Case Report

The Alternative Treatment of Intestinal Neuronal Dysplasia in Newborn Period: A Case Report

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Intestinal neuronal dysplasia is one of the most commonly encountered variant of Hirschsprung's disease but also in that untreated cases may prove fatal. The authors describe the operative treatment for a newborn who was diagnosed with long segmental intestinal neuronal dysplasia, as the remains of the normal bowel was no more than 35 centimeters from the DJ junction. The side-to-side enteroenterostomy of the affected intestine with Santulli enterostomy could rescue the baby from parenteral nutrition, regaining normal oral feeding.

Keywords: *Intestinal neuronal dysplasia, Neuronal intestinal dysplasia, IND, NID*

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Intestinal neuronal dysplasia (IND) is an inherited disease of the intestine that affects the rectum, colon, or the whole intestine due to complex alteration of the enteric nervous system and leads to functional intestinal obstruction. IND was first described by Meier-Ruge in 1971 as a malformation of enteric plexus and it can often be confused with Hirschsprung's disease[1,2]. The incidence of isolated IND varies from 0.3% to 40% of all suctional rectal biopsies in different centers[3]. The incidence varies considerably among different countries. Recently, there are many therapeutic efforts including medical treatment, sphincter operation, and resection of the affected intestine. However, results and follow-up reports have been described in very few cases and had limited success. Moreover, IND has rarely been reported in the neonate; therefore, this article describes the operative management in a newborn whose diagnosis was histologically confirmed as the long affected segment of IND.

Case Report

A 3-day old Thai baby girl was referred to the hospital with history of failure to pass meconium with abdominal distention. She was fully term with no other problems after birth.

The physical examination demonstrated a low grade fever and bilious content was detected in the orogastric tube. The abdomen was mildly distended, soft, and there was no redness of the abdominal wall seen. The anus was in its normal position. Rectal examination revealed a narrow anal canal with empty rectum and there was white mucous passing after per rectal examination. Plain abdominal radiograph was suggestive of intestinal obstruction (Fig. 1). After a single attempt of rectal irrigation, the abdominal distension failed to resolve; therefore, the patient underwent a contrast enema followed by fluid resuscitation and administration of intravenous antibiotics. The water soluble contrast enema showed a microcolon (Fig. 2). The exploratory laparotomy was performed, which showed marked dilatation of the small intestine from the ligament of Treitz to 35 centimeters of the proximal jejunum without any mechanical obstruction (Fig. 3). Consequently, serial full thickness intestinal biopsies with loop jejunostomy were performed. The pathology report indicated the presence of ganglion cells in all specimens and the final diagnosis of IND was determined. After the operation, the patient suffered from dehydration and electrolyte imbalance due to high output enterostomy (more than 1 mL/kg/hour); thus, parenteral nutrition was provided during the time until the next operation.

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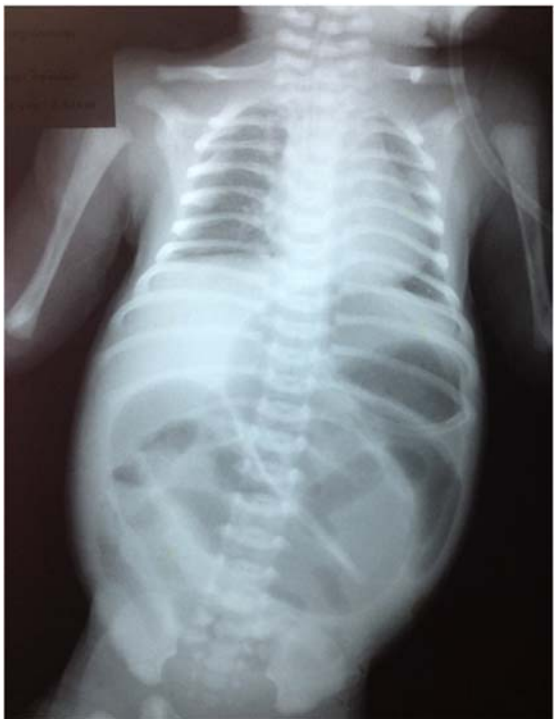

Fig. 1 The plain abdominal radiograph scout view showed small bowel dilatation in disparate sizes and no air passage in the colon.

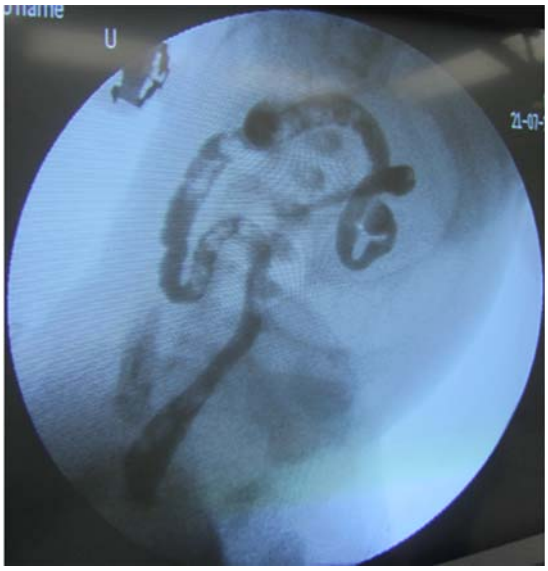

Fig. 2 A microcolon or unused colon was demonstrated in the contrast enema study.

At 2 months of age, the patient was taken to the second operation in order to widen the affected intestine which was still small (Fig. 4) by using the side-to-side enteroenterostomy. The operative procedure was performed by dividing the affected intestine (approximately 30 cm long each) into two segments and each segment was divided in a longitudinal plane. After that, these segments were patched together with preservation of the mesenteric blood supply to both portions of the intestine. The distal end of it was opened as an ileostomy. In addition, the previous proximal jejunostomy has remained as Santulli enterostomy (Fig. 5) in order to decompress the proximal intestine in the early postoperative period. Enteral feeding was started on postoperative day 7th then full oral feeding was attained approximately on postoperative day 26th and the parenteral nutrition was sustained. After 2 months of the procedure, the Santulli enterostomy was closed after the content was gradually reduced. Subsequently, the content can pass into the distal bowel before adaptation to normal stools was seen at the ileostomy. Therefore, the functional ileostomy is a good indicator and implied that the patient is well prepared for the pull-through operation in the future.

Discussion

Intestinal neuronal dysplasia (IND) is a disease of the enteric nervous system, which clinically resembles Hirschsprung's disease. Various terms have been used to describe these conditions: chronic idiopathic intestinal pseudo-obstruction, pseudo-Hirschsprung's disease, neonatal intestinal pseudo-obstruction, and intestinal hypoperistalsis syndrome[1,2]. Until recently, it is not well-defined whether IND is a congenital malformation or an acquired secondary condition related to some gastrointestinal problems.

The clinical characteristic of IND can be found in infants with a history of constipation and abdominal distention that mimic Hirschsprung's disease. Symptoms associated with IND were reviewed by Csury and Pena[4] who analyzed reports of 278 patients. The most frequently described symptom was constipation. Constipation was reported in 53%, obstruction in 20%, colitis in 12%, and bloody stool, diarrhea, or vomiting in less than 10% each. However, no pathognomonic symptoms for any specific neuronal intestinal disorder were identified in 203 patients by Ure and Holschneider[1]. Neither the radiological study nor anorectal manometry can be used to diagnose IND specifically. However, Montedonico et al[5] had mentioned a significantly higher rate of the absence of the internal sphincter relaxation in

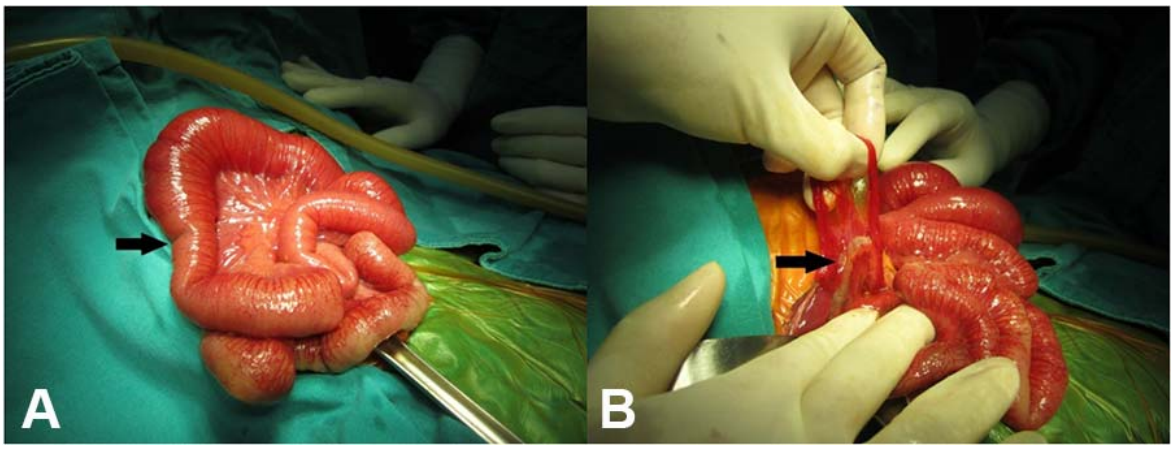

Fig. 3 A.The operative view identified the transitional zone (Black arrow) at upper jejunum, 35 centimeters from the ligament of Treitz. B.The colon (Black arrow) was in a very small caliber, less than 1 centimeter in diameter, consistent with a microcolon.

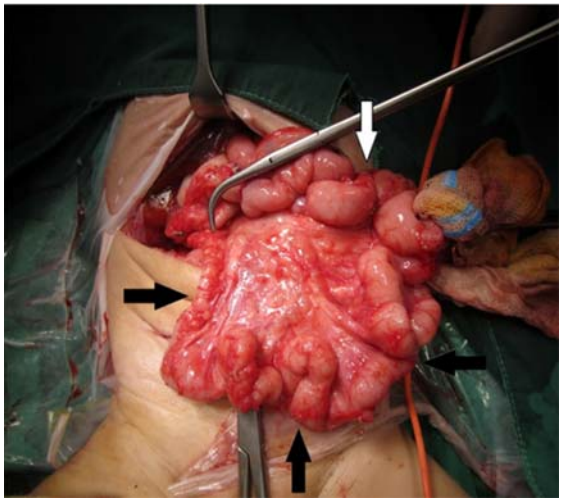

Fig. 4 The second operative view, the white arrow denotes the normal proximal bowel compared to the affected small intestine which was apparently small (black arrow).

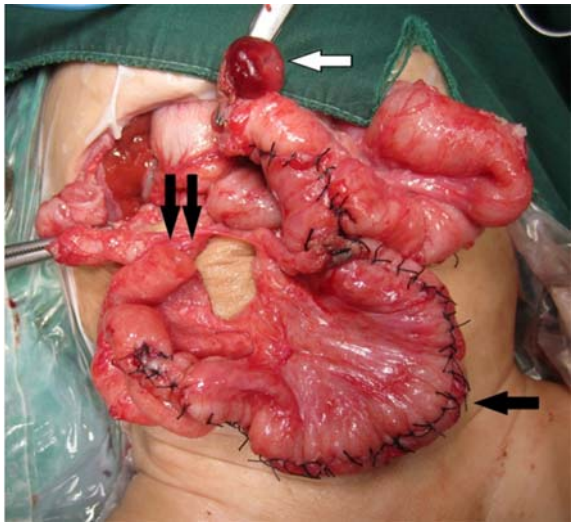

Fig. 5 The side-to-side enteroenterostomy procedure was performed by two segments of the affected intestine patched together (black single arrow) and a safety vent created with the Santulli enterostomy (white arrow) during the waiting period for the definitive operation. Note the double black arrows marked the distal enterostomy or ileostomy.

manometric presentation in the severe IND group but there was no pathognomonic pattern of the relaxation reflex; the findings could be either atypical or normal.

The principal methods employed for the diagnosis in disorder of intestinal innervation are the rectal biopsy, as including hematoxylin and eosin (H-E) and AChE histochemical staining[1,2]. Hyperplasia of the submucous plexus, giant ganglia or hyperganglionosis and increased AchE activity are the most important features for the diagnosis of IND. In the meantime, the neuronal markers and immuno-histochemical technique currently are being used to display the enteric nervous system to confirm the diagnosis[1,3,6].

Current treatments of IND have been subjected to medical management with laxatives and enemas. The operative interventions were used if the intestinal symptoms persist after at least 6 months of treatment, including internal sphincter myectomy and resection of the affected intestine with a pull-through

operation. The results obtained with these multiple treatments have been reported by Gillick et al[7], including a review of 33 patients with IND for the period ranging from 1 to 8 years. Twenty-one (64%) patients had good response to conservative treatment and currently have normal bowel habits. Twelve patients (36%) underwent internal sphincter myectomy after failed conservative management. Two patients were able to stay clean with regular bowel habits, whereas three patients, who continued to have persistent constipation after myectomy, consequently underwent resection of the redundant and dilated sigmoid colon. It could be said that IND is a distinct clinical entity, which had completely different outcomes.

In this case, the patient exhibited the clinical of intestinal obstruction at birth and the long segmental IND was definitively diagnosed. This entity was a rare condition and seldom reported in the neonatal period. The patient suffered from the initial management including dehydration and electrolyte imbalances because of the high output proximal enterostomy locating very close to the ligament of Treitz. The second surgical outcome of the side-to-side enteroenterostomy of the affected intestine with the proximal Santulli enterostomy was undoubtedly efficient. The patient was able to wean from parenteral nutrition and regain full enteral feedings. Finally, the patient can tolerate oral nutrition while continue to have normal growth. However, the prognosis in this case should be followed-up and monitored regarding growth and development in a long-term basis after the definitive surgery.

In conclusion, the side-to-side enteroenterostomy of the affected intestine with the Santulli enterostomy can be the alternative procedure to perform in the neonate who was diagnosed the long segmental IND. In addition, this surgical technique enables recovery of the intestinal continuity and decreases the complications of long-term parenteral nutrition during the waiting period for the definitive operation. Subsequently, the definitive surgery, including a pull-through procedure should be performed. However, the operation is technically demanding and carries significant morbidity.

Documentation of the IRB or Animal Care committee approval

The project entitled "The alternative operative treatment of the intestinal neuronal dysplasia in a newborn: A case report" has been reviewed by The Khon Kaen Hospital Committee for Human Research, based on the declaration of Helsinki (Protocol number: 26/01/2556).

What is already known on this topic?

Intestinal neuronal dysplasia (IND) is an inherited disease of the intestine that affects the rectum, colon, or the whole intestine due to the complex alteration of the enteric nervous system and leads to functional intestinal obstruction. IND patients have been subjected to medical management with laxatives and enemas as the primary treatments. If the intestinal symptoms persist at least 6 months after using medication, the operative interventions have been used for further treatment including internal sphincter myectomy and resection of the affected intestine with a pull-through operation.

What this study adds?

The side-to-side enteroenterostomy of the affected intestine in the long segmental IND with the Santulli enterostomy can be the alternative procedure to perform in the neonate. This surgical technique enables rapid recovery of the intestinal continuity and decreases the complications of long-term parenteral nutrition during the waiting period for the pull-through procedure.

Potential conflicts of interest

None.

References

1. Holschneider AM, Puri P, Homrighausen LH, Meier-Ruge W. Intestinal neuronal malformation (IND): Clinical experience and treatment. In: Holschneider AM, Puri P, editors. Hirschsprung's disease and allied disorders. 3rd ed. New York: Springer-Verlag Heidelberg; 2008: 229-51.
2. Puri P. Intestinal dysganglionosis and other disorders of intestinal motility. In: Adzick NS, Krummel T, Laberge J-M, Shamberger R, Caldamone A, editors. Pediatric surgery. 7th ed. Philadelphia: Saunders; 2012: 1279-87.
3. Puri P. Variant Hirschsprung's disease. J Pediatr Surg 1997; 32: 149-57.
4. Csury L, Pena A. Intestinal neuronal dysplasia: Myth or reality? Pediatr Surg Int 1995; 10: 441-6.
5. Montedonico S, Acevedo S, Fadda B. Clinical aspects of intestinal neuronal dysplasia. J Pediatr Surg 2002; 37: 1772-4.
6. Kobayashi H, Hirakawa H, Puri P. What are the diagnostic criteria for intestinal neuronal dysplasia.

Pediatr Surg Int 1995; 10: 459-64.
7. Gillick J, Tazawa H, Puri P. Intestinal neuronal dysplasia:Results of treatment in 33 patients. J Pediatr Surg 2001; 36: 777-9.

ทางเลือกในการรักษาทารกแรกเกิดที่เผชิญกับภาวะบกพร่องของระบบประสาทที่ควบคุมการเคลื่อนไหวของลำไส้: รายงานผู้ป่วย 1 ราย

จุฬาทิพย์ นาคาเริงฤทธิ์, มงคล เลาหเพ็ญแสง

ภาวะบกพร่องของระบบประสาทที่ควบคุมการเคลื่อนไหวของลำไส้ intestinal neuronal dysplasia (IND) มีผลทำให้ลำไส้ใหญ่หรือลำไส้ทั้งหมดเกิดความผิดปกติ โดยลำไส้จะสามารถหดรัดตัวได้แต่จะไม่มีการคลายตัว ส่งผลให้สิ่งบรรจุภายในลำไส้ไม่สามารถผ่านไปได้หรือผ่านได้ด้วยความยากลำบาก เกิดการอุดกั้นของทางเดินอาหารบางส่วนหรืออย่างสมบูรณ์ ผู้ป่วยที่มีภาวะนี้จะเกิดภาวะอุดกั้นของทางเดินอาหารถ่ายอุจจาระลำบากหรือท้องผูกเรื้อรัง

วิธีการผ่าตัดที่ใช้ในการรักษาทารกแรกเกิดที่ได้รับการวินิจฉัยว่ามีภาวะบกพร่องของระบบประสาทที่ควบคุมการเคลื่อนไหวของลำไส้ ในงานวิจัยนี้คือ side-to-side enteroenterostomy ในส่วนของลำไส้ที่ผิดปกติร่วมกับการยกลำไส้ไว้ที่ผนังหน้าท้อง (santulli enterostomy) โดยผู้ป่วยรายนี้เหลือลำไส้ที่ปกติเพียง 35 เซนติเมตร ซึ่งภายหลังการผ่าตัดผู้ป่วยสามารถรับประทานนมได้ตามปกติทางช่องปากและไม่จำเป็นต้องได้รับสารอาหารทางหลอดเลือดในที่สุด

วิธีการผ่าตัดที่ใช้ในการรักษาทารกแรกเกิดที่มีภาวะบกพร่องของระบบประสาทที่ควบคุมการเคลื่อนไหวของลำไส้ ในงานวิจัยนี้ได้ผลการรักษาดี ดังนั้นวิธีการผ่าตัดนี้น่าจะสามารถนำมาใช้เป็นทางเลือกในการรักษาเพื่อช่วยลดภาวะแทรกซ้อนจากการให้สารอาหารทางหลอดเลือดได้