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Annals of Pediatrics & Child Health

Case Report

Intestinal Transmesenteric Plication in a Cystic Fibrosis Child with Recurrent Distal Ileal Obstruction Syndrome

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Submitted: 17 August 2017

Accepted: 05 January 2018

Published: 08 January 2018

ISSN: 2373-9312

Keywords

- Cystic fibrosis
- Meconium ileus
- Distal ileal obstruction syndrome
- Child-phillips

Abstract

Recurrent intestinal obstructions are frequently reported in Cystic Fibrosis patients at any age. The most frequent diagnoses are constipation and distal ileal obstruction syndrome, however in previously operated patients, post-surgical adhesions must be considered in the differential diagnosis of recurrent abdominal pain. We report the case and the follow-up of a 4-years-old Cystic Fibrosis child with recurrent intestinal obstruction after neonatal abdominal surgery for meconium ileus. The decision making process to treat symptoms is described. The chosen surgical procedure proved to prevent any further occlusive episode in a ten years follow up.

ABBREVIATIONS

CF: Cystic Fibrosis; DIOS: Distal Ileal Obstruction Syndrome; MI: Meconium Ileus

INTRODUCTION

Cystic Fibrosis (CF) patients frequently suffer from recurrent intestinal sub-occlusive episodes during their lifetime; constipation and distal ileal obstruction syndrome (DIOS) are largely the most common diagnosis. Nevertheless these clinical events are more frequently reported in CF patients who presented at birth with meconium ileus (MI) requiring surgery in the first days of life [1].

MI, a complete intestinal obstruction caused by the intraluminal impaction of dehydrated and thickened meconium, is the earliest manifestation of CF [2]. Hyper echogenic fetal bowel with dilation detected by ultrasound during the second trimester of pregnancy must place the suspect for CF [3].

European CF patients' registry reports an 18% frequency of MI for European CF population in 2009, while a recent European multicenter study showed that about 40% of all CF patients suffering from DIOS presented at birth with MI [4].

These data confirm the high prevalence of DIOS in CF patients with MI as both these conditions probably share the same pathophysiology, such as slow intestinal transit and impaired intestinal secretion flow leading to sticky mucus.

In CF patients who underwent surgery for MI, differential diagnosis should include also postoperative adhesions syndrome, which is reported as a late complication of intraperitoneal surgery with a variable incidence depending both on the primitive disease and the surgical procedure [5].

Every recurrent occlusive episodes in CF/MI patients show grossly the same clinical presentation, making sometimes hard to differentiate between DIOS and a possibly post surgical adherence syndrome.

We report the case of a CF male patient with MI requiring surgery in the first day of life, who presented several intestinal occlusive episodes since the age of two years. An extended adhesiolysis together with a Childs-Phillips procedure (transmesenterical plication) as a prophylaxis against recurrencies solved the episodes of bowel obstruction.

CASE REPORT

L.C. was born at full term with proper weight for gestational age and presented a neonatal intestinal obstruction requiring surgery in the first day of life. Abdominal exploration showed a meconium ileus which required about 20 cm ileal resection with a temporary Bishop-Koop ileostomy.

Cystic Fibrosis wassuspectedby a positive newborn screening program (IRT/DNA) with two CFTR mutations (N1303K/711+1G→T). The infant was therefore refered to our

CF Center since the first weeks of life and the diagnosis of CF with pancreatic insufficiency was confirmed by the positive sweat test and the dosage of fecal pancreatic elastase (<15 mcg/g).

At 5 months of age he underwent again surgery for ileostomy take-down and the postoperative period was burdened by a Pseudomonas aeruginosa infection of the surgical wound.

Since the age of two years the child was admitted to our pediatric department nine times in 25 months, five of which were required for bowel occlusive episodes with abdominal pain, nausea and emesis. All the episodes were classified as constipation or DIOS, and were treated medically.

Despite optimization of pancreatic enzymes replacement therapy, regular use of stool softeners, osmotic polyethylene glycol laxative, and enemas, intestinal occlusive episodes presented with growing frequency.

Careful revision of clinical and radiological history by a committed team showed the presence of a firm bowel loop in several X-rays raising the suspicion of an adhesions syndrome instead of a more common DIOS.

As a consequence we decided to profit of the general anesthesia scheduled to remove bilateral paranasal mucoceles (a CF typical upper airways complication leading to recurrent upper and lower airways infections) to perform an abdominal exploration despite the absence of an acute occlusive episode. At the time of surgery the child was four years old and did not have lung infections or liver disease and the BMI was 10th centile (2000 CDC growth charts).

The procedure started in laparoscopy which showed more peritoneal scars then expected from previous surgeries, and it was converted to open surgery due to technical difficulties and a perforation in a loop which was firmly stuck to the anterior abdominal wall at the site of the previous wound.

All the bowel was mobilized through a transverse incision on the previous laparotomy; a 10 cm tract including the perforation was removed at about 60 cm from ileo-cecal valve and restored with an end-to-end anastomosis. The entire length of the mobilized small bowel was then properly repositioned and a Childs-Phillips procedure was performed, stabilizing the loops from Treitz to ileo-cecal valve with three transmesenteric stitches. The appendix was prophylactically removed and the abdomen closed by layers.

Early postoperative course was uncomplicated, no respiratory nor gastrointestinal symptoms were observed. Since that surgery the child experienced no further intestinal occlusive episodes in the following ten years. During the same period he needed significantly less admissions and medical evaluations (respectively from one every 3.3 months to one every 40 months, and from ten to four visits per year). Growth remained in the normal range, with height over the 50th centile within his genetic target and BMI over the 20th centile.

DISCUSSION

We reported the case of a CF child who had ileal resection in the first day of lifefor MI and ileostomy take-down at 5 months of age, whose recurrent intestinal occlusive episodes dramatically ceased after a surgical procedure to eliminate peritoneal adhesions and to stabilize bowel loops [6].

In the last decades MI patients presented a clear improvement in their prognosis mostly due to progress in medical and surgical procedures, early CF diagnosis and a timely start of both respiratory and nutritional management.

However a worse outcome of MI children is still reported and it can be linked to the sum of gastrointestinal complications possibly afflicting those patients [7]. Actually MI/CF patients are prone to frequent gastrointestinal CF-related complications as DIOS, constipation, liver disease and failure to thrive since the first years of life.Similarly to patients who underwent abdominal surgery for other causes, they may present also late surgical complications as recurrent occlusive courses, postsurgical adherence syndrome or short bowel syndrome. Post surgery adherence syndrome is the consequence of the occlusion of the bowel progression due to scars provoked by inflammatory process on the serosal surface conglutinating the intestine to adjacent structures and causing compression or kinking of one or more intestinal loops.

Factors affecting the severity and width of serosal inflammation usually are considered to worse the risk of adherence syndrome such as diffuse peritonitis, perforations, widespread dissections and bleedings; any of those can burden the surgical procedures for MI in newborns. In our patient the *P. aeruginosa* infection of the previous surgical wound may have played an additive role in entrapping the bowel loops just under the line of the laparotomy.

In MI patients recognizing the real cause of their postoperative occlusive episodes can be difficult because symptoms are the same both for the commonest DIOS and the less frequent adhesions syndrome. In our case clinical history, frequency of the episodes and radiological imaging lead us to the correct diagnosis. In our patient the wide dissection required left the bowel mostly uncovered by serosal surface, which is known as a major risk for occlusion recurrence, thus we decided to perform a Childs-Phillips procedure to rearrange the loops and prevent further obstructions.

Experience of such procedure in pediatric surgery is limited but some literature reports significant results with this extreme option in dealing with adhesions syndrome in adults [8].

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Cite this article

Padoan R, Poli P, Tonegatti L, Falchetti D (2018) Intestinal Transmesenteric Plication in a Cystic Fibrosis Child with Recurrent Distal Ileal Obstruction Syndrome. Ann Pediatr Child Health 6(1): 1138.