How to Manage Low Gut Obstruction in Neonates with Immature Ganglion Cells in the Colonic Wall?

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Background: *Neonates with immature ganglion cells in the colonic wall may have the clinical picture similar to Hirschsprung's disease, especially total colonic aganglionosis. Management of this entity depends on the judgment of each clinician.*
Objective: *The aim of this study was to review management of clinical low gut obstruction in neonates with presence of immature ganglion cells in the colon.*
Material and Method: *A retrospective study of neonates with clinical low gut obstruction due to presence of immature ganglion cells treated between 2007 and 2012 was reviewed.*
Results: *Six patients, one term and 5 pre-term neonates, were proven to have immature ganglion cells in the colonic wall. They presented with delay or failure to pass meconium, progressive abdominal distension and bilious vomiting after birth. Abdominal films showed generalized small bowel dilatation and barium enemas revealed a microcolon in 4 of them. They underwent laparotomy between 4 and 11 days of life. A microcolon with a transitional zone (Tz) was seen at the terminal ileum, 30-75 cm proximal to the ileocecal valve. Colonic biopsy and the appendix revealed presence of immature ganglion cells and ileal biopsy at the Tz showed presence of normal ganglion cells. An ileostomy was performed at the Tz in all of the 5 premature neonates, while an enterostomy was not done in term infant. Closure of the enterostomy in the 5 premature cases was performed after they had been proven to have mature ganglion cells in the colonic wall by a rectal biopsy after the age of 3 months. All of the 6 cases were doing well on the last follow-up between 1 and 3 years.*
Conclusion: *Functional low gut obstruction in neonates caused by immaturity of the colonic ganglion cells should be managed by laparotomy including biopsies of the colon, appendix and terminal ileum with enterostomy at the Tz. Closure of the enterostomy is done after presence of mature ganglion cells proven by a rectal biopsy after 3 months of age. Full-term neonates with immature ganglion cells in the colonic wall may be successfully managed conservatively without enterostomy.*

Keywords: *Allied disorders of Hirschsprung's disease, Total colonic aganglionosis, Immature ganglion cells, Functional gut obstruction*

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Hirschsprung's disease (HD) is one of the most common causes of neonatal intestinal obstructions. This entity is characterized by congenital absence of ganglion cells in the nerve plexus within the wall of the rectum and distal colon. It is associated with dilatation of the normal proximal colon due to a functional obstruction. The aganglionic segment may extend more proximally into the descending and transverse colon. Sometimes, the aganglionosis involves the entire colon and even the terminal ileum, which is called total colonic aganglionosis (TCA). The disease of colonic aganglionosis was first described by Herald Hirschsprung[1,2] of Copenhagen in 1886. The presentation of a curative operative technique was proposed by Swenson and Bill[3] in 1948. Zuelzer and Wilson[4] reported the first two examples of TCA in 1948 and Sandegard[5] presented the successful treatment of an infant with TCA in 1953.

The principal diagnosis of HD is dependent on the absence of ganglion cells just proximal to the spastic or aganglionic segment and with the presence of ganglion cells in all of the dilated or ganglionic bowels. This phenomenon is called "all or none law",

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as described by Hirschsprung. For this reason, a rectal biopsy is generally accepted to be the definite diagnosis of HD[6].

A minority of neonates, who have typical clinical presentations and radiological findings like Hirschsprung's disease, are proven to have ganglion cells from rectal biopsies. These ganglion cells show abnormal features such as hypoganglionosis, neuronal immaturity (immature ganglion cell), and intestinal neuronal dysplasia. Holschneider[7] categorized these abnormalities into allied disorders of Hirschsprung's disease. The authors have some experience in the case of premature neonates who developed clinical features of low gut obstruction, caused by immature ganglion cells in the entire colonic walls. We, herein, reviewed all of our patients in order to formulate a treatment guideline for this condition.

Material and Method

Medical records of the patients with Hirschsprung's disease and its allied disorders treated at Queen Sirikit National Institute of Child Health (QSNICH) and Samitivej Srinakarin Hospital (SNH) between 2007 and 2012 were reviewed. The study was begun after approval of the research proposal by the Institutional Review Board (Document No. 57-010). We focused on the patients who had a diagnosis of low gut obstruction and a pathological report of immature ganglion cells in the colonic wall. Demographic data, clinical presentations, radiological findings, operative procedures and results of the treatment were studied in details.

Results

Between 2007 and 2012, 145 patients were treated with classical Hirschsprung's disease and 12 patients with TCA at QSNICH, whereas 5 neonates (3.2% of HD and TCA) were treated with functional low gut obstruction due to immature ganglion cells in the colonic wall. Our one case with immature ganglion cells and low gut obstruction from SNH was included in the present study.

Of the 6 neonates with immature ganglion cells, 2 were males and 4 were females. They were born at the gestational age ranging from 27 to 38 weeks and their birth weights ranged from 1,040 to 2,750 grams (Table 1). One set of twins was noted in the study. Four cases were born by vaginal delivery, whereas two cases were born by cesarean section. There was no history of maternal magnesium sulfate administration during delivery in any of the cases.

All of the neonates presented with delayed or failure to pass meconium in the first 48 hours of life, progressive abdominal distension (Fig. 1) and bilious vomiting. Abdominal films revealed generalized small intestinal dilatation that was compatible with low gut obstruction (Fig. 2). All of the patients received conservative treatment with NPO, nasogastric tube decompression, parenteral nutrition and antibiotics. None of the babies showed any improvement after rectal irrigation with normal saline solution (NSS). Two neonates underwent laparotomy without contrast study, while 4 neonates underwent a further investigation with barium enema (BE). BE showed a microcolon in all of the 4 neonates (Fig. 3) and shortening of the sigmoid colon in 2 neonates. All of the patients underwent laparotomy with the pre-operative diagnosis of TCA.

Case No. 1, Twin A, was operated on the sixth day of life without frozen section for immediate pathological examination at the Eastern Regional Hospital. Operative findings revealed a microcolon with a transitional zone (Tz) at the terminal ileum, 30 cm above the ileocecal (IC) valve. A loop ileostomy was performed just above the Tz and a biopsy was also obtained from the right transverse colon and ileostomy site. The pathological report showed presence of immature ganglion cells at the right transverse colon and numerous normal ganglion cells at the ileostomy site. He was followed-up at the regional hospital and was eventually referred to QSNICH at the age of 5 months. Full thickness rectal biopsy was repeated at QSNICH. Pathological report revealed both mature and a few immature ganglion cells in the muscular layer of the rectum. The ileostomy was closed at the age of 7 months. The postoperative course was uneventful and the patient could pass stools normally. He was doing well at the 3-year follow-up.

Case No. 2, Twin B, underwent laparotomy at the age of 9 days at the same hospital as Case No. 1. A microcolon and a Tz at the terminal ileum, 40 cm above the IC valve were noted. An ileo-colonic patching was performed by modified Kimura's technique[8] (using 10 cm of the transverse colon patching with the terminal ileum). An ileocolostomy was opened at the right lower quadrant. The pathological report revealed mature ganglion cells at the Tz, whereas immature ganglion cells were noted at the transverse colon. He was referred to QSNICH at the age of 5 months. After obtaining a rectal biopsy, which showed mature ganglion cells in the muscular layer, the ileocolostomy was closed at the age of 7 months. He was doing well and had normal

Table 1. Demographic data, clinical presentations and radiological findings

Case No.	GA (weeks)	BW (grams)	Symptomatology	Plain abdomen	Barium enema
1 M (Twin A)	34	1,960	Delayed passage of meconium, abdominal distension, bilious vomiting	Marked small bowel dilatation with air fluid level	Not done
2 M (Twin B)	34	2,000	Delayed passage of meconium, abdominal distension, bilious vomiting	Marked small bowel dilatation without rectal gas	Not done
3 F	31	1,450	Delayed passage of meconium, abdominal dilatation, bilious vomiting	Generalized small bowel dilatation	Microcolon, shortening of the sigmoid colon
4 F	38	2,750	Delayed passage of meconium, abdominal dilatation, bilious vomiting	Generalized small bowel dilatation	Microcolon, good barium evacuation in the delayed film of 24 hours
5 F	27	1,040	RDS, respiratory failure, absence of meconium passing, marked abdominal distension, marked bilious content from the NG tube, clinical sepsis	Generalized small bowel dilatation without rectal gas	Microcolon, shortening of the sigmoid colon
6 F	35	2,290	Delayed passage of meconium, marked abdominal distension, bilious vomiting	Generalized small bowel dilatation without rectal gas	Microcolon, barium passing upwards to the terminal ileum just proximal 5 cm, to the IC valve

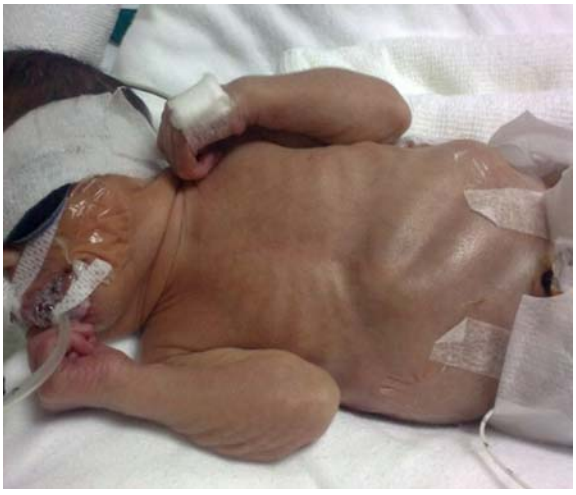

Fig. 1 Marked abdominal distension with visible bowel loops in the Case No. 5.

bowel function at the 3-year follow-up.

Case No. 3 was referred from SNH and underwent laparotomy at the age of 10 days at QSNICH. Operative findings revealed a microcolon with a Tz, 75

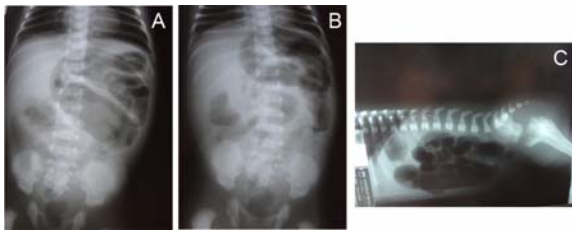

Fig. 2 Abdominal films showed generalized small bowel dilatation (A and B) without rectal gas (C) in the Case No. 6.

cm above the IC valve. All of biopsies from the sigmoid colon, terminal ileum (at the Tz) and appendix revealed presence of immature ganglion cells in the submucosal and intramuscular layers from the immediate frozen and permanent section of pathological examinations. A loop ileostomy was performed, 80 cm above the IC valve. She developed marked watery diarrhea and was diagnosed as short-bowel syndrome. Ileo-ileal patching by modified Kottmeier's technique[9] was done in order to correct watery diarrhea at the age of 3 months. Biopsy of the terminal ileum revealed presence of mature

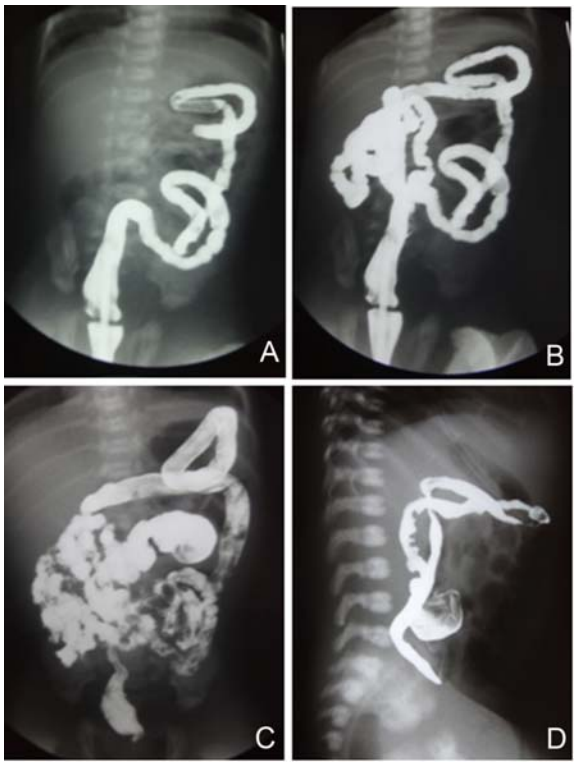

Fig. 3 Barium enema of the Case No. 4 revealed a microcolon and good barium evacuation in the delayed film of 24 hours ((A,B) present a microcolon, (C) immediate post-evacuation film, (D) delayed film of 24 hours).

ganglion cells, whereas biopsy of the transverse and sigmoid colon demonstrated both immature and mature ganglion cells in the muscular layer. Following the surgery, she tolerated oral formula and passed stool normally via the anus with minimal stool content passing via the ileostomy. Her ileostomy was closed when she was 5 months old. She was doing well with normal development at the last 3-year follow-up.

Case No. 4 partially responded to rectal irrigation with normal saline solution. BE showed a microcolon with some dilated loops of the small intestine and good barium emptying from the colon in the 24-hour-delayed film (Fig. 3). She underwent laparotomy on the eleventh day of life at QSNICH. The operative findings included a microcolon and a Tz at the terminal ileum, 30 cm from the IC valve. Pathological examination from the frozen section revealed normal mature ganglion cells at the sigmoid colon, appendix and the terminal ileum (Tz). The abdominal incision was closed without further procedure and conservative treatment was resumed. Five days after laparotomy, she could pass stool and tolerated oral feeding well without abdominal distension and vomiting. Pathological report from the permanent section showed presence of both numerous immature and a few mature ganglion cells in the intramuscular layer of all 3 biopsied sites. She was doing well at the 2-year follow-up.

Case No. 5 had a severe clinical course (Table 1). She developed clinical sepsis and apnea at the age of 9 days with decreased body weight to 900 grams. The authors decided to operate on that day at SNH. The operative findings revealed a microcolon with a Tz at the terminal ileum, 60 cm from the IC valve (Fig. 4). Pathological findings from the frozen and permanent sections showed immature ganglion cells at the sigmoid colon and appendix with numerous mature ganglion cells at the terminal ileum just above the Tz approximately 5 cm (Fig. 5). A loop ileostomy was performed at the biopsied site. She developed diarrhea with elemental formula. She was conservatively treated with parenteral nutrition for a duration of 3 months. A full thickness rectal biopsy was done and the results revealed mature ganglion cells in the submucosal and muscular layer of the rectum. The ileostomy was closed 2 weeks after the rectal biopsy. The postoperative course was uneventful and she passed stool on the fifth postoperative day. She was doing well with normal development at the 2-year follow-up.

Case No. 6 underwent laparotomy on the fourth day of life at QSNICH. A microcolon and a Tz at the terminal ileum, 30 cm from the IC valve were seen during the operation. Pathological reports from the frozen and permanent sections revealed immature ganglion cells at the sigmoid colon and appendix with normal mature ganglion cells at the Tz of the terminal ileum. A loop ileostomy was performed at the biopsied site. Three months after laparotomy, a full thickness rectal biopsy was done. The pathohistological examination revealed presence of rare mature ganglion cells in the submucosal and muscular layer of the rectum. At the age of 4 months, she had a problem of severe ileostomy prolapse. Exploratory laparotomy was performed with closure of the ileostomy. The postoperative course was uneventful. She could pass stool normally. She was doing well at the last one-year follow-up.

Discussion

The most common causes of clinical low gut obstruction in neonates are mechanical obstruction from intestinal atresias and functional obstruction from HD including TCA. Other functional low gut

Table 2. Operative findings and operative procedures

Case No.	Age at operation (days)	Operative findings	Pathological reports (permanent section)	Operative procedures
1	6	Microcolon, Tz at the terminal ileum 30 cm above the IC valve	Terminal ileum at the Tz - normal ganglion cells Right transverse colon - immature ganglion cells	Ileostomy at the Tz Rectal biopsy after 5 months old Closure of ileostomy at the age of 7 months old
2	9	Microcolon, Tz at the terminal ileum 40 cm above the IC valve	Terminal ileum at the Tz - normal ganglion cells Right transverse colon - immature ganglion cells	Ileostomy at the Tz with colonic patching Rectal biopsy after 5 months Closure of ileostomy at the age of 7 months old
3	10	Microcolon, Tz at the terminal ileum 75 cm above the IC valve	Terminal ileum at the Tz, appendix and sigmoid colon - immature ganglion cells	Ileostomy at the Tz (80 cm from the IC valve) Ileoileostomy (ileal patching) at the age of 3 months old Closure of ileostomy at the age of 5 months old
4	11	Microcolon, Tz at the terminal ileum 30 cm above the IC valve	Terminal ileum at the Tz, appendix and sigmoid colon - numerous immature and mature ganglion cells	Closure of abdomen without ileostomy
5	9	Microcolon, Tz at the terminal ileum 60 cm above the IC valve	Terminal ileum at the Tz - normal mature ganglion cells Appendix and sigmoid colon - immature ganglion cells	Ileostomy at the Tz Rectal biopsy after 3 months old Closure of ileostomy at the age of 3.5 months old
6	4	Microcolon, Tz at the terminal ileum 30 cm above the IC valve	Terminal ileum - normal mature ganglion cells Appendix and sigmoid colon - immature ganglion cells	Ileostomy at the Tz Rectal biopsy after 3 months old Closure of ileostomy at the age of 4 months old

obstruction in neonates result from a variety of meconium obstructions such as meconium ileus associated with cystic fibrosis or mucovisidosis[10,11] small left colon syndrome from diabetic mother[12] and meconium plug syndrome possibly related with maternal administration of magnesium sulfate during delivery[13]. Premature infants with low birth weights less than 2.5 kilograms are prone to develop functional intestinal obstruction due to inspissated meconium, immature bowel function for peristalsis and a disturbance of physiological process with morphologically normal gastrointestinal tract[14-17]. This clinical entity often shows a microcolon on BE and has been called "microcolon of prematurity" by Amodio[18]. Almost all of these patients are successfully managed by saline irrigation or water soluble contrast enema such as gastrograffin.

There has been another group of premature infants in which the nature of functional obstruction remains obscure. This condition does not appear to fit into any of the above groups. The patients develop marked abdominal distension, bilious vomiting in the first few days of life with failure to pass meconium. Plain films of abdomen are compatible with low gut obstruction and BE reveals a microcolon similar to TCA. The symptomatologies do not respond to rectal irrigation and require exploratory laparotomy. The operative findings reveal a microcolon and a Tz at the terminal ileum like TCA, but frozen section shows immature ganglion cells in the entire colonic wall with normal ganglion cells in the dilated ileum. The functional low gut obstruction results from immaturity of the colonic ganglion cells and functional immaturity of the large bowel[19]. Vanhoutte and Katzman[20] called this clinical entity "aganglionosis-like syndrome", but Holschneider[7] categorized this finding into an allied disorder of HD.

Okamoto and Ueda[21] described a process of

Fig. 4 A microcolon and transitional zone at the terminal ileum in the Case No. 5.

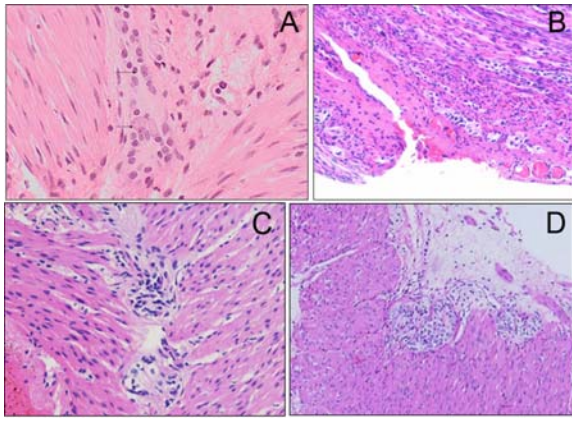

Fig. 5 Pathological examinations of the Case No. 5. A) ileum: intramuscular mature ganglion cells, having enlarged and eccentric nuclei with prominent red nucleoli and eosinophilic cytoplasm, B) appendix: immature with few mature ganglion cells in myenteric layers, C) sigmoid: immature ganglion cells in myenteric layers, having round centrally nuclei without prominent nucleoli, D) rectum: mature ganglion cells in both submucosal and myenteric layers (A-C: the first operation at 9 days old, D: rectal biopsy at 3 months old).

craniocaudal neuroblast migration, which originates in the vagal trunks approximately in the fifth week of gestation from the pharynx and esophagus to the distal segment of the intramuscular layer of the alimentary tract. Neuroblasts reach the rectum by the twelfth week of gestation. The last step is migration of the neuroblasts from the myenteric plexus to the submucosal plexus. The process of maturation of these neuroblasts also develops in a cephalocaudad direction. Neuroblasts in the esophagus and stomach are earlier mature than those in the colon and rectum[22]. Histological appearance of immature ganglion cells of the neuroenteric plexus results in functional obstruction of the bowel similar to HD and TCA. This entity is mostly found in the premature infants. However, it may be occasionally seen in full-term infants as in Case No. 4 of the present study. Smith[22] and Miyahara[23] have suggested that immaturity of the ganglion cells is physiologic and full maturation takes place gradually after one year of age.

Management of neonates with clinical presentations of low gut obstruction and the evidence of microcolon from barium enema is controversial. In our 6 patients, 5 were premature and one was full-term with body weight over 2,500 grams. All of the cases developed low gut obstruction that required laparotomy within 4-11 days after birth. Operative findings were positive for a microcolon and a Tz at the terminal ileum, 30 to 75 cm proximal to the IC valve, similar to TCA. Pathological examination revealed characteristics of immature ganglion cells in both the myenteric and submucosal plexuses of the colon. Our initial operative procedure was to correct functional low gut obstruction by a temporary ileostomy just above the Tz in the 5 premature infants. Rectal biopsy was performed at least 3 months after the initial ileostomy. Closure of the ileostomy were performed when the histopathology revealed presence of mature ganglion cells in the rectum. The full-term neonate was conservatively treated without ilesotomy because the frozen section revealed normal ganglion cells in all of the biopsied sites, along with evidence of good passage of the contrast material after BE. All of our 6 cases were doing well at the 1-3 year follow-up. In contrast, from our literature review, we see that many investigators did not use BE for diagnosis of these patients. They preferred initial management of functional intestinal obstructions by contrast (gastrograffin) enema including NSS enema for diagnostic and therapeutic purposes[17,18,24,25]. Rectal suctional biopsy may be done to rule out HD in the cases with no response to contrast and NSS enema. However, some patients might be prone to bowel perforation during contrast enema or delayed laparotomy in the cases with worsening clinical condition, especially in very low birth weight infants[16-18,22].

Conclusion

Neonates with low gut obstruction caused by immaturity of ganglion cells in the colonic wall should be managed by laparotomy, serial intestinal biopsy and enterostomy at the Tz. A rectal biopsy should be done after 3 months of age in order to confirm ganglion cell maturity before closure of the enterostomy. This condition in a full-term neonate may be successfully managed by conservative treatment without enterostomy.

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Potential conflicts of interest

None.

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จะรักษาโรคลำไส้ส่วนล่างอุดตันอย่างไร ในเด็กทารกที่มีเซลล์ปมประสาทตัวอ่อนในผนังของลำไส้ใหญ่?

รังสรรค์ นิรามิษ, อัจฉริยา ทองสิน, อัญชลีรัตน์ เลิศสถิตย์, ลั่นทม ตันวิเชียร, หทัยทิพย์ ชัยประภา, ไพโรจน์ จรรยางค์ดีกุล

ภูมิหลัง: ทารกที่มีเซลล์ปมประสาทตัวอ่อนในผนังของลำไส้ใหญ่ อาจจะแสดงอาการคล้ายโรค Hirschsprung's disease โดยเฉพาะภาวะไม่มีเซลล์ปมประสาทในผนังของลำไส้ใหญ่ทั้งหมด (total colonic aganglionsis หรือ TCA) การรักษาในโรคนี้ขึ้นอยู่กับการตัดสินใจของแพทย์แต่ละคน

วัตถุประสงค์: การศึกษาครั้งนี้มีวัตถุประสงค์เพื่อทบทวนแนวทางการรักษาโรคลำไส้อุดตันในเด็กทารกที่เกิดจากการมีเซลล์ปมประสาทตัวอ่อน ในผนังของลำไส้ใหญ่

ผลการศึกษา: ผู้ป่วยเด็กทารก 6 ราย เกิดครบกำหนด 1 ราย และเกิดก่อนกำหนด 5 ราย ได้รับการพิสูจน์แล้วว่ามีเซลล์ปมประสาทตัวอ่อนในผนังของลำไส้ใหญ่ ผู้ป่วยทั้งหมดมีอาการไม่ถ่ายขี้เทา ท้องอืดมากขึ้นเรื่อย ๆ และอาเจียนเป็นสีน้ำดีตั้งแต่เกิด ภาพรังสีของช่องท้องพบมีลำไส้เล็กโตขึ้นทั่วท้อง และการตรวจทางทวารหนัก โดยการสวนแบเรี่ยมในผู้ป่วย 4 ราย พบลำไส้ใหญ่ทั้งหมดมีขนาดเล็ก ผู้ป่วยทุกรายได้รับการผ่าตัดระหว่างอายุ 4 ถึง 11 วัน ผลการผ่าตัดพบว่าลำไส้ใหญ่มีขนาดเล็กทั้งหมดและพบลำไส้เล็กส่วนปลายขยายใหญ่ขึ้นชัดเจนตั้งแต่ 30 ถึง 75 ซม. เหนือรอยต่อของลำไส้เล็กและลำไส้ใหญ่ ผลการตรวจชิ้นเนื้อของลำไส้ใหญ่พบมีเซลล์ปมประสาทตัวอ่อนและลำไส้เล็ก ส่วนที่โตขึ้นมีเซลล์ปมประสาทที่เจริญเติบโตปกติ ทารกที่เกิดก่อนกำหนด 5 ราย ได้รับการรักษาโดยนำลำไส้เล็กที่โตขึ้นเปิดเป็นทวารเทียมไว้ที่ผนังหน้าท้อง ขณะที่ทารกเกิดครบกำหนดได้เย็บปิดหน้าท้องและรักษาโดยประคับประคอง ทารก 5 ราย ที่เกิดก่อนกำหนดได้รับการผ่าตัดปิดลำไส้เล็กที่เป็นทวารเทียม ภายหลังได้พิสูจน์พบว่ามีเซลล์ปมประสาทเป็นปกติแล้ว จากการตรวจชิ้นเนื้อที่ผนังของไส้ตรงเมื่ออายุมากกว่า 3 เดือน ผู้ป่วยทั้งหมด 6 ราย เป็นปกติดีจากการตรวจติดตามอาการระหว่าง 1 ถึง 3 ปี

สรุป: การอุดตันของลำไส้ส่วนล่างในเด็กทารกที่เกิดจากการมีเซลล์ปมประสาทตัวอ่อนที่ผนังลำไส้ใหญ่ ควรจะมีการรักษาขั้นต้นโดยการผ่าตัดเพื่อตัดชิ้นเนื้อที่ลำไส้ใหญ่ ไส้ติ่งและลำไส้เล็กบริเวณที่โตขึ้นส่งตรวจทางพยาธิวิทยาและยกลำไส้เล็กที่โตขึ้นเปิดเป็นทวารเทียมไว้ที่ผนังหน้าท้องก่อนผ่าตัดปิดทวารเทียมที่หน้าท้องควรพิสูจน์ว่าบริเวณไส้ตรงมีเซลล์ปมประสาทที่ปกติดีแล้ว โดยการตัดชิ้นเนื้อบริเวณไส้ตรงส่งตรวจเมื่ออายุมากกว่า 3 เดือน ในกรณีทารกที่เกิดครบกำหนดที่มีเซลล์ประสาทตัวอ่อนอาจจะรักษาได้สำเร็จ โดยวิธีประคับประคองโดยไม่ต้องยกลำไส้เล็กที่โตขึ้นเปิดเป็นทวารเทียมที่ผนังหน้าท้อง