Case Report

Enteral Autonomy in Surgical Short Bowel Syndrome from Necrotizing Enterocolitis: A Case Report

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Necrotizing enterocolitis (NEC) is one of the most common causes of short bowel syndrome in children. The prognosis of regaining intestinal function, weaning from parenteral nutrition and survival depend on multiple factors such as the remaining small intestinal length, the presence of ileocecal valve and overall intestinal adaptation. At present, more children with surgical short gut could transit to full enteral feeding. The minimal length of intestine to maintain full enteral autonomy has not been reported.

We present the case of a preterm baby who developed short bowel syndrome as a result of surgical management of NEC. With a remaining small intestinal length of 10 cm, she was dependent on parenteral nutrition for 2.5 years. She suffered multiple episodes of catheter related bloodstream infection and thrombosis. Ultimately she was able to achieve complete enteral autonomy.

Current surgical and nutritional management increases the survival rates and incidence of long-term intestinal autonomy even in the very short bowel patients. The presented case is an example of a surgical short bowel syndrome child and an optional surgical treatment for necrotizing enterocolitis to encourage pediatric surgeons to not lose hope in caring for these babies.

Keywords: *Short bowel syndrome, necrotizing enterocolitis, intestinal autonomy*

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Short bowel syndrome (SBS) is a clinical disorder defined as the insufficiency of the intestine to maintain normal nutrition and growth. It is characterized by malabsorption, malnutrition, metabolic disturbances and occasionally motility disorders[1-6]. The condition occurs as a result of functional or anatomical loss of the small intestine and possibly the colon. Necrotizing enterocolitis, intestinal atresia, aganglionosis and midgut volvulus are the common causes of short bowel syndrome in infant and small children. Predictors of intestinal function and survival depend on the remaining small intestinal length, the presence of ileocecal valve, spontaneous intestinal adaptation and the etiology of intestinal loss[1,2,4,5]. Current nutritional and surgical management can better support even the very SBS patients to have a longer survival, better quality of life and for some, the ability to regain intestinal autonomy[6]. The minimal small bowel length required for complete intestinal autonomy is difficult to determine owing to the diversity in the causes of intestinal loss and potential for adaptation[5]. The authors present a necrotizing enterocolitiscase to encourage other physicians to never give up hope in caring for these patients especially in developing countries, where intestinal transplantation is not yet an option for long term survival.

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Case Report

The following information has been reviewed and approved by the patient's family. According to the Siriraj Institutional Review Board policy as of 2016, an ethic committee approval was not required for a case report.

A 31 weeks premature female baby was delivered by Caesarean section due to maternal placenta previa totalis. Upon delivery, the APGAR score at 1, 5 and 10 minutes were 5, 6, and 10 due to skin complexion and tone. Her birth weight was 1,610 grams. She was provided intravenous fluids and total parenteral nutrition (TPN) via umbilical venous catheter for more than 3 weeks. After a few days, premature formula was initiated. She developed feeding intolerance and bilious vomiting on the 25th day of life. Per-rectal examination

showed a small amount of blood stained yellowish stool. Her feeding was sustained and intravenous antibiotics were started. Abdominal radiography demonstrated a distended stomach and several small bowel loops as shown in Fig. 1. There were no pneumatosis intestinalis or abnormal bowel gas in the following x-rays. She remained clinically active with bleeding per rectum and no obvious thrombocytopenia, thus an emergency upper gastrointestinal contrast study was done to rule out midgut volvulus. The contrast study revealed that water-soluble contrast passed freely through the stomach and distended proximal small intestine, although not passing through to the colon (Fig. 2A). Fourteen hours later, follow-up film showed contrast leakage into the peritoneal cavity (Fig. 2B).

On exploratory laparotomy there was near total small bowel gangrene sparing 3 viable jejunal and ileal segments of 1.5 cm each. The viable proximal jejunal stump was 3 cm from the duodenojejunal junction and the viable terminal ileal segment was 4 cm from the ileocecal valve (ICV). Several lengthy segmental small bowel resections, edge debridement and end-to-end anastomoses were accomplished using single layer interrupted absorbable suture stitches. At completion of the procedure, the total small bowel length was 10 cm altogether and the ileocecal valve was preserved. TwoPenrose drains were placed on each side of the abdomen for peritoneal drainage and nasogastric tube was ascertained its proper position for decompression. Post-operative recovery was uneventful.

During early postoperative period, intravenous fluid replacement with 80 to 100 mEq/L of sodium and 2 to 3 mEq/kg/day of possasium was given to maintain fluid and electrolyte balance due to high nasogastric tube output. Once her metabolic and cardiovascular statuses were stabilized, a tunneled central venous catheter was placed in the right internal jugular vein and TPN administration of 80 to 90 kcal/kg/day was provided. Postoperative ileus resolved within a couple of weeks, which she started to have bowel movement. Unfortunately breast milk was unavailable and small trophic feedings of diluted protein hydrolysate formula with medium chain triglycerides was initiated. The volume of her feedings could gradually be increased and continuous drips via nasogastric tube were tolerable. As enteral feeding was slowly advanced by concentration and volume, TPN was reduced by rate every other week. Large amount of loose stool of more than 40 ml/kg/day was replaced with isotonic solution[7]. Stool output increased as feedings were advanced. Intravenous proton pump inhibitor was switched to oral form and sodium supplements were adjusted to weekly labs. Small quantities of premature formula according to were fed

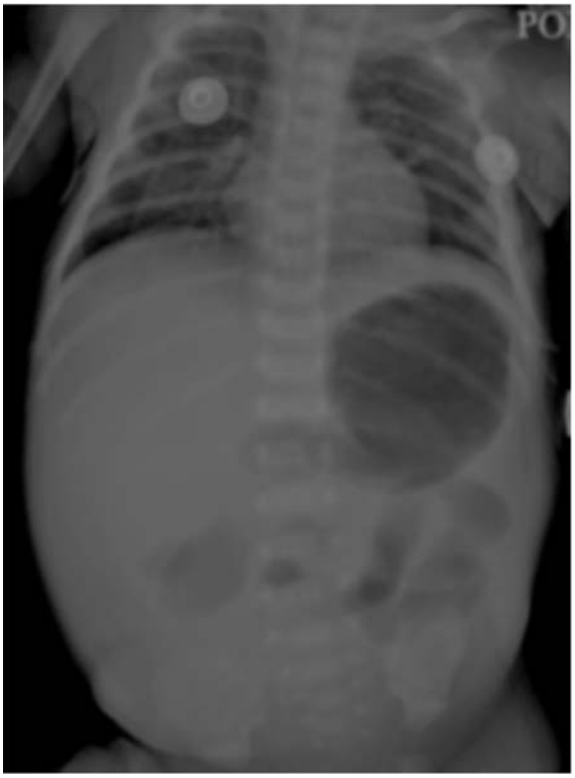

Fig. 1 Abdominal radiography on the 25th day of life after developing feeding intolerance and bleeding per rectum, demonstrated a distended stomach and several small bowel loops.

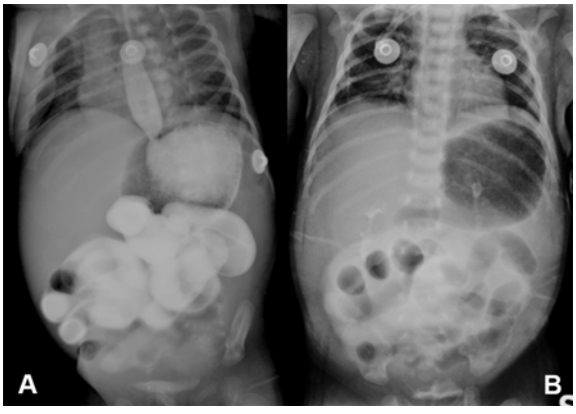

Fig. 2 Upper gastrointestinal study showed water soluble contrast passing through the distended stomach freely into the dilated proximal small intestine (A). Follow-up film 14 hours later showed contrast leakage into the peritoneum (B).

orally 2-3 times a day to stimulate sucking and swallowing. At 3 months of age, she was able to tolerate half of her daily volume requirements in 8 enteral feedings a day of 1-2 hours drip half strength hydrolyzed formula, having normal texture stool output. Although her weight and length was below the 3rd percentile for her age as demonstrated in the growth chart in Fig. 3, according to the normal growth reference, her growth increased slowly in her own pace.

Catheter related blood stream infection (CRBSI) and acute gastroenteritis were encountered every now and then, which significantly affected overall absorption and growth. The most severe episode progressed to respiratory distress syndrome and disseminated intravascular coagulopathy, which put her on high frequency ventilation for a week at 4 months of age. At 6 months of age, 2/3 of her energy requirements were from enteral feedings. Daily feeds included 1-hour drip fullstrength hydrolyzed formula given every 3 hours. Nocturnal feeds were given over 8 hours. Parenteral nutrition was given in daily cycles of 12 to 16 hours. At 9 months of age, oral bolus feeding slowly replaced tube drip feedings, which she was able to tolerate 3/4 of daily volume requirement in 20 calories per ounce of premature formula 8 meals a day. Although she did not gain weight well, there was measurable skin folds and muscle mass. One year after the surgery, long gastrointestinal contrast study demonstrated slight elongation and enlargement of the small bowel although not dilated which made none of the intestinal lengthening procedure suitable for her. She weighed 4 kilograms and was taking 2 onces of full strength premature formula 8 bottles a day. Initially, any blended complex carbohydrate food would cause flatulent and diarrhea. Over the second year of her life in and out of the hospital, she slowly tolerated puree food and ultimately small soft to regular diet meals having 1 to 2 bowel movements a day. At 2 to 3 years of age, she weighed 7-9 kilograms respectively (Fig. 3).

Follow-up on the liver functions during the first year of life showed elevation of Alanine aminotransferase 2 to 8 times the normal limit and total bilirubin and direct bilirubin were 2 to 7 and 5 to 20 times, respectively. Each episode of transaminitis or cholestasis was quickly managed by changing the parenteral fat component to lipid emulsion with fish oil, decreasing the protein and carbohydrate concentration or even sustaining the parenteral nutrition to a crystalloid solution for a few days. Later on, as enteral feedings increased, bilirubin levels and liver enzymes returned to normal. Finally at 30 months old the patient

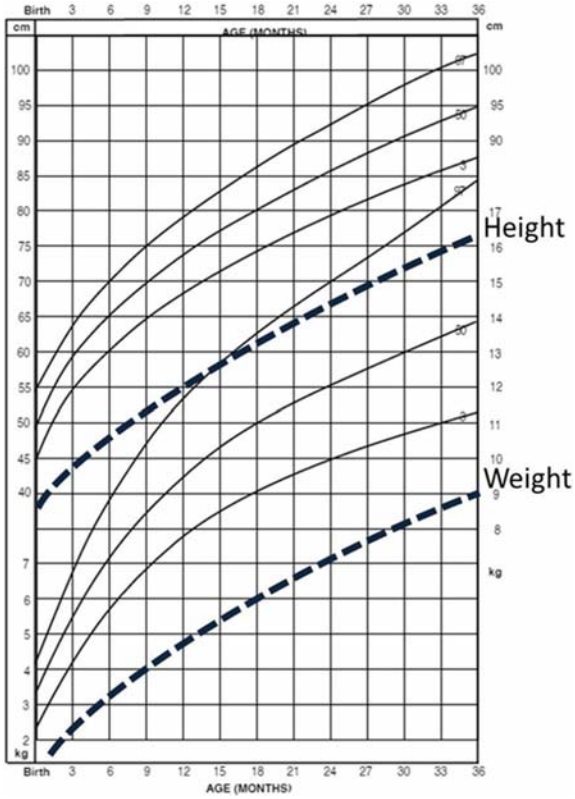

Fig. 3 National growth reference in weight and height for Thai girls from birth to 3 years old. The patient's weight and height was beneath the 3rd percentile as demonstrated in dashed lines.

completely weaned from TPN, liver function was normal. The patient recovered from TPN induced cholestasis. Catheter related blood stream infection has been difficult to avoid since the initiation of TPN. Despite strict protocol for catheter care, CRBSI occurred every 2-3 months. Intravenous third to fourth generation Cefalosporins or Carbapenem in combination with Aminoglycosides were frequently used for CRBSI. Metronidazole was added if enterocolitis or intestinal source of infection was suspected. Oral cyclic metronidazole was prescribed during the first week of the month for breakthrough enterocolitis in the first year of life and slowly tapered off when the episodes were less frequent. At 2 years of age she developed chronic central venous thrombosis after which catheter removal for a CRBSI incidence was followed by failure to insert another central venous catheter. CT venography demonstrated chronic venous thrombosis of bilateral internal jugular veins extending beyond the brachiocephalic trunk. There was also chronic venous thrombosis of both iliac veins extending into the inferior

vena cava.Initial antithrombotic therapy failed to recanalize the central veins in her neck; chest and femoral, thus peripheral lines were established when needed. Over the second year of her life, having cyclic parenteral nutrition 3 to 4 days a week, she had subcutaneous administration of low molecular weight heparin for the chronic venous thrombosis. Until she finally weaned from parenteral nutritionand at 33 months of age, her central veins recovered. Having a noticeable decrease in superficial dilated veins and head circumference proportionate to her body, a Doppler ultrasonography of the neck and groins confirmed recanalization of both jugular and femoral veins. The low molecular weight heparin was eventually discontinued.

The patient had delayed development in all aspects. The evaluation of her developmental milestones at 35 months old was comparable to a 24-month-old. The patient showed a high capacity to continue to improve in her development despite her petite body. At 40 months of age, she can communicate in sentences and recognizes her family members and caretakers throughout every visit she had to the hospital.

Upon review at 42 months, the patient was able to maintain her body weight without TPN for more than 18 months and there has been no further incidence of bloodstream infection since the removal of central catheter. Her body weight on enteral feeding alone was 8 to 9 kg and height 76 cm tall. Eating normal soft to solid food, she has regular bowel movements of 1 to 2 times a day. No secretory or infective diarrhea incidence after weaning from cyclic antibiotics. Her length increased more than weight in the last few months of review. Through investigations, she is dependent on oral sodium bicarbonate, vitamin D and monthly B12 supplements. Her growth and blood chemistry was regularly monitored every 3-6 months while she has been consuming conventional diet and living well. Re-evaluation by gastrointestinal contrast study demonstrated a small intestinal length of 18 cm with slightly distended caliber of both small and large intestine. The antegrade filling of contrast medium into the large bowel or transit time was 5 hours.

Discussion

Although the minimal small bowel length that defines SBS is difficult to determine, many studies suggested that term infants with as little as 15 to 20 cm of small bowel and an ileocecal valve (ICV) or more than 40 cm of small bowel without the ICV can be successfully weaned from parenteral support[1-5]. These outcomes, however, presumed normal functioning of the remaining bowel[5]. Our observation of over 100 pediatric surgical short bowel syndrome patients in 10 years at Siriraj Hospital, Thailand, suggested that a length of at least 40 cm of normal functioning small bowel is necessary to achieve long term intestinal autonomy either with or without the ICV. There is no better way to treat a short bowel patient than to prevent the short bowel from happening. Unfortunately it is inevitable in most cases. Surgeons well aware of SBS are meticulous in resection of the bowel to preserve enough length and tapering to appropriate size for best functional results.

Following massive surgical resection of the intestine, the remaining gastrointestinal tract adapts with epithelial cellular proliferation, reorganization of the crypt-villous histoarchitecture leading to increase villous height and diameter and enlongation of the crypts. Additionally functional changes in absorption and motility occurs[8,9]. Several studies demonstrated dilatation, thickening and lengthening of the remaining bowel[10,11]. Thepotential for intestinal adaptation varies among each individual depending on the underlying condition of the patient and the etiologies of intestinal loss, which NEC fared the best. Kurkchubasche A et al[5] proposed that 10 cm of small intestinal length to be the new lower limit for the bowel length necessary to achieve full enteral nutrition and that early institution of enteral feedings should be initiated to promote intestinal adaptation for a minimum of 2 years before transplantation is considered. The study was consistent with others[8,11] suggesting intestinal adaptation to be an ongoing process lasting from months to years of recovery. Early enteral feeding has been demonstrated to induce acute post-surgical intestinal adaptation, likely to improve functional capacity of the remaining bowel[8]. Massive intestinal resection was unavoidable in the presented case with necrotizing enterocolitis totalis. After the bowel edges were trimmed, it was decided to perform primary anastomosisfor the remaining small bowel segments were too short to create an ostomy and the only proximal diversion was the large nasogastric tube placed in the stomach for decompression in early postoperative period also with hopes to initiate enteral feeding later. Early enteral feeding was initiated and possibly contributed to an appreciable intestinal adaptation. It was proposed that early intestinal anastomosis and enteral feeding can promote substantial intestinal adaptation than a diverting ostomy. However, this is

not always the case in babies with necrotizing enterocolitis; since intestinal anastomosis would be considered only when there is undoubtedly intestinal viability with certainty. Oftentimes the remaining bowel length would be insufficiently shorten after debridement for anastomosis,which a proximal diversion or multiple ostomies could better preserve the questionable viable bowel for longer remaining intestinal length in future anastomoses or bowel lengthening procedures[12]. Therefore,especially in patients with necrotizing enterocolitis, the risk and benefits must be considered individually to avoid anastomosis leakage and stricture when considering primary anastomosis. On the other hand, preservation of sufficient intestinal length is of utmost importance to prevent future short bowel syndrome nevertheless inadequate debridement of necrotic tissue may be the source of worsening sepsis.

Conclusion

The presented case is an example of a surgical short bowel syndrome child and an optional surgical treatment of necrotizing enterocolitis, to encourage pediatric surgeons to not lose hope in treating even the very short bowel babies. For surgical short bowel syndrome, the small bowel length is not the only factor to predict the long term outcome. Early enteral feeding may contribute to acute post-surgical intestinal adaptation improving the functional capacity of the remaining bowel in the long term.

What is already known on this topic?

The treatment of necrotizing enterocolitis (NEC) is a challenge and unfortunately, often times the consequence leaves the child with short gut syndrome. The surgical treatment for NEC requires removal of necrotic bowel and all sources of infection at the same time preservation of the remaining bowel sufficient to avoid short gut syndrome.

What this study adds?

The presented case is an example of a surgical short bowel syndrome child and an optional surgical treatment for necrotizing enterocolitis, which the child eventually was able to tolerate enteral feedings and weaned off parenteral nutrition. An example case to encourage pediatric surgeons to not lose hope in caring for these babies.

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Potential conflicts of interest

None.

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ความสำเร็จในการหยุดให้สารอาหารทางเส้นเลือดในเด็กที่มีภาวะลำไส้สั้นภายหลังการผ่าตัดรักษาการอักเสบติดเชื้อรุนแรงในลำไส้: รายงานผู้ป่วย 1 ราย

บุนเหลือ ลิดดัง, นิรมล ตันเต็มทรัพย์

ภาวะการอักเสบติดเชื้อรุนแรงในลำไส้เด็กทารกเป็นหนึ่งในสาเหตุของการเกิดภาวะลำไส้สั้นในเด็ก โดยคำจำกัดความของภาวะลำไส้สั้นหมายถึงการที่ร่างกายไม่สามารถดำรงชีวิตอยู่และมีการเจริญเติบโตได้ โดยอาศัยพลังงานจากสารอาหารที่รับประทานอย่างเดียว จำเป็นต้องให้อาหารทางเส้นเลือดช่วย ภาวะลำไส้สั้นอันเกิดภายหลังการผ่าตัดลำไส้เป็นปัญหาที่พับบ่อยทางกุมารศัลยกรรม การพยากรณ์โรคขึ้นอยู่กับความยาวของลำไส้ที่เหลืออยู่การมีหูรูดลำไส้เล็ก ตลอดจนความสามารถในการฟื้นฟูสภาพของลำไส้เล็กที่เหลืออยู่

ผู้นิพนธ์ได้นำเสนอผู้ป่วย 1 รายที่เกิดการอักเสบติดเชื้อรุนแรงของลำไส้ จำเป็นต้องผ่าตัดลำไส้ที่เน่าออกเหลือความยาวลำไส้เล็กโดยรวม 10 เซนติเมตรและเกิดภาวะลำไส้สั้นตามมา ผู้ป่วยประสบปัญหาจากการดูดซึมไม่ดี มีการติดเชื้อในระยะแรกและได้รับการฟื้นฟูด้วยสารอาหารทางเส้นเลือดตลอดจนยาปฏิชีวนะได้รับสารอาหารผ่านทางเดินอาหารหลายรูปแบบ จนลำไส้สามารถปรับตัวย่อยอาหารและดูดซึมได้ ต่อมามีภาวะแทรกซ้อนจากสายให้สารอาหารทางเส้นโลหิตจนในที่สุดสามารถรับประทานอาหารทางปากเป็นอาหารที่เหมือนคนปกติ และไม่ต้องใส่สายให้อาหารทางเส้นโลหิตอีกต่อไป มีการเจริญเติบโตได้ถึงแม้จะช้ากว่าปกติ

รายงานผู้ป่วยรายนี้เป็นตัวอย่างของการรักษาที่ได้ผลดีแม้ในภาวะการติดเชื้ออักเสบรุนแรงของลำไส้ ส่วนหนึ่งน่าจะมาจากการผ่าตัดและต่อลำไส้ในการผ่าตัดครั้งแรก ตลอดจนการให้อาหารทางลำไส้โดยเร็วหลังผ่าตัดทำให้มีการฟื้นฟูสภาพของลำไส้ส่วนที่เหลือได้ดี นอกจากนี้ยังเป็นการให้กำลังใจแพทย์ที่ต้องดูแลผู้ป่วยกลุ่มนี้ไม่ให้สิ้นหวังแม้จะเหลือลำไส้สั้นมากก็ตาม