Concepts
Intestinal rehabilitation in children with short bowel syndrome/intestinal failure is a painstaking but ultimately rewarding process. The goal of therapy is the transition from parenteral support to full oral nutrition, while maintaining a high quality of life for patient and family and avoiding the multiple medical and surgical complications of these disorders and their treatment. This session will highlight several of the challenges of rehabilitation, as well as illustrate the benefits of a multidisciplinary approach.

Case 1
The patient was a 3 year old female with a history of gastroschisis diagnosed by prenatal ultrasound. She was born at 37 weeks gestation with a birth weight of 3.450 kg. At surgery in the neonatal period, she was found to have a concomitant ileal atresia. She underwent primary gastroschisis repair, resection of 14 cm of distal atretic ileum including the ileocecal valve, and placement of a diverting ileostomy. Residual small bowel length measured intra-operatively from ligament of Trietz to ileostomy was 125 cm. During the first four months of life, she received full parenteral nutrition and increasing amounts of enteral nutrition. She was weaned completely from parenteral nutrition at age 3 months. At 4 months of age she underwent a take-down of the ileostomy. At age 4.2 months she was discharged home on full oral nutrition (an amino acid-based formula).

1. What if any outpatient follow-up is indicated? What if any issues should be followed up?

At her first outpatient visit at age of 5 months, she was growing well and normal blood levels of zinc, vitamins A, E and B12.

2. What follow-up should occur now?

From 5 to 36 months of age she grew well, along the 50th % for weight and 10th % for length. She had no chronic abdominal pain, diarrhea or vomiting. She did not receive any dietary supplements during this period.

At 3 years of age she presented with fatigue, pallor and decreased activity. Her bowel movements were reported as occurring three to four times per day and were soft, light brown in color and sometimes foul smelling. She denied symptoms of pica, easy bruising, hematuria or hematemesis. She had no history of recent travel outside the United States. She had a good appetite and her diet consisted of 3 meals per day plus snacks, and included rice, chicken, sausage, fruits, meat, vegetables and beans. She
drank less than 16 oz milk/day. She had no known allergies and was on no medications. Her family history was notable for iron deficiency anemia in some relatives. She lived with both parents and was not exposed to lead at home.

On physical examination, her weight was 13.2 kg (50th %) and her height was 89.4 cm (10th %). She was noted to be pale, in no acute distress but tachycardic (heart rate = 126 bpm). She was afebrile had a normal respiratory rate. Her chest examination was clear. She had a grade II/VI systolic ejection murmur. Her abdomen was soft, non-tender and non-distended without hepatosplenomegaly. Her extremities showed normal capillary refill and her skin had no rashes, jaundice, petechiae or ecchymoses. Neurologically she was appropriate and alert. Rectal examination revealed no skin tags or fissures and stool tested guaiac positive.

3. What if any laboratory tests are indicated?

Case 2
An 18 year old boy presents with enteral feeding intolerance and abdominal distension. He was born at 28 weeks estimated gestational age and evolved severe necrotizing enterocolitis necessitating multiple operations. After spending the first three years of life in the hospital, he was weaned from parenteral nutrition (PN) and sent home on a combination of oral and G-tube feeds. At age 16 he began to experience weight loss and was eventually placed back on full parenteral nutrition. A recent upper GI with SBFT shows massively dilated small bowel but no obvious obstruction. A barium enema shows an increase in bowel diameter proximal to what appears to be a jejuno-colic anastomosis but contrast flows freely through it.

1. What medical therapies are available?

2. What are the surgical options?

3. In general what are the indications for bowel lengthening and tapering operations?

4. At laparotomy 140 centimeters of jejunum is the total length of small bowel found. It is dilated to 8 centimeters in diameter. What should be done?

5. What are the clinical outcomes following the serial transverse enteroplasty (STEP) operation?

Case 3:
The patient was a 20 mo female with a history of gastroschisis and intestinal resection in the neonatal period (reportedly 66 cm remnant small bowel remaining after surgery in continuity with the colon). She had a 7 month hospital stay in an outside NICU and our center before being sent home on PN under our multidisciplinary team care. Enteral feeds: Partially hydrolyzed 30 cal/oz formula @ 35 ml/hr x 24 hours (92 kcal/kg/day) TPN with Omegaven® 30 mL/hr x 20 hrs (55 kcal/kg/day). She had been following
closely in our Intestinal Rehabilitation Clinic, but then, suddenly in the last 2 months, had multiple new bouts of severe acidosis, and the parent was beginning to ignore usual anticipatory advice of what to do if fevers > 100.3.

1. What are the possible issues going on here?

2. If the CO2 was 15 at the office visit, how would your team proceed?

3. What members of your multidisciplinary team should be enlisted at this point (if not already a part of this child’s case)? What makes up a comprehensive, complete intestinal rehabilitation team?

4. What resources can be sought out for this child/family?

5. Is this a CPS case, or when should CPS become involved?

References:


