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Indian Journal of Pharmacy and Pharmacology

Journal homepage: <https://www.ijpp.org.in/>

Case Report

A case report on long segment hirschsprung's disease

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ARTICLE INFO

Article history:

Received 29-04-2021

Accepted 27-08-2021

Available online 04-09-2021

Keywords:

Hirschsprung disease

Intestinal obstruction

Phenotypes

Genetic predispositions

ABSTRACT

Hirschsprung disease is a complex congenital condition of the intestine, usually recognized to be genetic origin and reverberates from a disturbance of the normal development of the enteric nervous system. It is evident to be the most common cause of a low intestinal obstruction in the neonates as well as the older children. Study reveals that occurrence rate of cases in 70% may be associated with other congenital abnormalities together with number of syndromic phenotypes. A number of distinct genetic sites have been identified in these syndromic phenotypes, identifying potential genetic predispositions. Presenting hereby a case of neonatal hirschsprung's disease whereby the patient was surgically corrected along with stepwise pharmacological therapy and was able to get discharged with regular follow up advice.

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1. Introduction

Hirschsprung disease (HSCR) is defined as a functional intestinal obstruction that betides due to the congenital deficiency of the normal myenteric plexus parasympathetic ganglion cells which are located in the distal portion of the large intestine.

Hirschsprung disease is characterized by aganglionosis (absence of ganglion cells) in the distal colon and rectum. It is believed to either occur from a failure of neuroblasts in neural crest cells to migrate into bowel segments or already migrated neuroblasts degeneration. Both the cells in the myenteric and submucosal plexuses get affected. Hence, functional obstruction develops as a result of a spasm in the denervated colon.

Treatment of Hirschsprung's disease involves surgery which includes bypassing the part of the colon that's lacking nerve cells. There are two ways this can be done: a pull-through surgery or an ostomy surgery.¹ In pull-through

surgery, the lining of the diseased part of the colon is withered away followed by pulling the normal section through the colon from the inside and attaching it to the anus. However, under ostomy surgery, for the Children's who are sick surgery is usually done utilizing minimally invasive (laparoscopic) methods, operating through the anus. Surgery usually involves two steps; Firstly, the abnormal portion of the colon is removed and the top healthy portion of the colon is connected to an opening the surgeon creates in the child's abdomen causing stools to leave the body through the opening into a bag that attaches to the end of the intestine that protrudes through the hole in the abdomen (stoma). This allows time for the lower part of the colon to heal. Once the colon has had time to heal, a second procedure is done to close the stoma and connect the healthy portion of the intestine to the rectum or anus.²

2. Case History

1 Year old developmentally normal male child was diagnosed to have hirschsprung disease at neonatal period for which he underwent colostomy. In august, he had

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perineal pull through surgery and revealed biopsy of sigmoid colon which showed absence of ganglion cells.

On November he was admitted for multiple biopsy for colon and later admitted for further management. After pre operative investigation, including Covid-19, due to ongoing pandemic situation (reported negative), informed consent from the parents bought and after pre anesthetic evaluation, the child underwent Redo Duhamel Extended Pull Through for long segment Hirschsprung disease on (November 2020). Post operatively child kept in nil per mouth. Since child presented with fecal stained drain, he was continued on NPO and was started on TPN; close monitoring of vitals and abdominal girth were done. USG abdomen screening done twice for any intrapelvic collection showed no significant collection. Blood investigation showed elevated total count and elevated ESR, Child started on intravenous Meropenem after getting infectious disease opinion. On post operative day 9, child noted to have main wound bulge and on opening up, fecal discharge came. So that child taken for emergency diversion ileostomy on December 2020. Post operatively child shifted to HDU Unit for further management and 1 unit of fresh frozen plasma was given. On post op day 2, patient was shifted to ward and was continued on intravenous fluids support, TPN drain replacement and started on oral feeds. Intravenous and TPN stopped on November 2020 and started on high protein diet. Drain was removed; ileostomy bag care was taught and child was hemodynamically stable. Hence was discharged with further care advice.

3. Discussion

The occurrence rate of Hirschsprung's disease according to WHO is approximately 1 per 4000 live births. Studies reveal that this disease prevails more commonly in males than females. Mostly children get diagnosed during the very first 2 years of their life.³ Genetic predisposition also surprisingly prevails in larger numbers affecting the severity. Diagnosis of Hirschsprung's disease requires a histological verified absence of ganglion cells in either biopsy or surgical colonic specimens.

The definitive treatment of most HD patients as discussed earlier in the introduction part involves surgical procedure, it is a paramount factor to decompress the abdominal distension in order to prevent HAEC by gentle rectal washout during the waiting time for surgery, in cases of HAEC in addition to the rectal washout, patients will require broad spectrum antibiotics, intravenous fluids is required if a patient kept NPO and is a supportive measure throughout surgery.⁴ Severe cases of HAEC usually employ diversion colostomy. Care should be taken that HAEC cases don't get misdiagnosed as cases of gastroenteritis to avoid serious consequences. Here the patient undergone redo Duhamel extended pull through for

long segment Hirschsprung's disease as a tertiary surgery. He was primarily done colostomy followed by perineal pull through surgery. Redo pull-through operations for Hirschsprung's disease seems to be as effective as primary procedures in terms of continence and stooling frequency. The postoperative period in HDTCA is characterized by surging death rates, including food intolerance, electrolyte disturbances, and dehydration due to exacerbated intestinal secretion. Taking into account these complications prior to surgery decreases death rate, but in the long run, there is greater risk of stoma permanence for long periods in these patients.⁵ In the present case, the analysis of the proximal segment indicated hypoganglionosis, characteristic of the transition zone, which may explain the requirement for a second intervention. Here the patient got discharged with hemodynamically stable condition with advice to regular follow up.

Pharmacological treatment involves just supportive and symptomatic treatment approach. For patients developing enterocolitis- nasogastric decompression, intravenous fluids, antibiotics, and colonic lavage may be necessary. Few studies also suggest that sodium cromoglycate, a mast cell stabilizer, has also been reported to beneficialise these patients.⁶ Botulinum toxin injections within the contracted internal sphincter mechanism have been reported to induce more normal patterns of bowel movements in postoperative patients with obstruction or enterocolitis.

Postoperatively, patient receives IV fluids and broad spectrum antibiotic according to hospital framed protocol. Until the passage of flats & stool from the stoma post-op, nothing will be administered by mouth. Patient only gets discharged from hospital upon attaining full feedings, as here in this case patient did so within 72 hours of surgery. Keeping in mind the long term complications of surgery including intermittent enterocolitis, severe stool retention as well as intestinal obstruction, child was advised to be taken utmost care of with regular screening for their bowel habits as an outpatient monitoring. Prolonged laxative treatment may be usually required for patients in whom hypo motility is common post surgery.

4. Source of Funding

None.

5. Conflict of Interest

None.

6. Acknowledgment

We would like to thank Department of Pediatric Surgery for their immense help providing with the case details whenever required. We would also like to thank Apollo Children's Hospital in allowing us to publish this case report.

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doi:10.1007/BF02049889.

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Cite this article: Suganya D, Kannan R, Priya A, Arun Chander Yadav K. A case report on long segment hirschsprung's disease. *Indian J Pharm Pharmacol* 2021;8(3):235-237.