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Superior sternal cleft, cutaneous, and airway haemangiomas

A term baby, born to a 19 year old healthy mother by emergency caesarean section because of failure to progress, was noted to have a superior V shaped sternal cleft, measuring 4 cm across with lower ribs meeting in the midline (fig 1). The mother had no significant medical history.

Apart from a visible pulsatile heart, cardiological examination was normal. She did not have a midline abdominal raphe. At three weeks, she gradually developed superficial capillary haemangiomas over the lower sternum, lower lip, neck, and right side of the face (fig 1).

A computed tomography scan of the thorax at 2 months of age revealed a membrane in the place of the sternum, with normal lungs, major vessels, and vertebrae.

At 3 months of age, she presented with stridor. Excision of the sub-glottic haemangioma was performed, with partial excision of the vocal cord. She is fed by gastrostomy because of aspiration. She is developing well while awaiting repair of the sternal defect.

Superior sternal cleft is almost always an isolated abnormality but can occasionally be associated with cervicofacial haemangiomas (vascular dysplasia)¹ and PHACES (posterior fossa brain malformations, facial haemangiomas, arterial anomalies with coarctation of aorta, cardiac defects, eye abnormalities, sternal cleft, and supraumbilical raphe) syndrome.² The aetiology of this condition (OMIM 140850) is not known. Facial haemangiomas are common in female infants.³ Most cases are sporadic with few reported recurrences in siblings.⁴ Studies on ventral body development in mice indicate impairment in Hoxb gene expression as a possible cause of sternal defects.⁴

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Figure 1 Superior sternal cleft with superficial capillary haemangiomas over the lower sternum, lower lip, neck, and right side of the face. The patient's parents have given permission for publication of this figure.

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