different areas of small intestine including stomach(3).

Histological findings include absence of ganglion cells in the myenteric and submucosal region of the entire gastrointestinal tract, but hypertrophied nerve trunks seen in classic Hirschsprung's disease may be absent(1). Associated congenital anomalies are very rarely reported with total intestinal aganglionosis(1,3). Available evidence suggests a familial occurrence of the disease and the mode of inheritance is autosomal recessive with a significantly high risk of recurrence (25% in each pregnancy)(1). No satisfactory therapeutic procedure is yet available for total intestinal aganglionosis, however, long segment small bowel myectomy myotomy(2) with total parenteral nutrition have been reported as a successful measure for long segment small bowel aganglionosis. Intestinal transplantation may be a successful method of treatment in the future.

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Congenital Absence of Ribs

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Congenital anomalies of chest wall are a relatively common occurrence but unilateral agenesis of ribs with a "lung hernia" producing an unstable chest is one of the rare deformities encountered(1-6). We wish to present our experience with the use of a temporary external chest splint for stabilization of the congenital flail chest in two neonates.

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Case Reports

Case 1: A 2.1 kg male neonate was born at term after an uncomplicated pregnancy. Acute respiratory distress was noted within a week after birth from agenesis of ribs T_7 to T_{12} on left side with a lung hernia and paradoxical respiration with marked scoliosis of the thoracic spine (*Fig. 1a*).

The baby also developed recurrent respiratory tract infections and failure to thrive in addition to tachypnea, dyspnea and hypercarbia from paradoxical respiratory movements and higher attachment of left hemidiaphragm. A noninvasive method for temporary chest wall stabilization was initially achieved by using micropore adhesive strapping over the cotton gauze pad. As the delicate skin showed excoriation after two weeks, we devised as external chest wall splint for prevention of paradoxical chest movements. A vest made from a low temperature thermoplastic material was moulded to the contour of the child's left hemithorax with a shape so that full range of arm motion was possible and other activities remained unrestricted. The vest was secured with an adjustable belt wrapped comfortably around the entire chest and vest. The infant improved following application of this splint and was discharged. There was no lung hernia or paradoxical movements and scoliosis of spine showed improvement at 2 month follow up (*Fig. 1b*). The splint was worn continuously except for short periods during bathing. It appears that eventually he would out grow the original vest and a larger one would be required.

Case 2: A 2.5 kg full term male neonate, was born following an uneventful pregnancy and normal vaginal delivery at hospital. He was noted to have an abnormal bulge in the right anterolateral chest wall associated with paradoxical respiration. *X*-ray chest showed agenesis of ribs T_3 to T_6 on right side with elevation of

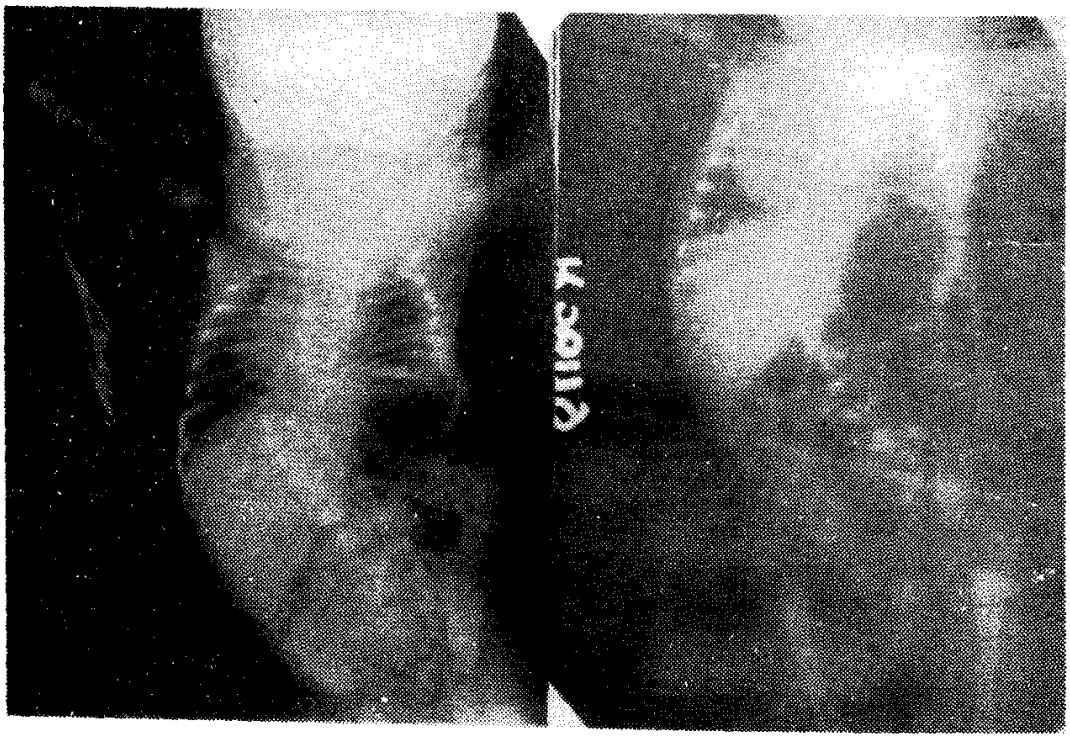

Fig. 1. Pre and post treatment infantogram of Case 1. Note absence of lower ribs on left side with lumbar hernia and intestinal contents in it and distended stomach below the left hemidiaphragm.

right side (*Fig. 2a*). He was put on conservative micropore adhesive strapping to stabilize the right chest wall but developed blisters and erythema of the underlying due to hypersensitivity. It was, therefore, thought to apply flexible plastic external splint similar to the one used in Case 1 with a sling passing over right shoulder and an adjustable velcro belt around the whole circumference of the chest wall. He remained asymptomatic and is thriving well at 4 month follow up. *X*-ray chest at follow up showed right hemidiaphragm in its usual position with heart in its normal position and no paradoxical movements. Expansion of both lungs was adequate and mild scoliosis of thoracic spine also got corrected (*Fig. 2b*).

Discussion

The treatment of congenital absence of ribs is to stabilize the chest wall and to prevent paradoxical respiration.

Life threatening respiratory insufficiency due to paradoxical respirations caused by large defects resulting in severe respiratory distress syndrome requires endotracheal intubation and ventilatory support until definitive chest wall reconstruction by an emergency operation can be performed(2,3,7).

Medium sized and small defects should also be corrected as soon as possible because some show a tendency to grow in size and correction is much more difficult in older children(8). However, the risks and challenges of complex reconstructive surgery and anesthesia in a neonate are well known.

It was, therefore, thought to develop a novel non-operative approach in neonatal period for prevention of paradoxical respiration by a method of temporary chest wall stabilization which would permit the child to grow and thrive at home. It is at times possible to minimize the constant shifting

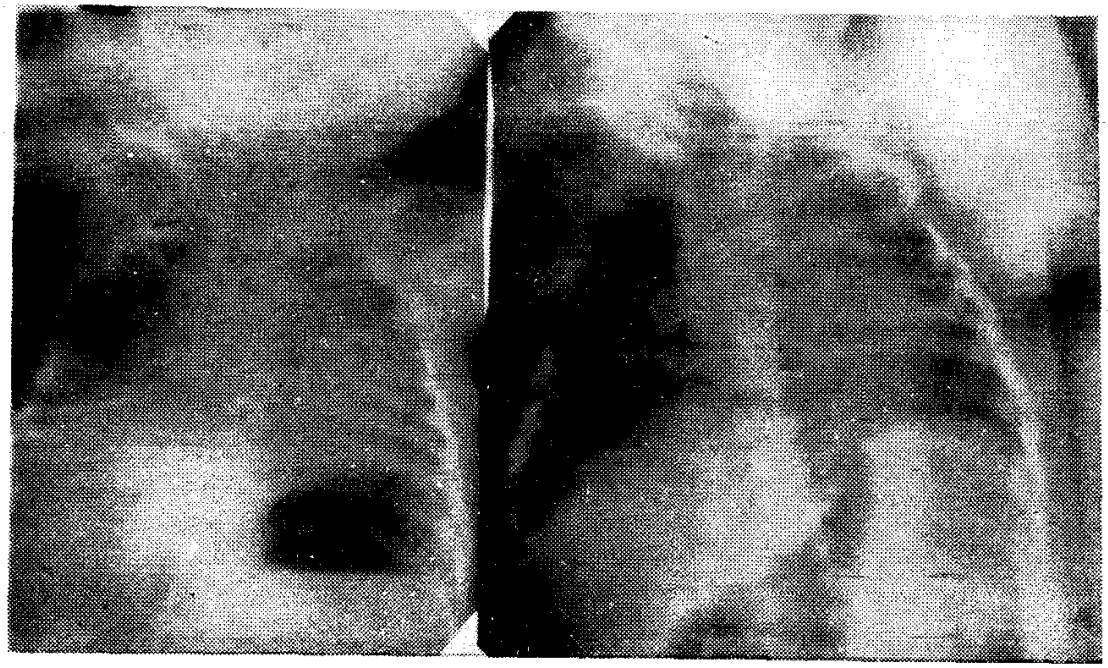

Fig. 2. Pre and post treatment X-ray chest of Case 2. Note absence of upper three ribs on the right side.

of the mediastinum by strapping the defect which may be sufficient in itself to relieve symptoms without any further treatment. Surgical alternatives could be considered for repair of the defect if recurrent chest infection continues in such a conservatively treated child, whatever the size of the defect.

The duration after which an infant does not require wearing of vest depends on site, size and extent of rib agenesis and the severity of associated bony, spinal, thoracic or muscular anomalies. Its use should be continued till the infant is asymptomatic or grown old enough to perform safe surgery. On removal, severity of respiratory distress and hypercarbia does not recur in most mild and moderate defects but may appear in severe defects to a lesser degree. These cases may require definitive surgery.

The prosthesis works very well until the patient grows older at which time change of the size of splint or elective surgical repair at regional centre may be performed if the child remains symptomatic. We feel that this non-operative conservative treatment demonstrates the usefulness of an external splint for chest stabilization permanently or temporarily and should be considered in neonatal period in distant rural set up where there is no easy access to safe surgery.

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