Case Series

Modified Duhamel with lateral anal sphincterotomy and coloanal stump for adult Hirschsprung’s disease: A case series

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A B S T R A C T

INTRODUCTION: Hirschsprung’s disease (HSCR) or megalacolon congenital is the most common congenital intestinal motility disorder and characterized by the absence of ganglion cells (aganglionosis) in the myenteric plexus and submucosa of the distal intestine.

PRESENTATION OF CASE: This study reports three cases of adult HSCR, with all young female patients who underwent colostomy for obstructive ileus. The chosen definitive therapies were Duhamel pull-through procedure combined with a temporary coloanal stump.

DISCUSSION: The three patients underwent Duhamel pull-through procedure with temporary anal stump in conjunction with stoma reversal. The temporary anal stumps were removed within 1–2 weeks after pull-through procedure. All patients were discharged from the hospital and underwent routine follow-up. All patients had fecal incontinence in early follow-up which resolved shortly afterwards. Long term follow-up showed normal intestinal functions and good cosmetic results.

CONCLUSIONS: The combination of Duhamel pull-through procedure with temporary coloanal stump in definitive therapy of adults with HSCR is a safe and effective technique.

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1. Introduction

Hirschsprung’s disease (HSCR) or congenital megacolon is the most common congenital intestinal motility disorder [1]. This condition is characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine, which makes fecal accumulation impact in the intestines proximal to the aganglionic rectosigmoid segment, resulting in passage blockage [1,2]. Adult cases of this condition are relatively rare, with only about 300 cases reported prior to 2016, mostly found in males (male:female ratio of 4:1) and associated with trisomy 21 or other genetic syndromes [3,4].

The therapy of HSCR aims to bring the ganglionic segment of the bowel to the dentate line, either by pull-through procedure or anastomosis of ganglionic segments. There are three procedures commonly performed in this case: Swenson, Soave, and Duhamel pull-through procedures. To this date, there are no long-term studies that compare complication rates and long-term outcomes of these procedures [1]. Therefore, in this study, we report three cases of HSCR in adults treated with Duhamel procedure combined with lateral anal sphincterotomy and temporary anal stumps. This study is reported in line with PROCESS checklist [5].

2. Case presentation

We present three cases of adult HSCR from 19, 20, and 28-years-old female patients. All patients underwent urgent colostomy diversion due to obstructive ileus. The definitive therapies were Duhamel pull-through procedure with lateral anal sphincterotomy and temporary anal stumps. All cases showed good results of intestinal function and anal appearance with mild fecal incontinence complaints at early follow-up which resolved shortly.

2.1. Case 1

A 27-year-old female came to the hospital with sudden inability to defecate and bloating. The patient was able to defecate every five days routinely before the complaint. Colon-in-loop (CIL) examination showed short segment Hirschsprung disease (Fig. 1a). The patient underwent Hartmann procedure and biopsy (Fig. 1b). Pathological examination showed no Meisner plexus nor ganglion and only a few Auerbach plexuses without ganglion.
Six months after colostomy, the patient was readmitted for stoma reversal and coloanal pull-through (Fig. 1c). The patient was treated in the hospital for five days after the two procedures before being discharged. Two weeks later, the patient was readmitted to undergo coloanal stump removal (Fig. 1d). Stump tissue pathology examination showed sufficient Auerbach plexuses and ganglions, and sufficient Meisner plexuses and a few ganglions. The patient was treated in the hospital for three days after stump removal before being discharged. At two weeks follow-up, the patient had fecal incontinence complaint which resolved shortly afterwards and mild anal stricture which resolved after rectal dilatation.

In the further follow-up, there were no complaints. The patient showed normal function of the intestinal tract after 2 years of follow-up and the anus looked normal (Fig. 1e).

2.2. Case 2

A 15-year-old female came to the hospital with inability to defecate for the last 20 days. CIL examination showed short segment Hirschsprung disease (Fig. 2a). The patient underwent sigmoid colostomy and biopsy (Fig. 2b). Pathological examination showed no Meisner plexus nor ganglion and only a few Auerbach plexuses without ganglion.

Eight months after colostomy, the patient was readmitted for stoma reversal and coloanal pull-through (Fig. 2c). The patient was treated in the hospital for 10 days after the two procedures before she underwent coloanal stump removal (Fig. 2d). Stump tissue pathology examination showed several Auerbach plexuses and ganglions, and several Meisner plexuses and a few ganglions. The patient was treated in the hospital for four days after stump removal before being discharged. At 20 days follow-up, the patient had fecal incontinence complaint which resolved shortly afterwards. In the further follow-up, there were no complaints. The patient showed normal function of the intestinal tract after 3 years of follow-up and the anus looked normal (Fig. 2e).

2.3. Case 3

An 18-year-old female came to the hospital with inability to defecate for the last 15 days and bloating. The patient was able to defecate every five days routinely before the complaint. CIL examination showed short segment Hirschsprung disease (Fig. 3a). The patient underwent sigmoid colostomy and biopsy (Fig. 3b). Pathological examination showed no Meisner plexus nor ganglion and only a few Auerbach plexuses without ganglion.

Five months after colostomy, the patient was readmitted for stoma reversal and coloanal pull-through (Fig. 3c). The patient was treated in the hospital for 11 days after the two procedures before she underwent coloanal stump removal (Fig. 3d). Stump tissue pathology examination showed several Auerbach plexuses and ganglions, and several Meisner plexuses and a few ganglions. The patient was treated in the hospital for four days after stump removal before being discharged. At 3 weeks follow-up, the patient had fecal incontinence complaint which resolved shortly afterwards. In the further follow-up, there were no complaints. The patient showed normal function of the intestinal tract after 3 months of follow-up and the anus looked normal (Fig. 3e).

Surgical procedures
All surgeries of presented cases were done in the following general steps:

i. We did exploratory laparotomy and adhesiolysis procedure due to previous surgery.
ii. We identified and ligated the inferior mesenteric artery and vein.
iii. Incision was made on the white line, splenic flexure, and gastrocolic to mobilize left colon. Intestine vascularization from the artery of Drummond was maintained.
iv. We performed lateral anal sphincterotomy and dilatation to loosen the sphincter tonus and anal opening (b) (Fig. 4a and c).
v. We dissected the distal colostomy segment until posterior/sacral side of the anal floor (c→a) and removed the aganglionic intestine closer to the pull-through opening (e) (Fig. 4a).
vi. The proximal colostomy segment was pulled through to the posterior side of rectum. Transanal excision was done to the rectum with diameter 2 times the pulled-through intestine diameter (a→c) (Fig. 4a). The intestine was pulled until at least 10 cm outside the anal canal to prevent retraction (d) (Fig. 4b).

vii. Three-step suturing was performed to the anastomose proximal and distal segment between the distal rectum and anal canal using absorbable multifilament 2.0.
viii. Ten days after initial procedures, we did the stump removal procedure:
ix. The stump was cut at the 12 o’clock point to the cranial direction until anal canal. Suture was made from the mucous layer to the serous layer at the end of the cut.
x. From the end of the cut, stump removal was done circularly in both left and right directions until 6 o’clock point.
xi. Simple sutures were made from the mucous layer to the serous layer at 8 points along the circular cut.

3. Discussion

Hirschsprung’s disease (HSCR) is a congenital disease characterized by the absence of ganglion cells in the distal colon leading to fecal obstruction due to accumulation of fecal mass proximal to the aganglionic segment. In some cases, this condition might remain until adolescence or adulthood due to compensation of hypertrophic normal bowel segment proximal to the aganglionic bowel with recurrent complaints of constipation, abdominal distension, or abdominal pain [6,7]. In China, adult cases of HSCR are defined as HSCR that occurred in the population above 14 years old, and the treatment can be based on the HSCR treatment in children with some adaptation [3,7]. The gold standard of HSCR diagnosis is full thickness biopsy showing the absence of ganglion cells in submucosa layer or the increasing activity of acetylcholinesterase. However, this kind of examination is invasive and inconvenient for patients. The rectal suction biopsy is one of the alternative examinations which show equal sensitivity and specificity compared to the previous examination [8].

In this study, all three cases of adult HSCR involved females. An urgent colostomy diversion was performed due to repeated constipation caused by impacted stool leading to obstruction and ileus. The definitive therapies of all cases were performed with Duhamel pull-through procedure combined with lateral anal sphincterotomy and temporary anal stumps. The stumps were removed 10–14 days after pull-through surgery. After stump removal, the patients were observed for 3–4 days and no abnormalities were found. In early follow-ups which were done 2–3 weeks after the procedures, all patients had fecal incontinence complaints and one patient had mild anal stricture. All fecal incontinence resolved shortly afterwards and the anal stricture problem was solved by rectal dilatation. Long term follow-up after 2–3 years showed normal intestinal function without complaints and good cosmetic appearance of anus which looked normal. Long term follow-up of case 3 was limited to 3 months due to the recent date of procedure, but the patient had no further complaints, good intestinal function, and good anus appearance.

All the HSCR cases in this study were treated with modified Duhamel with lateral anal sphincterotomy and temporary anal stump procedures. In the Duhamel technique, the ganglionic colon...
segment is pulled through the posterior wall of the rectum at the anal floor to the anal canal and the rectum and colon are anastomosed using a crushing clamp, thus preserving the aganglionic rectum to act as a pouch reservoir [9]. In this technique, the ganglionic segment can be anastomosed as close as possible to the anal canal, thus reducing the risk of recurrence.

Traditionally, temporary ileostomy is made to divert fecal mass in order to protect the new anastomoses after the procedure. The purpose of fecal diversion is to lower the risk of leakage, fistula, and wound infection of the anastomoses [10,11]. However, the ileostomy has some disadvantages, including risk of dehydration, electrolytes imbalance, possible malnutrition due to high fluid output, patient discomfort, psychological impact, and other daily life disturbances. In addition, the temporary ileostomy is maintained until 8–12 weeks before being reversed. The use of temporary coloanal stumps instead of diverting ileostomy in this study were able to help patients to avoid ileostomy disadvantages while accelerating recoveries with faster stump removal, thereby reducing the risk of retraction, and optimizing post-operative defecation function.

There are other procedures to treat HSCR available, such as Soave and Swenson procedures. Soave procedure is done by removing rectal mucosa while retaining the muscular cuff, then anastomose the ganglionic colon to the anal canal mucosa. Swenson procedure is done by removing aganglionic rectum, pulling the ganglionated colon, and connecting it to the anus. This procedure involves mucosal biopsies assessed by frozen sections to determine the ganglionicated segment [12]. One of the recurrence factors of HSCR is the remaining aganglionic section due to insufficient removal at the surgery, resulting in the need for reoperation. For this reason, the availability of a frozen section facility is an important factor in the consideration of HSCR treatment options [13]. Due to our current study is still in the form of case series and the evidence of its effectiveness is limited, further study with larger sample size is needed to confirm and clarify our findings.

4. Conclusions

Duhamel combined with temporary coloanal stump is a practical and efficient option for the treatment of Hirschsprung’s disease in adults. We believe that this procedure gives satisfactory results and remains well suited to the adult with Hirschsprung’s disease. The findings of this study indicated that Duhamel and temporary coloanal stumps in definitive therapy for adult patients with Hirschsprung’s disease are safe, effective, and able to reduce the risk of recurrence.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

The informed consent form was declared that patient data or samples will be used for educational or research purposes. Our institutional review board also do not provide an ethical approval in the form of case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Authors contribution

Adedejus Yuda Handaya conceived the study. Aditya Rifqi Fauzi drafted the manuscript. Ahmad Shafa Hanif and Joshua Andrew critically revised the manuscript for important intellectual content. All authors facilitated all project-related tasks.

Registration of research studies

The manuscript is a case report, not considered a formal research involving participants.

Guarantor

Adedejus Yuda Handaya.

Provenance and peer review

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