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Resolution of infantile intestinal pseudo-obstruction in a boy

G. Angsten [a, *], E. Gustafson [a], N. Dahl [b], R.H. Christofferson [a]

[a] Section of Pediatric Surgery, Department of Women's and Children's Health, Uppsala University, Uppsala, Sweden
[b] Department of Immunology, Genetics and Pathology, Uppsala University, Uppsala, Sweden

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ABSTRACT

A term boy with spontaneous passage of meconium exhibited episodes of abdominal distension and diarrhea. Due to failure to thrive and suspicion of Hischsprung's disease he was referred to our university hospital at five months of age. Rectal biopsies were normal. Laparotomy revealed dilation of the small bowel and colon without any mechanical obstruction. Full thickness bowel biopsies were taken and a loop ileostomy was constructed. Histopathology revealed fibrosing myopathy, Cajal cell hypertrophy, and neuronal degeneration in both the large and small bowel. The small bowel showed mastocytosis without inflammation. A central venous catheter was placed for vascular access, replaced three times and later switched to a subcutaneous venous port. Catheters were locked after use with vancomycin-heparin and later taurolidine. The individually tailored home parenteral nutrition contained unsaturated fatty acid lipids to reduce cholestasis. Initial insufficient growth was improved after correction of partial parenteral nutrition based on a metabolic balance study.

The ileostomy was revised once and finally taken down at 11 years of age following one year without parenteral support. At follow-up at 13 years of age he has episodes of moderate abdominal pain and has entered puberty and reports a high quality of life.

1. Introduction

Intestinal pseudo-obstruction in children is characterized by bowel dilation, abdominal pain, and bowel failure, leading to impaired growth and development. In most cases, intestinal pseudo-obstruction is considered to be chronic condition (CIPO), and treatment is parenteral nutrition or bowel transplantation [1,2]. The CIPO diagnosis is suspected to be incorrect in anecdotic cases of resolution. The intestinal morphology in pseudo-obstruction is under revision, and is currently descriptive and quantitative rather than diagnostic [3], but it is traditionally separated into neuropathies, myopathies, or combinations of them. The aim of this report is to present a well-documented case of resolving infantile intestinal pseudo-obstruction.

1.1. Patient

The boy F.M. was born at term (40 weeks) after a normal pregnancy, birth weight 3450 g and birth length 50 cm. He was the first child of a healthy mother, and he was breastfed from start with spontaneous passing of meconium within the first 36 h after birth. His first six weeks in life were normal, after which he developed progressive episodes of vomiting several times a day and abdominal distension. He had pain and his stools were alternately hard or watery. A regime with a milk-free formula was tried for two months without any improvement of his symptoms. He was then referred to the section of pediatric surgery at our university hospital at five months of age due to failure to thrive. A plain radiograph of the abdomen revealed massive bowel dilatation (Fig. 1) and a contrast enema suggested Hirschsprung's disease with a transition zone located to sigmoid colon. Though, the following rectal suction biopsies confirmed presence of ganglion cells and absence of nerve hypertrophy. Exhibiting all signs of bowel obstruction, he was subjected to semi-emergent explorative laparotomy at five months of age. There was considerable dilation of the small bowel, but also colon was dilated without any transition zone. A loop ileostomy was constructed on the dilated distal, but not terminal, ileum. Full thickness intestinal wall biopsies were taken from jejunum, ileum, ascending -, transverse -, and sigmoid colon. A 4 Fr tunnelled central venous catheter (CVC) was placed for parenteral nutrition.

Small bowel manometry wasn't available at our department at this time, but could have characterized the disease.

* Corresponding author. Section of Pediatric Surgery, Children's Academic Hospital, SE-751 85, Uppsala, Sweden.
E-mail address: gertrud.angsten@kbh.uu.se (G. Angsten).

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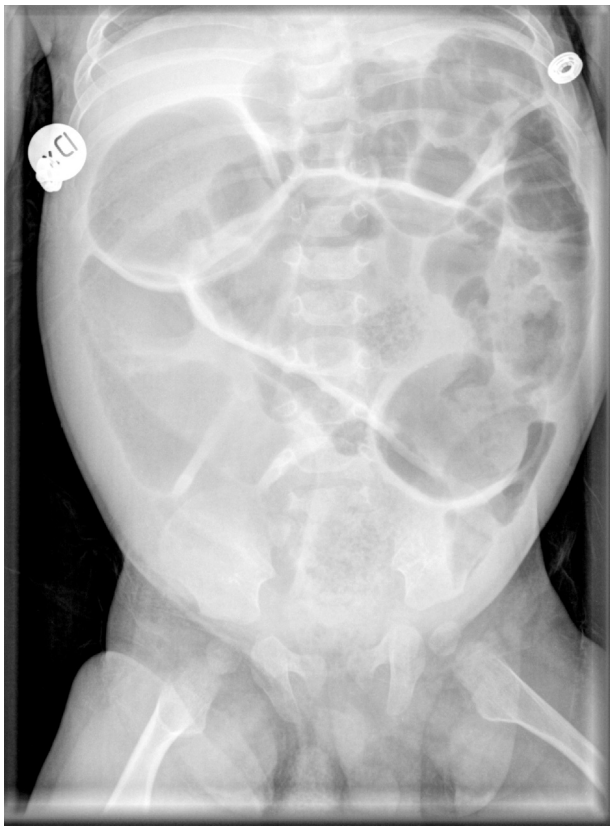

Fig. 1. A plain radiographic of the abdomen.

Postoperatively he received total parenteral nutrition (indications given in Table 1) for neonates with essential nutrients and soybean oil-based fat emulsion. There was no stomal output the first week. The parents were taught home parenteral nutrition (HPN) during their three weeks stay in hospital. His weight had increased to 5200 g at discharge, when he was put on a breast-milk and cow's milk-free formula, to which a high-calorie, medium (MCT) - and long chain triglyceride (LCT) supplement was added (~500 ml), and partial parenteral nutrition (450 ml).

1.2. Histopathology

Pathologist's report on the full thickness biopsies stated fibrosing myopathy, most pronounced in the small bowel, interstitial Cajal cell hypertrophy and hyperplasia, and signs of limited neuron degeneration in both the large and small bowel. There were more mast cells (16 vs. normally 5 per high-power field of vision; mastocytosis) in the fibrosis between muscles layers in the small bowel, but there were no signs of inflammation. The mucosa and submucosa were normal in all biopsies.

1.3. Metabolic balance

At 1.5 years, a metabolic balance study was performed because of poor growth (weight −4 S.D., length −3 S.D.; Fig. 2). Among blood tests, concentrations of e.g. hemoglobin, leukocytes, platelets, aspartate aminotransferase, alanine aminotransferase, albumin, bilirubin, sodium, potassium, creatinine, alkaline phosphatase and thyroid stimulating hormone and acid-base-balance, were measured. Urine and faeces were collected during three days for analysis of sodium, potassium and chloride as well as fecal fatty acids and trace elements. The mother kept a diary over his oral food intake, and the dietician calculated the intake of calories. The volume, content and energy in his daily HPN was known and kept constant. The basal metabolic rate was measured in a Deltatrac® apparatus with a pediatric mixing chamber (Datax Division Instrumentarium Corp., Helsinki, Finland) respirometer. The boy's total energy intake and losses per day were calculated and were lower and higher, respectively, compared to those expected at his age. The dietician improved his oral energy intake, and a new HPN was composed. The lipid emulsion was changed to a structured mixture of MCT/LCT (Structolipid ®; Fresenius Kabi Marknadsbolaget, Uppsala, Sweden), supplemented with carnitine. The boy's oral intake continued to be poor, but after six months he had grown 6 cm in height and increased 3 kg in weight (Fig. 2).

1.4. Management

He was medicated with oral ranitidine, 60 mg daily; ursodeoxycholic acid, 50 mg 3 times daily, and intravenous metronidazole (20 mg/kg) as a monthly bolus to reduce enteral bacterial overgrowth. He was vaccinated against pneumococci. Hematological and biochemical parameters were normal apart from consistently low leukocyte counts, which were further reduced at frequent episodes of viral infections and some bacterial septicaemias, and a bone marrow biopsy showed a slightly reactive marrow.

At 2 years and 8 months of age, a revision of the ileostomy was made due to stricture and prolapse. He also received a gastrostomy because his oral intake was minimal, and parenteral metronidazole was switched to 5 ml of trimethoprim-sulfamethoxazole mixture daily in the gastrostomy. He received the same HPN for three years, adjusted to his current weight. He also received 60 ml formula and 10 ml water four times daily in his gastrostomy. He was followed-up every 6 months with blood samples and clinical examination at the university hospital and sometimes also at the local hospital.

At 3 years of age he began to eat a little, speak two word-sentences, and continued to gain weight (Fig. 2). Due to elevated concentrations of liver enzymes, the lipid emulsion in HPN was switched to 50% MCT and 50% LCT (Vasolipid®, B Braun Medical AB, Danderyd, Sweden), and liver enzymes were normalized. He received HPN for 16 h daily. Later Omegaven® (Fresenius Kabi Marknadsbolaget, Uppsala, Sweden), a lipid emulsion rich in omega-3 fatty acids derived from fish oil was added to the HPN.

The parents raised the feasibility of bowel transplantation, and also met a clinical geneticist to discuss the risk of CIPO in a sibling. In this context, it deserves to be mentioned that the boy had not and has not bladder or urinary symptoms. The etiology to CIPO is heterogeneous and most cases are sporadic without family history of the condition. In our case, there was no family history of CIPO or CIPO-associated symptoms. However, rare familiar cases of CIPO have been reported that follow X-chromosome linked, autosomal recessive or dominant inheritance [4–8]. The few genes associated with CIPO over the last years were not known at the time for the genetic counselling in this boy and no genetic analysis was performed [9]. The family has currently chosen to wait until the boy is older and can make his own decision regarding genetic analysis.

At 4 years, he began day nursery and gave his first birthday party. His growth charts were good (Fig. 3) and HPN was reduced to 14 h daily 6 days a week, and 6 h the seventh day to improve appetite (see Table 2 for clinical criteria for weaning off parental nutrition). The HPN was further reduced to 13 h daily 5 days a week and 6 h for 2 days. He ate three meals and two between-meals every day (see Table 3 for a nutritional overview). Appetite was better on days with reduced HPN. His balance and motor activity improved. His speech developed slowly and a speech therapist was