Extended Small Bowel Hirschsprung's Disease

Extended small bowel Hirschsprung's disease (ESBHD) is a considerable challenge with a high mortality rate (53%), despite the improvements seen in treatment modalities. Long-term total parenteral nutrition (TPN) is required in all cases. As the length of the left over ganglionated SB must be a compulsory point, the surgical treatment become very complex and tedious. The patient also has to face short bowel syndrome following the surgery. For jejunoileal disease with less than 40 cm ganglionated small bowel, Kimura-Stringel's procedure and extended myotomy-myectomy is adapted, but no operative technique was found to have a superior outcome. Currently, following unsuccessful attempts of intestinal salvage, small bowel transplantation or liver intestinal transplantation is used. Survival has been reported to be improved through the recent years and is currently > 80% after one year and > 50% following 5 years.

A 5 days old baby girl delivered after a 34-week gestation with a birth weight of 2.44 kg, presented in a private hospital with complaints of reluctance to feed since birth and delayed passage of meconium, previously she was admitted somewhere else for above complaints and due to suspicion of duodenal stenosis, surgery was advised but parents left against medical advice. There was no history of birth asphyxia. On examination, vitally stable active baby with slightly protuberant abdomen more in the upper part was seen. On digital rectal examination, the baby passed small amount of stool. All the systemic examination were unremarkable. Her plain X-ray abdomen showed gut loops upto the pelvis. General management of intestinal obstruction was started. Upper GI contrast study revealed dilated jejunal loops with persistence of contrast on delayed films. With provisional diagnosis of ileal stenosis, patient underwent laparotomy.

Per-operatively, distal to ligament of Treiz (50 - 60 cm) length of jejunal loops were found moderately dilated while, the ileal loops were tapered and the caliber of gut decreased to a pencil tip. Distal lumen patency was checked and multiple biopsies including appendectomy and rectal biopsy were taken. Stoma was fashioned in the most distal dilated jejunal loop. Biopsy report was indicative of absence of ganglion cells from ileum to rectum. Baby was kept on TPN. Counselling about poor prognosis and future treatment options were explained to parents. The case was referred to higher centres; parents consulted different consultants and after hearing the same prognosis, they gave up hope and became reluctant for surgery. The conservative treatment continued. Eventually the child developed sepsis and succumbed to it at 2-1/2 months of life.

There is a suggestion that there should be a strong strategy at the national level to deal with such rare cases as a large sum is required for prolonged hospital stay and the cost of TPN. Finally, the complex surgical procedures make the survival of this disease an unfulfilled dream in a poor set-up like Pakistan. Every case should be recorded so that their incidence can be analyzed and a specialized local treatment protocol might be developed. Even patients with rare diseases should be facilitated for definite treatment abroad to good centres in the world involved in the treatment of ESBHD. For patients with rare diseases having 100% mortality and with no treatment options here in Pakistan, such facilities are needed on humanitarian and sympathetic grounds.

REFERENCES

Yaqoot Jahan

Correspondence: Dr. Yaqoot Jahan, Paediatric Department, Dow University of Health Sciences and Civil Hospital, Karachi. E-mail: iqabyj47aug@yahoo.com

Received May 25, 2011; accepted May 09, 2012.