An Update

SHORT BOWEL SYNDROME IN CHILDREN

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ABSTRACT

Short bowel syndrome, a devastating outcome after major intestinal resection, manifests as malabsorption and diarrhoea. Improved outcome in recent years has come about with better understanding of the disease and its sequelae. Medical therapy using H₂ receptor antagonists, loperamide and growth hormone analogues has been found useful. Total parenteral nutrition has revolutionized the outcome of this crippling disorder. Diverse spectrums of surgical procedures aimed at deceleration of intestinal transit and intestinal lengthening have not found favour in clinical practice. Intestinal transplantation remains the only viable option when intestinal adaptation is inadequate and complications of total parenteral nutrition ensue.

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KEY WORDS : Adaptation; Intestinal transplantation; Small bowel syndrome; Small gut syndrome; Total parenteral nutrition.

Introduction

Short gut syndrome in paediatric age group can be a management dilemma. Major intestinal resection may result in diarrhoea and malabsorption, a clinical entity known as short gut syndrome - bigger challenge in the infant or the growing child than an adult. Nutritional prognosis after massive intestinal resection depends on the extent of resection, the nature of the intestinal segment removed (jejunum or ileum) function and adaptive capacity of the remaining intestine and preservation of the colon/ileoceleal valve. Adequate management of this dreadful syndrome requires a multidisciplinary effort between various medical specialties viz. paediatric surgery, intensive care unit, nutritionist, psychologist, paediatric gastroenterologist and transplant immunologist [1].

Aetiological Spectrum

Perinatal causes of loss of massive short gut include multiple intestinal atresias, abdominal wall defects, malrotation with midgut volvulus and segmental volvulus. Other acquired causes with or without predisposing factors include necrotizing enterocolitis, inflammatory bowel disease, abdominal trauma with venous or arterial thrombosis.

Short gut syndrome -its consequence

Significant loss of small intestine results in an inability to maintain nutrition through the residual intestinal tract. A short mucosal contact time and an insufficient mucosal surface area (enterocyte mass) results

in rapid intestinal transit with inadequate digestion and malabsorption.

The metabolic consequences of massive intestinal loss occur as a result of substrate, vitamin and mineral malabsorption. Gastric acid hypersecretion in small gut syndrome has been attributed to hypergastrinemia. Loss of ileum results in hyperoxaluria, vitamin B12 deficiency and malabsorption. Loss of ileocaecal valve results in an overgrowth of colonic bacteria retrogradely into the residual small bowel further compounding the malabsorption state [2].

The neonatal small bowel measures an average of 250-300 cm. Independent survival following resection depends on the length of remaining bowel and the presence or absence of ileocecal valve. No survival is possible with less than 15 cm of small gut. The minimum bowel length for survival is 15 cm with TPN support and 30 cm without TPN support in a setting where ileocecal valve is present. When the ileocecal valve has been lost, 40 cm of small gut is compatible with survival with TPN support and 50 cm without TPN support [3].

Carbohydrate malabsorption and steatorrhoea are common features resulting in diarrhoea and failure to thrive. Malabsorption primarily results from decreased mucosal surface but other factors including bile acid deficiency and bacterial overgrowth also contribute to problems of malabsorption.

Loss of distal small bowel is more serious than loss

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of proximal jejunum. The jejunum is relatively incapable of compensating for ileal loss because ileum is the sole site for absorption of bile salts and therefore essential for the normal process of fat absorption and fat soluble vitamins.

Therapy - Medical

As mentioned earlier, the clinical presentation and physiological consequences after massive gut resection depend heavily on segment of bowel removed and extent of resection. The remaining intestine has the capacity to adapt anatomically and functionally and stimulating the adaptation process is a major component of therapy. Medical therapy of diarrhoea consists of H₂ receptor antagonist, loperamide and secretion inhibitors (somatostatin). Enteral nutrition necessitating the use of pre digested formulae like *pregestimil* or elemental diet like *vivonex* are helpful in initiation of the adaptation process. Recent reports suggest use of growth factor as an adjunct may be useful in enhancing intestinal adaptation. The evolution of institutionalized and home TPN has revolutionized the enteral recovery from this crippling syndrome. Notwithstanding the success story of TPN, life threatening vascular, hepatic and metabolic complications can sometimes ensue and then in these circumstances intestinal transplantation remains the only viable therapeutic option. Clinical management of long-term complications like bacterial overgrowth, nutrient deficiency states and TPN induced liver disease are key outcome determinants [4,5,6].

Surgical

Surgical correction of SBS in the past has looked at procedures which result in deceleration of intestinal transit (like vagotomy or pyloroplasty, segmental reversal of small intestine, recirculating intestinal loops, colonic interposition, constitution of sphincters and valves) and intestinal lengthening procedures [7,8]. Despite enormous literature on these procedures their utility in clinical practice remains to be established, even today.

Autologous gastrointestinal reconstruction by augmenting blood supply of an isolated bowel segment, sequential intestinal lengthening and composite bowel tubes are an exciting new era of experimental surgical successes in this decade [9].

In contrast to other solid organ transplantations, slow progress has been seen in introducing intestinal transplantation from an experimental level to clinical practice [10]. In the last 9 years less than 200 transplantations have been performed worldwide with a 3

year survival of approximately 50% [11,12].

The main problem of intestinal transplantation is immunological. Large amount of lymphatic tissue transplanted along with the intestinal grafts, increases the risk of acute rejection and necessitates high doses of immunosuppressive regimes liable of inducing serious side effects. The immunocompromised recipient is vulnerable to various infections, particularly CMV enteritis. Improved immunosuppression by a combination of drugs like tacrolimus and mycophenolic acid may however change the entire spectrum of immunosuppressive drug therapy. Emphasized antiviral prophylaxis and treatment, improved preservation and prevention of ischaemia-reperfusion injury are other means presently available to obtain better results after intestinal transplants in the near future. Functional grafts resulting in gastrointestinal autonomy being the objective, the next century may well see this dream fast become a viable reality.

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