Congenital Thoracic Wall Deformities

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Congenital absence of ribs producing herniation of the lung through the gap in the rib cage and paradoxical respiration is rare and so is the anterolateral protrusion of chest wall due to abnormal prominence of costochondral junction. We report one case belonging to each of these categories treated successfully.

Case Reports

Case 1: A 2-year-old female child, was admitted with repeated attacks of respiratory tract infection, respiratory distress, failure to thrive and bulging of right sided chest wall in the axilla with progressive deformity of thoracic spine since birth. There was an obvious soft tissue swelling due to herniation of lung through the lateral thoracic wall with paradoxical respiration, confirmed on screening.

X-ray chest showed a wide gap between right second and third ribs with soft tissue bulge, fusion deformities of right fourth, fifth and sixth ribs as well as seventh and eighth ribs, congenital absence of two lower ribs of right thoracic cage, contralateral mediastinal shift and scoliosis of thoracic spine (*Fig. 1a*).

At operation, osteotomies and approximation of 2nd and 3rd ribs after splitting them longitudinally provided bony struts across the defect, corrected the herniation of the right-lung and reduced the contralateral shift of mediastinum and the scoliosis (*Fig. 1b*). The repair was reinforced and strengthened by covering the defect by onlay muscle flaps from neighbouring muscles. The fusion deformities were also corrected by splitting the fused ribs at the point of fusion laterally. The procedure was done extrapleurally through right posterolateral approach. The postoperative period was uneventful. The patient is asymptomatic and thriving well after one year of follow up.

Case 2: An 8-year-old male child had an abnormally prominent right sided cen-

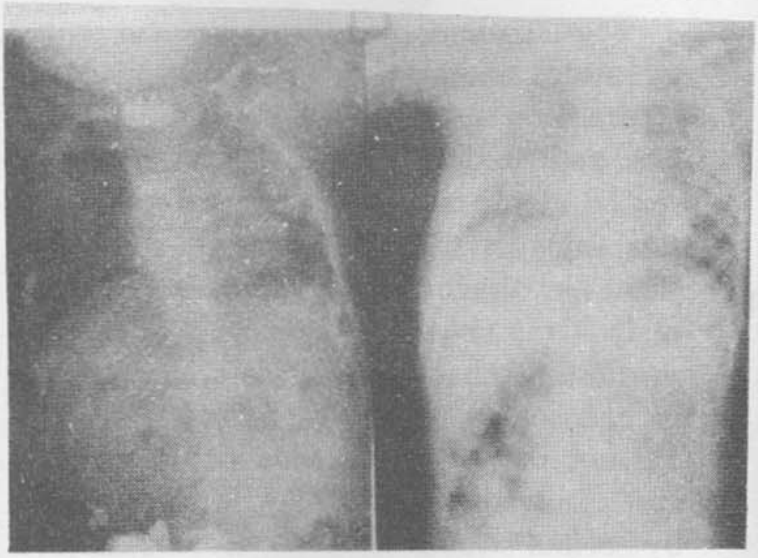

Fig. 1. Pre and one year follow up chest X-ray of case 1. Note absence of herniation of right lung, shift of mediastinum and correction of scoliosis at one year follow up.

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tral portion of anterior thoracic cage since birth. He was diagnosed as a case of rickets in infancy and that of tuberculosis of thoracic cage later. However, treatment for both these conditions did not give any relief. Moreover, unilateral pathology with normal clinical, laboratory and radiological findings were puzzling. He was referred to us finally. On examination, there was abnormal prominence of 3rd to 6th costochondral junctions of right thoracic cage, leading to obvious unilateral anterolateral bulge of the chest wall. The patient was asymptomatic but the parents were upset because of psychological and cosmetic reasons.

Corrective surgery included subperiosteal excision of deformed costal cartilages from 3rd to 6th rib through a small transverse submammary incision extrapleurally under ketamine anesthesia. The postoperative recovery was smooth and the patient was discharged home on the following day. Histological examination of the excised deformed costochondral junctions showed normal histology with no evidence of rickets or tuberculosis. The parents were pleased by excellent cosmetic and psychological results.

Discussion

Very few reports of congenital absence of ribs as well as thoracic cage deformities have been reported in the Indian Literature(1). Most defects of the ribs lie parasternally and usually involve the lower ribs between 6 and 12(2-4).

Congenital absence/fusion deformities of ribs leave pediatric surgeon with the challenging problem of reconstruction. The aims and objects of construction of these defects are:

(*i*) to provide stability to chest wall in order to prevent paradoxical respiration;

(*ii*) to cover and protect vital organs;

(*iii*) to achieve the above in a cosmetically satisfactory way; and

(*iv*) to prevent or correct secondary spinal deformities.

Treatment depends on the size and site of the defect, associated malformations, general state of health of patient and age of the baby. The defect should be corrected as soon as possible because some of them may grow in size and also correction is more difficult in older children(2-4).

It is at times possible to minimise the paradoxical respiration and hence the constant shifting of the mediastinum by strapping the defect in neonatal period(4). The operative treatment of large defects, as seen in our case, requires longitudinal splitting of the ribs adjacent to the defects and their anterior and posterior osteotomies to provide bony struts across the defect and have been very successful. Bone graft operations using bone either from adjacent, opposite or lower ribs with costal margin or other parts of the skeleton have been advised in severe defects. Covering the defect is further strengthened by the overlying muscles and their flaps sutured across the defect.

Prognosis is good if associated malformations are absent and the treatment carried out at an earlier age. Recurrence is rare but the frequently associated scoliosis can become progressively worse and may need early corrective treatment.

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Bernard Soulier Syndrome

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Bernard-Soulier (BS) Syndrome is one of the hereditary hemorrhagic disorders(1). Similar to von-Willebrand's disease, there is a defect in platelet adhesion in BS Syndrome. Earlier two siblings with this disorder were reported from India(2). Here we report 4 unrelated patients detected in our laboratory in last 15 years.

Case Reports

Case 1: A 6½-year-old male child presented with history of ecchymoses on different parts of body and bleeding from mucosal surface of lips since one year of age. There was also prolonged bleeding following tooth extraction. At each episode, the bleeding had continued for 12-16 hours. One month back, he had prolonged gastro-intestinal bleeding and two units of whole blood with hematolcele and streptochrome were given. His two female siblings 11 and 8½ years old, had similar hemostatic defect. History of consanguinity was absent.

Case 2: A 2½-year-old male child presented with repeated episodes of excessive bleeding in last one year following history of blunt trauma. Epistaxis occurred once only. Six months back he had prolonged gum bleeding and was controlled with 2 units of whole blood transfusions. His brother (10 years age) was apparently asymptomatic. There was no history of consanguinity.

Case 3: An 18-year-old female, presented with history of gum bleeding since 4 years of age. At menarche she had menorrhagia. It was controlled by combined hormonal pills (Ovral). Other bleeding problems were of mild in nature and did not necessitate any blood transfusion. In the past one year, she had been put on multiple courses (4-6 weeks each) of oral corticosteroid therapy with symptomatic relief. There was no family history of any hemostatic defect or consanguinity.

Case 4: A 7-year-old male child, presented with bleeding for 48 hours from gums following tooth extraction. History of prolonged bleeding on injury was present since 2 years of age. On examination, a small hematoma was present on forehead. She did not need any blood transfusion in the past. There was no family history of excessive bleeding or consanguinity.

Screening tests of coagulation (*Table 1*) were done by standard techniques(3). Platelet factor 3 (PF_3) availability was determined by the method of Hardisty and

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