presence of true *ectopia cordis* necessitates a challenging surgical reconstruction. Attempts to correct *ectopia cordis* have generally been unsuccessful because of the presence of multiple cardiac and extracardiac anomalies^{17,19}; a marked contrast to operations for cleft sternum which are usually successful.

Major defects of the sternum make the chest wall unstable and allow the thoracic viscera to move paradoxically during respiration. Some infants tolerate this paradoxical chest movement well, but others suffer attacks of cyanosis and develop recurrent chest infections. Symptomatic infants clearly require early operation. Asymptomatic infants pose a more difficult problem. Although long-term asymptomatic survival has been recorded in untreated cases,²⁰⁻²⁴ there are good reasons to advise early operation. First, the appearance of the child with the heart bulging through the chest wall is very disturbing to the parents. Second, the heart is denied its normal protection of the sternum and is at increased risk from direct trauma. Third, the patient may be predisposed to recurrent chest infections by paradoxical movement of the lungs. Finally, it is simple to correct the abnormality in the first four weeks of life.

The type of operation required to correct a complete cleft of the sternum depends on the age of the patient. Within the first month of life the defect can be closed as one would close a median sternotomy in a child of the same age. In addition, attention must be paid to the midline skin abnormality which is a consistent feature of this type of cleft sternum. Its appearance varies from that of a healed scar to an actual skin deficiency. The area is probably best excised taking care to avoid the pericardium, which is adherent to it. In older children it is not usually possible to approximate the sternal bands without causing distress. This is because of the rapid growth of the intrathoracic organs and the tendency of the defect to separate as a result of the weight of the shoulders pulling laterally.¹⁵ The sternal defect can only be closed then if relaxing costal chondrotomies are performed,^{13,14,18} or part of the defect is filled rather than closed.¹¹ The results of treatment of partial sternal clefts indicate that this first type of operation using relaxing chondrotomies could probably be used in children up to the age of 4 years²⁵ although no case of complete cleft sternum has been operated on after 2 months of age. A variety of other procedures has been used to correct sternal defects

in still older children. Defects have been filled with autogenous cartilage and bone grafts,^{26 27} and various prosthetic materials such as Marlex mesh, Teflon felt,¹⁵ and Acrylic plate.⁷

The long-term results of surgery for complete clefts of the sternum are largely unknown as only two of the reported cases had a documented follow-up of more than a few months. The case of Ravitch^{5 15} followed for 12 years after correction of multiple sternal, pericardial, diaphragmatic, and abdominal wall defects, has had a good result although additional Teflon felt was required one year after the primary procedure. Our own case has been followed for five years now, and the result has been good, with normal growth of the chest wall.

In common with most other authors,^{9 12 13} we believe that the best time to operate on a child with a complete cleft of the sternum is in the neonatal period, whether the child is symptomatic or not. The operation is simple at this age and relatively safe in the context of a neonatal referral centre. If the child is for some reason unfit for operation at this stage or presents after the neonatal period, the case will have to be assessed on its own merits. Reconstruction will still be possible, but it will be a little more difficult to produce an aesthetically pleasing result.

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