June 2012

Duhamel's Procedure for Adult Hirschsprung's Disease

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INTRODUCTION
Hirschsprung’s disease (HD) is a congenital aganglionosis of submucosal and myenteric neural plexuses, principally affecting distal sigmoid colon and rectum. It is usually recognized in first few years of life, but can occasionally be encountered in adults as misdiagnosed cause of refractory constipation. The term “Adult Hirschsprung’s Disease” has been arbitrarily applied to cases in which patient is more than 10 years of age when diagnosis is first established. The first well-documented case of Adult HD was reported in 1950s by Rosin. In these cases, disease may be of milder form and may go unrecognized, until the proximal colon can no longer propel faeces distally, leading to faecal retention and/or intestinal obstruction. These patients usually have typical medical history of life-long chronic constipation, with very frequent use of enemas and cathartics for relief of constipation since early childhood. Definitive diagnosis is established on full-thickness rectal biopsy; barium enema and manometric studies being useful investigations. Duhamel’s procedure is recognized as most reliable and safe surgical procedure for treatment of Adult HD with excellent long-term functional outcome.

We report a case of adult Hirschsprung’s disease successfully treated with Duhamel’s procedure.

CASE REPORT
A 17-year-old male presented in outpatients’ clinic at the Aga Khan University Hospital (AKUH), Karachi, Pakistan, in October 2003, with abdominal mass for one year and chronic constipation since early childhood, requiring frequent use of enemas for relief. He had been seen by many doctors for evaluation of chronic constipation during early childhood and thereafter, but no diagnosis could be established. He was therefore, admitted at AKUH for evaluation. On examination, abdomen was distented, and a firm, non-tender, hemispherical mass measuring 12 cm x 14 cm was palpable in umbilical and hypogastric region. Bowel sounds were absent.

Plain abdominal radiograph revealed a well circumscribed ovoid mass in the left hemi-abdomen, with well-defined margins. Barium enema study revealed narrowed distal sigmoid colon with significant proximal sigmoid dilatation with faecal loading. A computed tomographic scan confirmed severe colonic dilatation with extensive faecal loading. At this stage, the cause of chronic constipation was obscure. However, clinical features, radiographs and barium enema studies were consistent with Hirschsprung’s disease. Therefore, with a provisional diagnosis of HD, faecal washout with full-thickness rectal biopsy was planned. However, patient was lost to follow-up for 3 years.

In July 2007, patient presented with absolute constipation, non-bilious vomiting, abdominal pain and distension in the emergency room. On examination, the abdomen was distended, diffusely tender and a firm, hemispherical mass was palpable in umbilical and hypogastric region. Bowel sounds were absent. Digital rectal examination revealed empty rectum. Plain abdominal radiographs re-demonstrated very large descending and sigmoid colon with extensive faecal loading (arrow, Figure 1). Patient was kept on low-volume nasogastric suction and enemas to relieve constipation.

Once constipation was relieved, exploratory laparotomy with resection of affected sigmoid colon, and end colostomy were performed. A huge phytobezoar...
measuring 15 cm x 12 cm was found in the left hypo-
chondrium. An intussusception was noted in proximal
jejunum, which was reduced. Sequential rectal biopsies
were obtained during the procedure, which revealed
absence of ganglion cells in affected segments, thus
confirming the diagnosis of HD. Two months later,
Duhamel’s procedure with a diverting loop ileostomy
was performed successfully.

Eight weeks following Duhamel’s procedure, a barium
enema study revealed an intact rectal pouch (blue
arrow; Figure 2), with no evidence of leakage from either
the rectal pouch or anastomosis (red arrow; Figure 2).
Thus, diverting loop ileostomy was successfully
reversed. Sigmoidoscopy and anal dilatation were also
performed.

Patient was followed-up weekly initially, and monthly
thereafter. At the time of last follow-up 3 years after the
procedure, patient had no bowel complaints and
reported exceptional improvement in bowel function and
quality of life.

DISCUSSION
The diagnosis of Hirschsprung’s disease in adults is
much more challenging than in infants owing to rarity of
the disease, and higher incidence of short/ultra-short
segment aganglionosis in adults.1,2 A typical adult
patient with HD has history of long-term constipation
since infancy or early childhood; other symptoms include
abdominal pain/discomfort and distention.1,2

There is no obvious best choice of surgical procedure for
Adult HD, and surgical outcome depends as much on
the skills of multiple surgeons involved, as on chosen
surgical procedure involving variety of techniques.6

Principal surgical procedures for Hirschsprung’s disease
have been described by Soave and Duhamel.7,8

Duhamel’s recto-rectal pull-through procedure has
excellent results in adult patients with much lower
incidence of postoperative complications.6,8 The primary
complication is anastomotic leakage,6 leading to fistula
or abscess formation. This procedure also has
advantage of minimal pelvic dissection.6 The literature
concentrating on adult follow-up of patients with HD who
underwent Duhamel’s procedure is scarce, and it has
been reported that long-term outcome is quite variable,
and that functional outcome improves as duration of
follow-up increases.4,5 Duhamel’s procedure is most
widely accepted surgical procedure, provides better
results in adult HD and is also associated with lower
post-operative morbidity rates.1-5

In this case, clinical features were similar to those
reported in literature. It is still unclear whether adult HD
is due to failure of diagnosis when patients present with
chronic constipation in early childhood, or in fact a late-
onset disease.1-3,9 Of note is that the literature reveals
that most patients diagnosed with adult HD fall under the
former category.1-3,9

In addition to refractory long-standing constipation, our
patient also presented with abdominal distension, and a
firm, hemispherical abdominal mass. Duhamel’s
procedure was chosen over other procedures described
in literature due to its widely accepted, good long-term
functional outcome and much lower incidence of
postoperative complications. More importantly, it also
meant minimal pelvic dissection to be done in a young
male patient, consequently sparing pelvic nerves. The
outcome of Duhamel’s procedure was excellent in our
patient with complete resolution of symptoms and
dramatic improvement in bowel function.

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