LETTER TO THE EDITOR

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Long Segment Hirschsprung's Disease with Difficult Feeding Progress

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DEAR SIR

Total colonic aganglionosis (TCA) is a relative uncommon form of Hirschsprung's disease occurring in 2 to 13% of cases and, total colonic and small bowel aganglionosis (TCSA) is an even rarer condition. The former is defined by the absence of neural ganglionic cells from the anus to the ileocecal valve, but no more than 50 cm proximal to the ileocecal valve, while the latter can extend from anus to duodenum. It represents a great challenge for both pediatricians and surgeons not only because it is difficult to diagnose, but also because of pre and post-operative complications.[2]

A term boy born by vaginal delivery (3935g and 50 cm) presented with bilious vomiting and abdominal distension in the first 36 hours of life. After anal stimulus, he passed meconium but the vomiting continued. An orogastric tube was placed for gastric decompression and the child received intravenous hydration. After five days, the abdominal distension persisted; an abdominal X-ray showed dilated intestinal loops with air-fluid levels with suspicious of pneumatosis intestinalis in hypochondrium.

The baby received antibiotics for 12 days along with total parenteral nutrition but abdominal distension persisted. Exploratory laparotomy was performed and biopsies of jejunum, ileum, caecum, ascending colon, transverse colon, descending colon, sigmoid and rectum were taken with suspicion of long segment aganglionosis. Frozen section biopsies revealed absence of ganglion cells in all segments except in the jejunum, confirming the diagnosis of TCSA. Despite the absence of neural ganglionic cells in a segment of the ileum, the surgeon decided to perform a decompressing ileostomy approximately 20cm from the ileocecal valve.

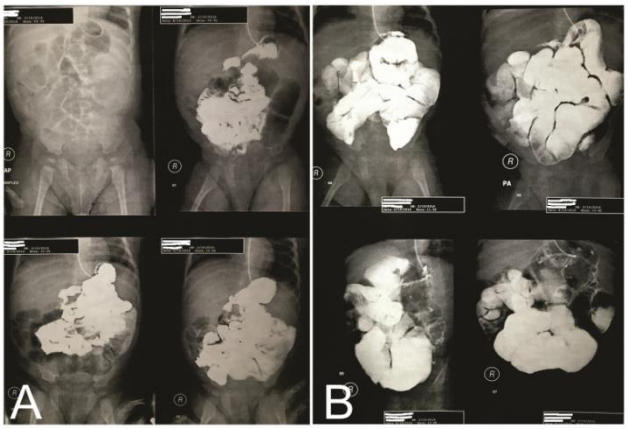

Figure 1. Small bowel follow–through studies. (A) showing was normal progression of contrast through jejunum. (B) showing distended ileal loops full of contrast.

The ileostomy didn't work properly since the beginning, and was emptied daily by saline washouts. On failure to establish enteral feeding, parenteral nutrition was started. Despite four months of hospitalization with treatment for cellulitis of his surgical wound, urinary tract infection by Klebsiella pneumoniae and Enterococcus faecalis and skin colonization by multi-resistant Enterobacter

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cloacae with multiple blood transfusions, the child reached normal nutritional status (7705 grams of body weight). The baby was transferred to a pediatric intensive facility of a different hospital. An abdominal ultrasound showed air-liquid distension in small bowel loops and a sequential small bowel follow-through demonstrated the presence of obstruction due to lack of contrast reaching the ileostomy site (Figure 1).

At redo-surgery, no ileal stenosis was found but a dilated segment followed by narrowed segment of small bowel was observed; the ileal segment near stoma site was also found rotated. A 28cm ileal loop was resected and a more proximal ileostomy was made. The new biopsies confirmed the absent neural ganglionic cells in an ileum segment close to the previous ileostomy, however, presence of regular neural ganglionic cells in the remaining ileum and jejunum were found. The child was further managed with IV antibiotics for a surgical wound infection by Escherichia coli and a central venous line infection by Pseudomonas aeruginosa. During a second period of four-month hospital stay, he had two episodes of enterocolitis managed successfully and was able to tolerate total feeding volume by nasogastric intermittent delivery feeds. Loperamide was instituted to control the high ileostomy output and, cholestyramine for bile-acids skin lesions near the ileostomy. He was discharged with a body weight of 8570 grams and length of 70cm after this last step.

After hospital discharge, he was followed at the paediatric gastroenterology department of a tertiary hospital. He was maintaining weight and height between 10th and 25th percentiles for age and sex on oral feeding. He required few inpatient treatment for episodes of secretory diarrhea and weight loss; and anemia refractory to oral ferrous sulfate and coagulopathy, successfully treated with IV iron and vitamin K treatment.

The typical clinical presentation of short segment Hirschsprung's disease is functional intestinal obstruction shortly after birth.[1] Our case was initially treated as necrotizing enterocolitis which was probably a Hirschsprung's associated enterocolitis. During first surgery, the frozen section could not find ganglion cells in ileum, however, it was found near proximal and mid ileum during second surgery. This reflect limitations of frozen sections biopsy. Moreover, transitional zone was not observed during the first surgery adding further to the difficulty in deciding level of stoma. This finding is in agreement with literature, as has been accurately determined in only 25% of cases.[4]

Surgical treatment of long segment has not yet reached a consensus. [6] A staged approach is commonly performed for long segment aganglionosis and was chosen for our patient.[7] After the child gets older and gains adequate weight, our team expects to retain a patch of left/right colon on the pull through ileum for enhancing water absorption and minimizing fecal fluid loss as a definitive surgery procedure. [8].

Consent: Authors declared that they have taken informed written consent, for publication of this report along with clinical photographs/material, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient however it cannot be guaranteed

Authors' Contribution: All authors contributed equally in concept, design, literature review, drafting the manuscript, and approval of the final manuscript.

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