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## Total Intestinal Aganglionosis

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Total intestinal aganglionosis with absence of ganglion cells from duodenum to the rectum has been reported in thirteen cases in the literature prior to 1985 with uniformly fatal outcome(1,2). Senyuz *et al.* have reported a case of total intestinal

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aganglionosis with involvement of the stomach(3). Absence of any satisfactory treatment makes it a unique but non salvageable form of intestinal aganglionosis. We report a case of total intestinal aganglionosis.

### Case Report

A full term second born eight-day-old female child weighing 2.8 kg, presented with bilious vomiting and abdominal distention since the second day of life. She passed small amount of meconium on the third day of life. The other sib, a one-year-old boy, was normal. There was no family history of Hirschsprung's disease or any major congenital anomalies in the near relatives.

At hospital, the child passed small amount of meconium but continued to have bilious vomiting and progressively increasing abdominal distention with radiological evidence of low small bowel obstruction. At laparotomy the upper small intestinal loops were dilated with apparently narrowed terminal ileum, but cecum, appendix and rest of the colon were of normal calibre and there was no evidence of intrinsic obstruction. An end ileostomy proximal to the narrowed segment and distal ileal mucous fistula with biopsy from both the ends of bowel and appendectomy was performed.

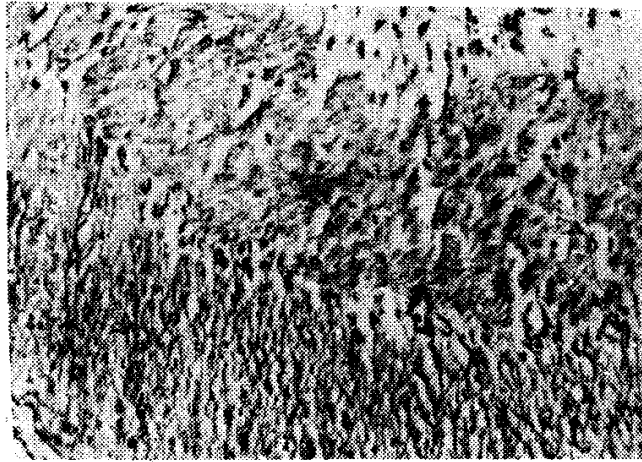
The ileostomy did not function and the child remained obstructed. The histopathology revealed absence of ganglion cells in the appendix as well as both the stomas. An abdominal re-exploration and frozen section biopsy from multiple sites of the small intestine did not reveal any ganglion cells in the proximal intestine. A more proximal stoma was performed which did not function and the child died at 3 weeks of age.

Multiple sections from the entire length of small and large intestine obtained at post mortem showed a total absence of ganglion cells in the myenteric and submucosal regions (*Fig. 1*). Occasional nerve fibres were seen in the myenteric plexus. Section from the duodenum showed a few small group of immature cells in the region of myenteric plexus (*Fig. 2*), but there were no ganglion cells. The stomach showed the presence of fully formed ganglion cells in the myenteric plexus (*Fig. 3*).

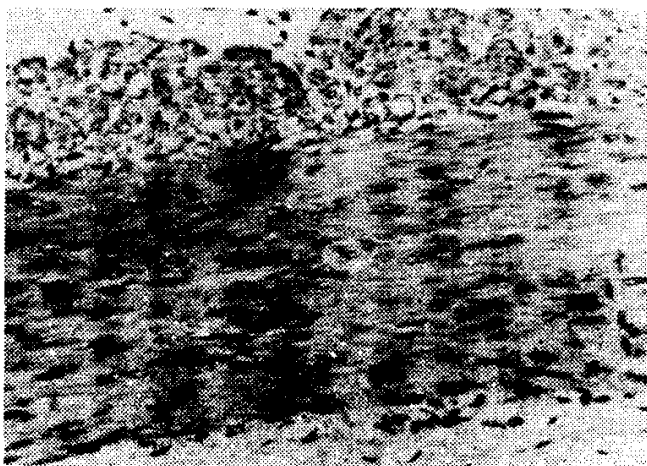
### Discussion

Absence of ganglion cells in the entire intestinal tract represents one of the rare variant of Hirschsprung's disease. The outcome of this extreme form of disease is uniformly fatal(1,2). In 75% of patients, aganglionosis is restricted to the rectosigmoid, in 17% to the sigmoid, splenic flexure or transverse colon, and in 8% to the distal ileum(4,5). Involvement from duodenum to the rectum is a unique form of this disease reported very rarely(1,6-9). One case has been reported with involvement of the stomach(3). Most of the cases present in the early neonatal period with

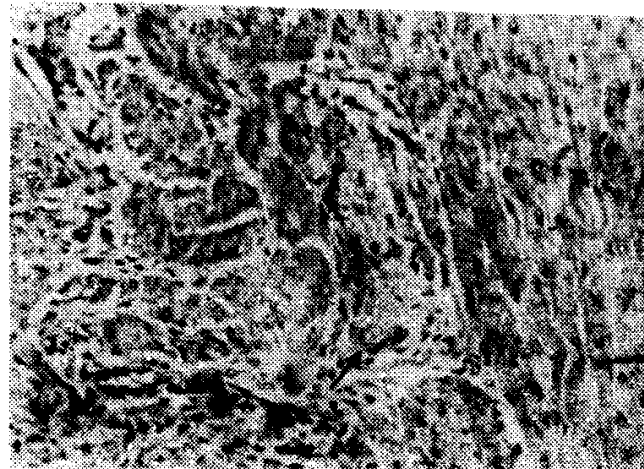
the features of intestinal obstruction. Some of them may pass meconium between 1st to 3rd day of life. The striking findings at laparotomy are absence of intestinal distention and obstruction or a transition zone(1). The small intestine may appear contracted but intraluminal patency is maintained throughout its length. Diagnosis at laparotomy can be substantiated by multiple frozen section biopsy taken from



*Fig. 2. Section of the duodenum showing small groups of lymphocyte like immature cells in the region of myenteric plexus (H & E  $\times 140$ ).*



*Fig. 1. Muscularis propria of the intestine showing a total absence of ganglion cells (H & E  $\times 140$ ).*



*Fig. 3. Microphotograph of stomach showing a mature ganglion cell (arrow) in the myenteric nerve plexus (H & E  $\times 140$ ).*

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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