

IMAGES IN CLINICAL PRACTICE

CONGENITAL MIDLINE MALFORMATION IN A NEWBORN

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A full-term boy was born following an uncomplicated pregnancy with normal prenatal ultrasound findings. After birth on examination an U-shaped upper sternal cleft, with atrophic skin covering the defect and a small area not covered by skin was seen. There was a two centimeters gap between the sternal laminae. A mediane abdominal raphe extended from the sternal defect to the umbilicus was also seen (Figure 1). A cardiac murmur was detected during auscultation. No other abnormalities were acknowledged on physical examination. A transthoracic echocardiography was performed which revealed a large perimembranous interventricular communication, right midventricular obstacle and an ostium secundum interauricular communication. He started to follow-up in pediatric cardiology and cardiothoracic surgery consultations. He had chronic heart failure and failure to thrive. At 5 months of age, primary closure of the sternal cleft along with correction of heart defect was performed. Currently, two years post surgery, he is asymptomatic, with adequate height-weight progression.

What are sternal clefts? What is the best approach?

Sternal clefts are a rare congenital malformation, with an incidence of <0.15%, that results from a failure in the fusion of the sternal bands at an early stage of embryonic development.^{1,2} It may present isolated (27%) or be associated with other malformations (73%) such as hemangiomas, PHACES syndrome, cantrell pentade, abdominal raphe or cardiovascular abnormalities.^{3,4} When associated with cardiac abnormalities, the prognosis may be unfavorable.¹ Patients are usually asymptomatic in the neonatal period. On physical examination, a paradoxical midline thoracic bulging with protrusion of the mediastinal viscera during expiration may be seen. If not timely diagnosed and treated, respiratory symptoms (dyspnea and cough), impaired gas exchange or lung infections may arise.^{4,5}

Surgical correction is therefore recommended and should be performed early because the greater sternal

Figure 1. Mediane abdominal raphe extended from the sternal defect to the umbilicus.



plasticity allows for primary closure without the use of autologous grafts/prosthetic material.¹ Indications for surgery are not only cosmetic but also for the improvement of respiratory dynamics, protection of mediastinal structures from direct injury and positive impact on growth.⁶

Compliance with ethical standards

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