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Upper sternal cleft associated with congenital heart disease

Introduction:Sternal cleft is a rare congenital malformation due to partial or total failure of fusion of the sternum in the early stages of embryological development. Upper clefts are mostly isolated, unlike total clefts, which are often associated with syndromes and whose incidence of heart disease is close to 8%. Although easily identifiable at birth, 1/3 of clefts are not diagnosed before one year of life. Case presentation: A 10-month-old female infant was brought to the clinic with a history of delayed weight and height development and fatigue during breastfeeding. On physical examination, a pulsatile mass was seen in the upper third of the sternum and a systolic murmur was heard in the lower left sternal border. The investigation was continued and the x-ray showed cardiomegaly, the transthoracic echocardiogram showed a 6 mm ostium secundum atrial septal defect and a 12 mm perimembranous ventricular septal defect with hemodynamic repercussions, and the chest tomography showed an upper sternal cleft, and the patient was referred for cardiac surgery. The ventricular septal defect was repaired with a bovine pericardial membrane, followed by direct suture of the atrial septal defect and the cleft was repaired by debridement of the upper third of the mediastinum, direct approximation of the sternal borders and fixation with steel wires. The patient progressed well, without complications, and was discharged from hospital on the 7th_{the}postoperative day. At 6 years of age, he presents discrete pectus excavatum and absence of residual shunt on the control echocardiogram. Discussion: Sternal cleft correction is indicated to restore bone protection of the heart and great vessels, improve respiratory dynamics by preventing paradoxical movement of the mediastinal viscera, and eliminate the visible deformity. Primary repair is more easily performed in the neonatal period due to greater compliance of the osteocartilaginous structures and less compression of the mediastinal structures. However, it can be postponed until the age and weight are appropriate for correction of associated heart disease, as occurred in this case due to late diagnosis. Final comments: Primary cleft repair is a safe procedure with good immediate and long-term results, which can be performed simultaneously with the correction of the heart defect. It allows adequate growth and development of the sternum with good aesthetic and functional results, and is the procedure of choice.

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References:

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