CASE REPORT

Ultrashort Bowel Syndrome: long-term survival without ParenteralNutrition

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ABSTRACT

Short Bowel Syndrome (SBS) is a condition in which nutrients are not absorbed (malabsorption) because a large part of the small intestine is missing or has been surgically removed, developing an Intestinal Failure (IF), that is defined as a loss of absorptive capacity secondary to obstruction, dysmotility, surgical resection, congenital defect, or mucosal disease resulting in chronic diarrhea, dehydration, electrolyte abnormalities, micronutrient imbalance, and malnutrition. (1)

The long-term survival without parenteral nutrition in patients with the diagnosis of ultrashort bowel syndrome is difficult.

This is a case, first reported in the country, of a 63 years old woman with ultrashort bowel syndrome who after three abdominal surgeries was left with only about 35 cm of small intestine with a jejune-transverse anastomosis. During the first surgery the right colon was removed.

From the beginning she received Total Parenteral Nutrition (TPN) lasting about three months, at which time she presented a serious reaction after which she refused further TPN treatment. She lived with oral feeding including nutritional supplements for almost five years, and one significant complication,diarrhea, was managed with loperamida and octreotride.

Keywords

short bowel syndrome; total parenteral nutrition.

When areas of the small intestine are removed by surgery, or they are missing due to a birth defect (congenital defect), there may not be enough surface area left in the remaining bowel to absorb enough nutrients from food.

This condition is likely to develop when one-half or more of the bowel is removed during surgery.

The average length of the adult human small intestine is approximately 600 cm, as calculated from studies performed on cadavers. According to Lennard-Jones and to Weser, this figure ranges from 260-800 cm. (2). Any disease, traumatic injury, vascular accident, or other pathology that leaves less than 200 cm of viable small bowel or results in a loss of 50% or more of the small intestine places the patient at risk for developing SBS.

SBS is a disorder clinically defined by malabsorption, diarrhea, steatorrhea, fluid and electrolyte disturbances, and malnutrition. The final common etiologic factor in all causes of SBS is the functional or anatomic loss of extensive segments of small intestine so that absorptive capacity is severely compromised. Although resection of the colon alone typically does not result in SBS , the condition's presence can be a critical factor in the management of patients who lose significant amounts of small intestine. (3,4)

Massive small intestinal resection compromises digestive and absorptive processes. Adequate digestion and absorption cannot take place, and proper nutritional status cannot be maintained without supportive care. Today, the most common causes of SBS in adults include Crohn disease, (5) radiation enteritis, mesenteric vascular accidents, trauma, and recurrent intestinal obstruction. In the pediatric population, necrotizing enterocolitis, intestinal atresias, and intestinal volvulus are the most common etiologic factors. Other conditions associated with SBS include congenital short small bowel, gastroschisis, and meconium peritonitis.

Not all patients with loss of significant amounts of small intestine develop SBS. Important cofactors that help to determine whether the syndrome will develop or not include the premorbid length of small bowel, the segment of intestine that is lost, the age of the patient at the time of bowel loss, the remaining length of small bowel and colon, and the presence or absence of the ileocecal valve.

In the majority of cases they eventually present complications related with TPN for a long time, like a liver failure, osteoporosis, osteomalacia, etc. (6,7,8)

CASE REPORT

In this case of a 63 year old woman, diagnosed with ultrashort bowel syndrome who survived almost 5 years without long-term parenteral nutrition.

The patient was 59 kg and had good muscle mass (tennis player). She underwent surgery with a diagnosis of epigastric tumor, found a mesentery tumor like a fibroma.

During surgery, a portion of the small intestine was removed and a transverse hemicolectomy was performed with terminal ileostomy and ileotransversostomy plus tumor resection was performed.

On the 5th day post-op, she had a thrombosis of the superior mesenteric artery and went back to the operating room where a Myocardial Mesenteric was found during an exploratory laparotomy, and almost all the small bowel was removed leaving approximately 12 cm of jejune. In the same surgery a right colectomy was performed with a jejune-transversostomy.

Six days after the second surgery she had post-operative complications of two high output enterocutaneous fistulas that closed with an aspirative system plus TPN. She was admitted to the Intensive Care Unit (ICU) for a period of two weeks, presenting other complications during her stay in the ICU. (Nosocomial Pneumonia and left pleural effusion). Then was transferred to a regular ward, and continued to receive TPN for four weeks more. And then went home to Home Parenteral Nutrition (HPN).

On the 5th day at home, she started oral feeding and continued with Cyclic Parenteral Nutrition (nocturnal) and also she started industrial semielemental formulas with good tolerance. For a period of 40 days she was with oral feeding and a low fiber diet and semielemental industrial formulas. Her usual weight was 59 kg, after 4 months she had lost 10 kg, and then after 7 months she lost 7 kg more. The patient performs daily physical therapy to strengthen her muscles.

The patient always received parenteral hydration at the same time.

She continued with the previously mentioned nutritional support until on two occasions she presented an allergic reaction with chills, hypotension, and sense of death, the first at 15 minutes after beginning the Parenteral nutrition (PN) and the second, after infusion of 16 ml of PN. From that moment, the patient decided she did not want to receive PN anymore, even knowing the risks of death from the disease being untreated.

From this moment was added electrolytes, calcium, phosphorus, magnesium, zinc, trace elements and multivitamins daily to her hydration. (She had a multi-puncture catheter)

The patient had several bowel movements per day: ranging in frequency but most times of a liquid consistency, controlled by loperamide dose/response.

After 6 months she developed liver failure, managed by the hepatologist. One year later, she had an upper endoscopy and a chronic erosive gastritis, alkaline gastritis, and chronic duodenitis was found, that was treated with omeprazole.

Three months after the endoscopy, she had intestinal transit that showed a symmetrical swallowing, a good esophageal and cardial passage. No filling defects or parietal deformations of the lesional character were observed in the esophagus.

Stomach, duodenal bulb and arc without gross abnormalities. Two jejunal loops and permeable anastomosis ileotransverso with a normal passage of contrast into the colon were identified.

One year after the first surgery she returned to the hospital to receive TPN, because she was afraid to receive it at home despite having a good team of nurses. She received TPN (without multivitamins in the bag) without issue for a period of one month. With this treatment she recovered, she felt stronger and had more energy, gaining a small amount of weight.

It was proposed a nutrition plan consisting of 3 days with TPN, three days with Hydration and one day at rest per week but she refused the plan due to fear.

For an entire year she went without TPN.

In one occasion she presented an uncontrollable diarrhea even using the maximum doses of loperamida but with a monthly dose of octreotride it was controlled.

Throughout the duration of the time, the albumin and pre-albumin fluctuated with a tendency to drop.

From the last hospitalization, she continued parenteral hydration and oral diet low in fiber plus semielemental industrial supplements. In addition, she received octrotride plus loperamide.

On several occasions she had strong joint pain, treated with anti-inflammatories. Later the pain prevented her from walking despite physiotherapy.

She continued losing weight, although we were unable to quantify it because she refused to get on the scale. After almost 5 years of the diagnosis of ultrashort bowel syndrome, she experienced an infection forcing her to return to the hospital where she received TPN for 5 days and was treated with antibiotics. Then she acquired a nosocomial pneumonia requiring ICU and mechanical ventilation. She could never improve due to her deteriorated state and after three days she died.

DISCUSSION

In patients with ultrashort bowel syndrome survival is so difficult without TPN, because the length is not the only factor that causes difficulty, the remaining portion of the bowel and the presence of the ileocecal valve will influence the absorption of the nutrients.

In this case the diarrhea was an important factor that contributed to her rapid nutritional deterioration, because once it controlled, the patient improve her nutritional state.

Another important aspect to mention is the difficulty arises when the patient does not accept treatment prescribed, knowing well the risks involved in not accepting it. But on the other hand, it is valid to respect patient autonomy as one of the principles of Medical Ethics.

This case could demonstrate the importance of team effort and that it can have a positive impact on patient survival.

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