Congenital absence of sternum in an infant

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DESCRIPTION

A female baby aged 6 months, born to non-consanguineous parents presented with anterior chest wall deformity since birth. On examination, an absent palpable sternum along with paradoxical respiratory movements with visible cardiac contractions and vasculature were seen beneath a thin layer of skin which led to the diagnosis of congenital absence of the sternum (figure 1 and video 1). The rest of the examination was found to be normal with no other obvious congenital anomalies. CT scan of the thorax showed mild shift of the mediastinum towards the right side with the pericardium abutting the anterior chest wall and absence of ectopiacardis. There was no other associated underlying malformation.

Congenital complete sternal cleft results from failure of the process of midline mesenchymal



Figure 1 The absent sternum along with visible cardiac contractions and vasculature were seen beneath a thin layer of skin.



Video 1 The absent sternum along with visible cardiac contractions and vasculature were seen beneath a thin layer of skin.

strip fusion during embryonic development.¹ These patients are at increased risk of mediastinal injury, hypothermia and insensible fluid losses.² ³ It may be associated with Cantrell's pentalogy, PHACES syndrome and Poland syndrome. Reconstructive surgery of absent sternum should be performed by primary closure using combined periosteal advancement flap and sliding osteochondroplasty during the neonatal period when the chest wall is highly compliant and closure can be achieved without significant cardiopulmonary compromise.² ³

Learning points

- Congenital absence of the sternum is a rare malformation of the chest wall which results from failure of the process of midline mesenchymal strip fusion during embryonic development.
- Patients with complete sternal cleft are at increased risk of mediastinal trauma, hypothermia, increased insensible fluid losses, cyanosis and recurrent infections of the chest.
- ▶ With increasing age, surgical correction becomes difficult due to a decrease in compliance of the chest wall necessitating early surgical correction in the neonatal period.

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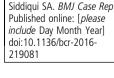
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