Pediatr Cardiol 23:472–473, 2002
DOI: 10.1007/s00246-002-1470-z

Pediatric Cardiology

Images in Pediatric Cardiology

Congenital Sternal Clefts

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Sternal clefts are rare congenital malformations caused by the failure of fusion of the sternal elements. They may be classified as partial or complete. Partial sternal clefts may involve the superior or inferior aspect of the sternum. Superior sternal clefts have been associated with other minor lesions, including vascular dysplasias and supraumbilical raphe. In contrast, inferior sternal clefts are often associated with other significant midline field defects, such as ectopia cordis. Pentalogy of Cantrell is the constellation of an inferior sternal cleft, ectopia cordis, midline abdominal defect or omphalocele, a pericardial defect allowing communication between the pericardial and peritoneal cavities, and one or more cardiac defects. Complete failure of sternal fusion, or asternia, is the rarest form.

The diagnosis and workup of a patient with a sternal cleft are straightforward. The defect is readily apparent on physical exam. Chest radiograph is of

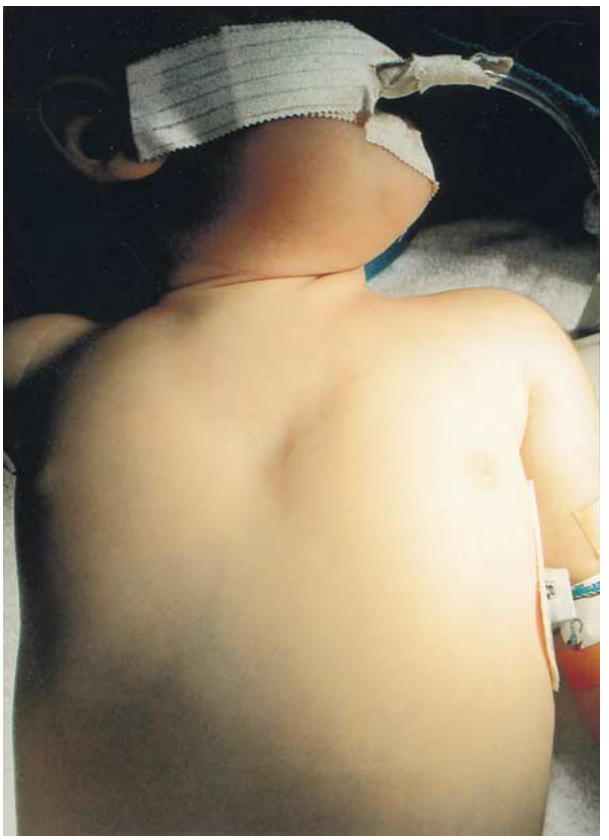

Fig. 1. Preoperative image demonstrating the V-shaped depression of the sternal cleft.

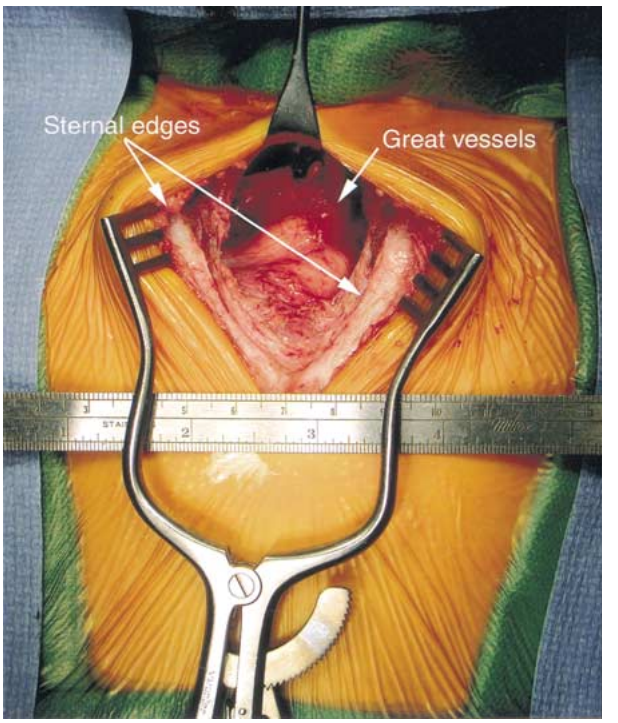

Fig. 2. Intraoperative image showing the V-shaped sternal defect with underlying heart and great vessels.

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limited use in further defining the defect but may delineate situs or reveal other evidence of cardiac defect. An echocardiographic examination is warranted, particularly in the presence of an inferior cleft. Further workup in patients with more extreme variants, such as pentalogy of Cantrell, is individualized.

Sternal clefts are closed to protect the underlying heart and great vessels from trauma and to improve cosmetic appearance. In addition, respiratory dynamics may potentially be compromised due to paradoxical movement of the thoracic organs during respiration. Rarely, patients may present with symptoms of respiratory compromise or recurrent respiratory infections.

The goals of therapy should be to safely provide adequate protection of the thoracic viscera, maintain the growth potential of the chest wall, avoid the use of prosthetic material when possible, and attain a satisfactory cosmetic result. In the neonate, the ideal procedure is primary closure. When diagnosed in the neonatal period, sternal clefts should be repaired early, when the defect is most amenable to simple closure. During this time, the chest wall is maximally flexible. In addition, over time the thoracic organs accommodate to the size of the thoracic cavity and defect, making primary closure more difficult. In the older patient, many different techniques utilizing autologous and prosthetic materials have been described, has, as rarely, primary closure. However, prompt recognition of the sternal cleft with referral for early primary closure results in the optimal outcome for this rare malformation.

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