

Post operatively, oral feeds were started on 1/4th strength pregestemil on the 6th post operative day. Immodium one drop B.I.D. was started on the 12th post operative day, and intravenous fluids were discontinued on the 5th post operative day. He was discharged on 3/4th strength pregestemil and Immodium 1 drop B.I.D. His weight on discharge was 2.8 kgs and the total length of hospitalization was three and a half months. This youngster is doing well three years later.

#### CASE NO. 4

A.F., a three day old male weighing 2.7 kgs., presented with bilious vomiting and abdominal distention. A plain X-ray abdomen showed dilated loops of small bowel and a barium enema showed microcolon. An exploratory laparotomy was carried out, and Type IV ileal atresia was found which was resected and jejuno ileal anastomosis done. The total length of small bowel left was 122 cms. Post operatively he continued to have intestinal obstruction. A re-exploration was carried out three weeks later which revealed dense adhesions between the loops of bowel and there was kinking up of the anastomosis. The adhesions were lysed and the anastomosis untwisted. He still continued to have intestinal obstruction post operatively. Hirschsprung's disease was suspected and two weeks later, a full thickness rectal biopsy was taken. At the same time he was re explored and biopsies from the transverse colon and terminal ileum were taken, and all these were sent for frozen section which revealed aganglionosis in rectal, colonic and ileal biopsies and this confirmed the diagnosis of total colonic aganglionosis. An end jejunostomy was then made. On the 6th post operative day, TPN was started which was continued and he was started on oral feeds of 1/4th strength pregestemil one month later. Due to frequency of stools through the jejunostomy 1 drop Immodium BID was started fifteen days later. The strength of pregestemil was slowly increased. TPN was discontinued at five months of age, and this was when he started slowing steady weight gain. He was discharged at six months of age on 3/4 strength pregestemil and 1 drop BID of Immodium. His weight on discharge was 3.8 kgs. His post discharge course consisted of two admissions one for diarrhea, and the other one for diarrhea with dehydration and metabolic acidosis. This was treated with parenteral fluids. He was also admitted once a formal closure of the gastrostomy, as it was continuously leaking after removal of gastrostomy tube. He is doing well one year following discharge.

#### CASE NO. 5

S.T., a 1 day old female weighing 2.8 kgs. presented with bilious vomiting, physical examination revealed abdominal distention and a small firm mass palpable in the right lower quadrant. Plain X-ray abdomen revealed dilated loops of small bowel. An ultrasound abdomen was unremarkable. A barium enema outlined a microcolon upto the cecum. On exploratory laparotomy, a meconium pseudocyst was found adherent to an atresia of the distal ileum. The pseudo cyst was removed, and ileocecal anastomosis done. The 105 cms. of small bowel which was left behind consisted of jejunum and proximal ileum. Ileocecal valve could not be salvaged.

Post operatively oral feeds were started on 1/4th strength pregestemil on the 7th post operative day. She started to stool frequently, and cholestyramine 150 mg. BID was started on the 20th post operative day. Although the frequency of stools became less, they continued to remain watery and therefore a month after surgery Immodium 1 drop BID was started and the dose of cholestyramine increased to 175 mg. BID. She improved thereafter and was discharged at 39 days

of life. Her weight on discharge was 3 kgs. The post discharge course was complicated by three episodes of diarrhoea at 15 days, 1 month and 3 months respectively. She needed hospitalization for dehydration secondary to diarrhoea, occurring at 1 month after discharge. She is doing well one year post operatively.

#### DISCUSSION :

As in the West, the most common causes of short gut syndrome in neonates remain small bowel volvulus and congenital intestinal atresia<sup>2</sup>. Loss of the ileum is metabolically more significant than loss of the jejunum as the former is the selected site for absorption of intrinsic factor bound Vitamin B-12 as well as conjugated bile salts.<sup>2,3</sup> Two youngsters G.A and A.F among the present group in which only the jejunum was left behind had longer periods of hospitalization. Similarly, the importance of the ileocecal valve was borne out by our experience. Loss of the ileocecal valve increases bacterial colonization of the small bowel, deconjugation of bile salts and reduction of small bowel reabsorption, and shortens the transit time of luminal contents.<sup>3,4</sup> Our patients A.F and S.T. whose ileocecal valves could not be salvaged had difficulty following discharge with recurrent episodes of diarrhoea and dehydration for an initial two to three months.

Due to non availability of parenteral hyperalimentation for the first 4 patients, these infants were handled postoperatively with dilute medium chain triglycerides formula which was gradually increased in volume and concentration. The feeds were begun as early as possible to prevent the structural changes that has been described in bowel unused for a prolonged period<sup>5</sup>. A gastrostomy tube was routinely placed in all infants allowing a continuous drip of formula in the face of intractable diarrhea, to avoid the effects of a bolus of feeds. In infants where a significant part of the ileum were resected, use of cholestyramine to control secretory diarrhoea also proved efficacious, as did the judicious use of Immodium. The most recent infant with short gut syndrome and total colonic aganglionosis was supported with total parenteral hyperalimentation till six months of age. A follow up of the patients one year to three years following discharge reveal all to be showing satisfactory weight gain and development.

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# SHORT GUT SYNDROME : THE AGA KHAN UNIVERSITY HOSPITAL EXPERIENCE.

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## ABSTRACT :

Despite recent advances in post operative care, problems associated with a short gut remain difficult to manage in neonates and infants. Five neonates with short gut syndrome secondary to various congenital intestinal anomalies were managed at the Aga Khan University Hospital between July 1986 and November 1988. The total length of the small bowel ranged from 55 cms to 130 cms and one patient had concomitant total colonic aganglionosis. The period of hospitalization ranged from 36 days to 6 months. Post operative management included the judicious use of Immodium, Cholestyramine and formulas containing medium chain triglycerides. Total parenteral nutrition was employed in one patient. Neonates, with retained ileum and ileocecal valve had smoother post operative courses and shorter hospitalization. All patients are showing satisfactory weight gain and development following discharge.

**Key words :** Short gut syndrome - Parenteral feeding - Cholestyramine - Immodium.

## INTRODUCTION :

The normal neonate is born with 200 to 250 cms. of small bowel. The short gut syndrome is defined as a condition of malabsorption and malnutrition following major resection of small intestine resulting in loss of 50% or more of intestinal length. The introduction of parenteral hyperalimentation by Dudrick in 1968<sup>1</sup> was a major breakthrough in management of short gut syndrome in children. However non-availability and inhibitory cost, when available, of parenteral hyperalimentation solutions pose specific problems in the management of these patients in the developing countries.

## PATIENTS AND METHODS :

Five infants with short gut syndrome were managed at the Aga Khan University Hospital between January 1987 and October 1988. A retrospective review of their cases records forms the basis for this study.

### CASE NO. 1

Q.S., a one day old male infant weighing 2.5 kg. presented

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with bilious vomiting and generalized abdominal distention. A plain x-ray abdomen showed dilated loops of small bowel with multiple air fluid levels. Barium enema revealed a microcolon. An exploratory laparotomy was done, and jejunal atresia and an antenatal perforation with meconium peritonitis was found. Due to the matting up the proximal ileum could not be salvaged. The remaining 45 cms. of jejunum was anastomosed to distal 16 cms. of ileum, that is the total remaining small intestine was 62 cms. of small bowel. The ileocecal valve was intact.

Post operative oral feeds were started with 1/4th strength pregestemil on the 7th post operative day. On the 26th post operative day Cholestyramine was started with a dose on 250 mg. thrice a day. Intravenous fluids were discontinued on the 26th post operative day. After a total hospital stay of 36 days, he was discharged on 3/4th strength pregestemil and 250 mg TID of cholestyramine. On discharge his weight was 2.8 kgs. Since then the post operative course has been unremarkable and he is showing normal growth two and half years later.

### CASE NO. 2

A.Z. a two day old male infant weighing 2.5 kg. presented with bilious vomiting and progressive abdominal distention. A plain X-ray abdomen showed dilated loops of small bowel with air fluid levels. A barium enema was deferred because of gross abdominal distention. Laparotomy findings were of a midgut malrotation and volvulus with gangrene and perforation in the distal jejunum and proximal ileum. After reducing the volvulus and lysing Ladd's bands, the non viable small bowel was excised and an jejuno-ileal anastomosis done. The remainder was 130 cms. of small bowel consisting of proximal jejunum and terminal ileum. The ileo-cecal valve was intact.

Post operative oral feeds were started with 1/4th strength pregestemil on the ninth post operative day. Cholestyramine 250 mg. tid was started on the 24th post operative day. Intravenous fluids were discontinued on the 29th post operative day and he was discharged on full strength pregestemil and cholestyramine 250 mg. tid, the total length of his stay being 37 days. His weight on discharge was 3.6 kgs. He is doing well a year and a half later.

### CASE NO. 3

G.A., a two day old male weighing 1.6 kg. presented with bilious vomiting and abdominal distention. A plain X-ray abdomen showed dilated loops of small bowel and a barium enema outlined the large bowel and distal 20 cms. of ileum. An exploratory laparotomy revealed antenatal ileal perforation and meconium peritonitis with matting up of loops of small bowel. A large portion of nonviable ileum was resected and 53 cms. jejunum was anastomosed to distal 2.5 cms. of ileum. The total length of small bowel was 55.5 cms. The ileocecal valve was intact.