

Special Article

Recent Advances on the Surgical Management of Common Paediatric Gastrointestinal Diseases

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Abstract

Diseases of the gastrointestinal (GI) tract remain a major part of the paediatric surgical caseload. Hirschsprung's disease (HSCR) and imperforate anus are two indexed congenital conditions which require specialists' management, while gastro-oesophageal reflux (GOR) is a commonly encountered problem in children. Recent advances in science have further improved our understanding of these conditions at both the genetic and molecular levels. In addition, the increasingly widespread use of laparoscopic techniques has revolutionised the way these conditions are treated in the paediatric population. Here, an updated overview of the pathogenesis of these diseases is provided. Furthermore a review of our experience in the use of laparoscopic approaches in the treatment is discussed.

Key words

Anorectal anomaly; Gastro-oesophageal reflux; Hirschsprung's disease

Introduction

Congenital anomaly of the gastrointestinal (GI) tract is a major category of the paediatric surgical diseases. Conditions such as Hirschsprung's disease (HSCR), imperforate anus and gastro-oesophageal reflux are specific to the paediatric population and require specialists' management. In this article we aim to provide an updated overview of these three index conditions and review our experience in the use of new respective laparoscopic surgical approaches in the treatment of these diseases.

I. Hirschsprung's Disease

HSCR is one of the most common causes of intestinal

obstruction in the neonates. It occurs in about 1 in 5,000 live births.¹ HSCR is characterised by the absence of ganglion cells in the submucosal and myenteric plexuses of the distal bowel, resulting in functional obstruction due to the failure of intestinal relaxation to accommodate the GI contents from the proximal bowel. The rectosigmoid colon is the most common site of involvement.

Around 80-90% of the patients with HSCR present in the neonatal period with symptoms and signs of intestinal obstruction, of which approximately 6-14% present with enterocolitis. However, enterocolitis seems to be less common among Chinese patients (unpublished data, Tam PKH). Clinical presentation may be subdominant. For example, 10-20% of the HSCR patients may present in late childhood or adulthood with chronic constipation, without overt signs of intestinal obstruction; these late presenters often have malnutrition and failure to thrive which are readily reversed upon surgery for HSCR.

HSCR is now recognised as a complex multigenic disorder. The major susceptibility gene for HSCR is RET; mutations of RET are found in 50% of familial cases and 7-35% of sporadic cases.² Other genes involved in HSCR pathogenesis include endothelin receptor B (EDNRB),³ SOX10,⁴ glial cell line derived neurotrophic factor (GDNF).⁵ Most recently, we have identified an association of PHOX2B polymorphisms with HSCR.⁶

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Investigations for diagnosis of HSCR include imaging studies of the lower GI tract and pathologic examination. Studies of barium enema usually, though not always, reveal a transitional zone, i.e., the junction between the proximal dilated and distal collapsed colon. However, the 'gold standard' for diagnosis is rectal biopsy that shows the absence of ganglion cells in the submucosa layer and thickened nerve fibers in either mucosa or submucosa. Anorectal manometry may provide additional information: in the presence of normally functioning ganglion cells, dilatation at the rectum induces relaxation of the internal sphincter, manifested as a drop of the internal sphincteric pressure the rectal inhibitory reflex. Therefore, an increase of the sphincteric pressure in response to rectal dilatation induced by intrarectal balloon inflation strongly indicates absence of intact innervation at the rectoanal region.

Conventional treatment for HSCR is to create initially a diverting colostomy (or ileostomy in the case of total colonic aganglionosis) proximal to the transitional zone to decompress the obstructed intestine and restore enteral feeding. At a later stage, the GI continuity can be reconstructed by a pull-through operation (e.g., Duhamel, Swenson or Soave procedures) to excise the aganglionic segment and bring the ganglionic segment down to the anus. The colostomy can be closed afterwards.

HKU Experience

Laparoscopic Duhamel pull-through procedure has been performed in our centre recently. Between December 1997 and June 2002, 16 children with Hirschsprung's disease, including 10 boys and 6 girls, were treated laparoscopically.⁷ The ages at operation ranged from 2 months to 5 years (median 6 months). One patient was converted to the open method due to severe intra-abdominal adhesions. The ages at operation ranged from 2 months to 5 years (median 6 months). One patient was converted to the open method due to severe intra-abdominal adhesions. A de-functioning stoma had been previously performed on 14 of them. In the early post-operative period, 3 patients had complications related to the procedure, including pelvic abscess, enterocolitis and severe diarrhoea. Compared to previous open procedures, a reduction of postoperative analgesia was noted. The mean follow up time was 30 months (4 to 44 months). All but two (86%) patients had daily spontaneous defecation with an acceptable frequency of bowel openings (1-4x/day).

Our preliminary experience revealed that laparoscopy-assisted Duhamel procedure is a safe and effective treatment option for children suffering from Hirschsprung's disease.

A randomised controlled trial is planned to evaluate its role in replacing the conventional methods.

II. Imperforate Anus

Imperforate anus or congenital anorectal malformation occurs in 1:5000 children,⁸ an incidence similar to that of HD. The relationship between the level of the atretic rectum and the pubococcygeus muscle categorises patients into the "high", "intermediate" and "low" types. Imperforate anus is usually presented in the newborn period during routine physical examination. Subtle malformations such as an ectopic anus and anal stenosis may present later in life. Associated fistula connecting the rectum and genitourinary tract may present with passage of stool at the urethral or vaginal opening (commonly recto-urethral fistula in males and rectovaginal fistula in female). Imperforate anus may occur independently, but not infrequently is it associated with anomalies of the other organs, such as vertebrae, heart, kidney, trachea, and bones etc. (VACTERL syndrome).

Invertogram (plain X-ray with the baby prone and its pelvis raised) usually gives the distance between the most distal rectal gas bubble and the perineum. CT scan⁹ and MRI¹⁰ provide clear information on the relationship between the atretic part and the intrapelvis sphincteric muscles, such as levator ani and puborectalis, and sometimes the fistula tract may also be delineated.

An ectopic anus usually is not associated with a rectal fistula, so treatment with a simple Y-V anoplasty and anal dilatation may be adequate. However, a colostomy is required for all patients of imperforate anus without an anal opening, followed by anorectal reconstruction at a later stage. Methods for the repair of high- and intermediate-type imperforate anus have evolved from abdomino-perineal pull-through and sacroperineal pull-through or their combination,¹¹⁻¹³ posterior sagittal anorectoplasty (PSARP),¹⁴ to the recently reported laparoscopy-assisted pull-through anorectoplasty (LAR).¹⁵ All these procedures aim at establishing a normal anatomical outlook and socially acceptable continence. To achieve this, precise placement of the pull-through recto-colonic segment within the center of the sphincteric complex and minimal damage of the intrapelvic muscle floor and its innervation are prerequisites.

Compared with abdomino-perineal and sacro-perineal approaches, PSARP is thought to be able to provide better continence as the reconstruction of pelvic muscles and external sphincter complex are performed under direct vision. However, long-term continence after PSARP is less

than ideal.^{16,17} For example, only about 25% of boys with a rectoprostatic fistula have normal continence and significant soiling occurred in more than half of this patient population. Recently, laparoscopy-assisted LAR has been designed and appears to provide surgical advantages of excellent visualisation of the rectal fistula and accurate placement of the pull-through segment at the center of levator sling without the need to divide the muscles controlling continence, which is inevitable in PSARP.

HKU Experience

We have performed laparoscopy-assisted anorectoplasty on nine patients (6 males and 3 females) from July 2000 to March 2001 one to six months after creation of diverting colostomy. The procedures were as described by Georgeson et al¹⁵ with minor modifications. Briefly, after induction of anaesthesia, pneumoperitoneum was created to a pressure of 8 to 10 cmH₂O using a 5-mm trocar (Endopath, Johnson & Johnson, USA) inserted through a semilunar skin incision along the inferior margin of the umbilicus. A laparoscope was introduced through the umbilical trocar, and two to three additional 5-mm trocars were inserted for use as working ports, one at the left lower quadrant and one or two at the right lower quadrant. Laparoscopic dissection was begun at the peritoneal reflection by hook cautery and a harmonic scapel (Ethicon, Endosurgery, Cincinnati, OH, USA) and the rectal wall was dissected circumferentially. The rectourethral or rectovaginal fistula was clip-ligated at its insertion at the posterior urethra or vagina, and was divided sharply. The levator ani sling was then readily identified in the center of the pelvic cavity under laparoscopic visualisation. The center of external sphincter complex was determined by electrostimulation with a Pena muscle stimulator in the perineum, and a cruciate skin incision was made. A Veress needle was advanced through the center of the external sphincter complex under the combined guidance of perineal electrostimulation and telescopic light shining downward from inside until the tip of the needle emerges into the pelvic center. A self-expanding 10-mm port (Steptrocar, Ethicon, USA) was introduced through this tract. The colon was pulled down through this tunnel and an anoplasty was fashioned accordingly.

Our preliminary results show that in the early post-operative stage, patients repaired with LAR had more favourable findings in anorectal manometry than patients repaired with PSARP.¹⁸ Long-term follow-up studies to confirm a superior defecation continence achieved with LAR is being conducted.

III. Gastro-oesophageal Reflux (GOR)

GOR results from inadequate sphincteric function at the gastro-oesophagus junction to prevent reflux of gastric contents into oesophagus. Most paediatric patients with GOR have other underlying abnormalities. Around 75% of our patients receiving anti-reflux surgery had associated medical problems, among which cerebral palsy was the most common. Symptoms presented in older children are the same as adults with heartburn, epigastric pain, waterbrash or acid brash. By contrast, infants usually present with persistent vomiting or regurgitation, recurrent aspiration pneumonia or feeding problems with failure to thrive.

The diagnosis relies on the clinical history and the demonstration of reflux of stomach contents into the oesophagus, either by a 24-hour pH study or by a radioisotope labelled milk scan. These examinations are particularly important for children with atypical symptoms to determine the severity of GOR and to confirm that GOR is responsible for atypical presentation. Upper endoscopy can demonstrate acid induced oesophagitis or Barrett's oesophagus. Barium swallow study is useful to exclude other congenital malformations and provide information on gastric emptying.

Treatment of GOR starts with modifications of feeding style. Medications such as antacids, alginic acid, histamine receptor antagonists or proton pump inhibitors are commonly used and may alleviate symptoms. Indications for surgical treatment include persistent symptoms despite medications, Barrett's oesophagus, recurrent aspiration pneumonia and failure to thrive.

The principles of anti-reflux surgery are to correct anatomical abnormalities such as a hiatal hernia and to augment the resistance to reflux at the gastro-oesophageal junction while preserving the patient's ability to swallow normally. Several procedures for anti-reflux surgery are practiced, and the Nissen fundoplication is the most widely used.¹⁹ The use of laparoscopic Nissen fundoplication for gastro-oesophageal reflux disease is well established in adults. This procedure involves wrapping the gastric fundus around the intra-abdominal oesophagus and the gastro-oesophageal junction laparoscopically. Usually 4 trocars are required for manipulation and a 30-degree telescope is used for visualisation. The 5-mm trocars are used in paediatric patients. The benefits of the procedure include better cosmetic outcome, early resumption of enteral feeding, better post-operative pain control and minimising intra-abdominal adhesion. Funduplications are considered

more risky in the paediatric population as most of them have underlying neurological diseases with seizure disorders associated with oesophageal dysmotility. This may contribute to a higher incidence wrap disruption.²⁰ Therefore laparoscopic fundoplication was not done in paediatric population until around ten years ago. However, the benefits of laparoscopic procedure over the open method are definite,²¹ especially in decreasing the incidence of intra-abdominal adhesion in children. Therefore laparoscopic Nissen fundoplication has been gaining in popularity among this patient group.

HKU Experience

From June 1998 to the end of 2002, a total of seventeen patients with GER in our centre underwent laparoscopic Nissen fundoplication, accounting for 70% of all the anti-reflux surgery performed during this period. There were 13 boys and 4 girls. The average age at the procedure was 7 years 9 months. The youngest patient was only 9 months old while the oldest was 20 years old. Indications for surgery included severe symptoms of vomiting or regurgitation (10 patients), failure to thrive (8 patients) and recurrent aspiration pneumonia (7 patients). Many patients had overlapping symptoms. Thirteen patients (76.5%) also had other underlying abnormalities. Eight patients had cerebral palsy. Two patients had degenerative brain disease. One patient had tracheo-oesophageal fistula with resection of the fistula and primary anastomosis done. One had chromosomal abnormality and one had non-specific developmental delay. All patients underwent 24-hour pH study to confirm the diagnosis of gastro-oesophageal reflux before definitive surgery.

The mean operative duration was 180 minutes (120 minutes for open fundoplication). Three patients (17.6%) needed conversion to open fundoplication. In one of the patients, it was due to difficulty in visualising the gastro-oesophageal junction at laparoscopy. Another conversion was needed because the operative field was obscured by a loop of dilated bowel. The most recent conversion was due to multiple adhesions as a result of previous laparotomy in a child with previous necrotizing enterocolitis. There were four postoperative complications (23.5%). One patient had wound infection, which resolved with antibiotic treatment. Another two patients had persistent symptoms. One of them was later found to have achalasia. The other patient with persistent symptoms finally required a redo fundoplication. Intra-operatively it was found that there was a small hiatal defect between the right crus and the oesophagus. The patient became symptom free after the redo operation. The

fourth complication was due to too tight a wrap around the gastro-oesophageal junction. The patient presented with dysphagia post operatively. Dilatation of the gastro-oesophageal junction was done but symptoms did not resolve. Finally a redo operation was decided with subsequent good outcome. Overall, failure rate of laparoscopic fundoplication to provide symptomatic relief was only 11.8%. The rate for redo fundoplication was also 11.8%. In conclusion, our initial experience with laparoscopic fundoplication was encouraging. Both the operative time as well as the specific complication rate was comparable to other major centers in the world.²² Furthermore, the majority of patients became symptom free after the surgery. The complication rate and operative duration will further improve as our technique matures. We therefore envisage the continued widespread use of this technique in the paediatric population.

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