Pull-through Procedure for Hirschsprung’s Disease with Simultaneous Stoma Closure: Insights for Limited-Resource Settings from Mbarara, Uganda

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Research Article

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Abstract

**Background:** In many resource-limited settings, patients with Hirschsprung’s Disease (HD) undergo initial diverting colostomy, followed by pull-through, and lastly, colostomy closure. This approach allows for decompression of dilated and thickened bowel and improved patient nutritional status. However, this 3-stage approach prolongs treatment duration, with significant stoma morbidity, costs, and impact on quality of life (QOL).

**Aim:** To determine whether pull-through for HD can safely be performed with simultaneous stoma closure, reducing treatment approach from three to two stages.

**Patients and Methods:** Children with HD and diverting colostomy were prospectively followed as they underwent pull-through with simultaneous stoma closure. Their in-hospital course, and 3-month outpatient course, were assessed for postoperative complications. Patients with total colonic HD, redo pull-through, and residual dilated colon were excluded from the study.

**Results:** Of the 20 children, seventeen were male (n = 17, 85%). All patients had rectosigmoid HD. The median weight, age at colostomy formation, and age at pull-through were 11.05 kg (interquartile range [IQR] 10-12.75), 0.9 years (IQR 0.25-2.8), and 2.08 years (IQR 1.28-2.75) respectively. Mean duration with colostomy before pull-through was 1.1 years (SD 1.51). Median hospital length of stay was 6 days (IQR 5-7). Early complications included anastomotic leak (n=1), perianal skin excoriation (n=2), surgical site skin infection (n=3), and burst abdomen (n=1). Longer-term complications included stricture (n=1, 5%) and enterocolitis (n=2, 10%).

**Conclusion:** In this small case series, we have demonstrated that pull-through with simultaneous stoma closure can be safely performed in resource-constrained settings. Further studies are needed to understand the QOL and economic impact of this change in management for HD patients.

Introduction

Hirschsprung disease (HD), also known as congenital megacolon, is characterized by the absence of ganglion cells in the myenteric and submucosal plexuses in distal bowel and extending proximally for varying distances [1][2][3], with an incidence of 1:5000 live births and more commonly in boys than girls with a ratio of 4:1 [3][4]. The ‘transition zone’ between normal and aganglionic bowel lies in the rectum or rectosigmoid colon in over 75–80% cases, while 10–17% have more proximal colonic involvement, and 5–10% have total colonic aganglionosis with variable involvement of the distal small intestine [1][3][5]. Patients may present during the neonatal period or childhood. Neonatal presentation includes delayed passage of meconium beyond 24 hours, abdominal distension, bilious vomiting, feeding intolerance, and Hirschsprung-associated enterocolitis (HAEC) [1]. Childhood presentation frequently involves constipation, history of delayed passage of meconium at birth, failure to thrive, abdominal distention, and dependence on enemas [1][5][6]. Patients in low- and middle-income countries (LMICs) have a higher rate
of delayed presentation, with the attendant complications, compared to high-income countries (HICs), where one stage pull-through is typically performed in infancy.

It is safer in older children with delayed presentation to delay pull-through until the extremely dilated colonic diameter has decreased sufficiently to perform a safe procedure. This may take weeks or months by irrigation, or by initial diverting colostomy [1][5]. This is an acceptable standard of care in our setting given the late presentation. It also gives time to improve the nutritional status of children who present with malnutrition and failure to thrive. At the pull-through operation, the majority of children are subjected to another proximal stoma to protect the anastomosis and allow for early oral feeding since many LMIC settings lack parenteral nutrition. Patients with a diverting proximal stoma will subsequently need another major operation to reverse the stoma. This subjects them to potential future complications such as prolapse, retraction, skin breakdown, and other issues causing significant morbidity and cost [7][8][9] especially in low income countries where stoma appliances and stoma care specialists are lacking. In addition, elective lists involving stoma related surgeries may get cancelled due to priority of emergency cases, upper respiratory infections at the time of planned surgery, insufficient theatre time and space, blood availability, and other anesthesia related factors [10][11][12][13][14][15]. This all translates into prolonged time living with stomas, increasing the waiting period for the patients, frustration and mental stress to the patients and their families, and increased cost and waste of hospital consumables.

Stomas have become over time a significant lifesaving treatment option in several gastrointestinal conditions [16]. However, these stomas are not without complications themselves. Neonatal stomas are frequently placed in HD to decompress dilated large bowel [17] and can cause significant complications [18][19]. Stomas impact negatively on the patient's quality of life (QoL). These include: change of body image, loss of control of flatus and fecal matter, stoma care, among others [20][21][22][23][24][25]. In Uganda stoma care and management is limited with scarce and expensive stoma appliances only affordable by selected few. The presence of a stoma limits social life interactions such as recreational sports, leisure activities, public transport. Affected persons feel embarrassed and physically restrained [26][27]. Stomas further limit interpersonal relations of children with their friends, relatives and family members. Most of these children delay starting school, returning to school or completely drop out of school [19][28][29] and their fathers leave families [30]. As a result, any change in our standard of care for HD patients that reduces the time with stomas, or the number of operations required, can have a significant impact on patient outcomes, as well as quality of life. This study sought to assess the effectiveness of pull-through with simultaneous stoma closure to minimize the challenges of stoma care faced by parents as well as to maximize the limited pediatric surgical services in the country.

**Methods**

Having observed challenges of stomas in our setting, we decided to pilot the Swenson pull-through procedure with simultaneous stoma closure and to assess the associated morbidity and mortality such as anastomotic leakage, bowel obstruction, wound dehiscence, incisional hernia, longer hospital stays, and deaths [2][31]; and the socioeconomic challenges faced by most of our patients. Here we are
reporting a prospectively followed consecutive case series. Ethical approval was obtained from Mbarara University research ethics committee # 02/05–19. Patients with total colonic HD, those for redo pull-through, and those with dilated colon upstream from stoma at surgery were excluded from the study. We monitored all patients closely and were prepared to place a diverting stoma in case of any complications. We do not have pathology laboratory available to send a biopsy for frozen section to confirm presence of ganglion cells when a transition zone is encountered intra-operatively. We relied on two main things: 1) once the temporary diverting stoma is functional, this confirmed we were above transition zone. 2) pulled through stoma, but took an additional 5–10 cm above that level to ensure that transition zone wasn’t involved in the pull-through before water tight two layer anastomosis was performed.

All cases were performed by the author and his supervisors. Patients were nil per os as instructed by anesthesia provider before surgery. No bowel preparation was done. A combined abdominal and trans-anal (perineal) approach was taken and the Swenson pull-through procedure was performed in all cases [32][33]. Figures 1 & 2

Patients were kept nil per os for 3–5 days while initiating oral feeds as the child tolerated feeds and generally discharged on days 5–7. Those with complications stayed longer.

The children were followed in outpatient clinic after discharge at 2 weeks, 1 month, and 3 months. They were assessed for early postoperative complications (anastomotic leakage, anal strictures, and perineal excoriations), and late complications (ongoing obstructive symptoms, frequent bowel movements, incontinence, soiling, and enterocolitis) [34][1][35][36], and death. The parents were educated about signs and symptoms of postoperative enterocolitis, since this can result in rapid severe illness and death.

Results

Of the 20 children recruited, seventeen were male (n = 17, 85%) and three were female (n = 3, 15%). All patients had rectosigmoid Hirschsprung’s disease. The median weight, age at colostomy formation, and age at pull-through were 11.05 kg (Interquartile Range [IQR] 10-12.75), 0.9 years (IQR 0.25–2.8), 2.08 years (IQR 1.28–2.75) respectively. Mean hemoglobin (Hb) was 11.02mg/dl (SD 2.31) and mean duration with colostomy before pull-through was 1.1 years (SD 1.51) Table 1.
Table 1

| Characteristics                        | Median (interquartile range) | Mean  | SD    |
|----------------------------------------|-----------------------------|-------|-------|
| Weight (Kg)                            | 11.05 (10-12.75)            | 12.8  | 5.12  |
| Hemoglobin (g/dl)                      | 11.55 (9.4–12.9)            | 11.02 | 2.31  |
| Age at Colostomy (years)               | 0.9(0.25–2.8)               | 1.56  | 2.6   |
| Duration of colostomy (years)          |                             | 1.175 | 1.51  |
| Age at Pull-through (years)            | 2.08(1.28–2.75)             | 2.7   | 2.7   |
| Duration of surgery (hours)            | 3.17 (2.91–3.79)            | 3.35  | 0.65  |
| Gender                                 |                             |       |       |
| Male (n = 17, 85%)                     |                             |       |       |
| Female (n = 3, 15%)                    |                             |       |       |

Thirteen had colostomy creation performed at our facility (Mbarara Regional Referral Hospital [MRRH]) while 7 were at others outside facilities. All colostomies performed at MRRH were leveling colostomies. Of the stomas placed elsewhere three had end stomas and 2 had transverse supraumbilical incisions during stoma placement. Four children were transfused (20%) post-operatively, all patients got intravenous paracetamol for pain control and intravenous metronidazole plus ceftriaxone as antibiotics. All patients were switched to oral paracetamol and ampiclox (ampicillin-cloxacillin) once oral feeds were started. Antibiotics were continued for five days postoperatively. One patient went to pediatric intensive care unit (PICU) due to hemodynamic instability after anastomotic leak, had a diverting ileostomy placed, and recovered uneventfully.

The mean duration of surgery was 3.3 hours (SD 0.65), minimum 2.5 hours and maximum 5 hours. The median length of hospital stay was 6 days (5–7), minimum stay 3-days and maximum stay 15-days.

**Complications:**

During the post-operative period one child had an anastomotic leak, 2 had perianal skin excoriation, 3 had a surgical site skin infection (SSI) of the stoma site and 1 had burst abdomen Table 2.
Table 2

| Postoperative complications | Frequency | Percentage (%) |
|-----------------------------|-----------|----------------|
| Anastomotic leak            | 1         | 5.0            |
| Perineal excoriation        | 2         | 10.0           |
| SSI                         | 3         | 15.0           |
| Burst abdomen               | 1         | 5.0            |

SSI were treated with daily dressing, burst abdomen was returned to the operating room (OR) for second look and closure and recovered uneventfully. Anastomotic leak was recognized on the fifth post-operative day, and proximal diversion was performed.

The majority of children were on nil by mouth (NPO) for 2 days (n = 12, 60%), they had clear fluids on day three and breast milk for those still breastfeeding (n = 5, 20%). Soft feeds were started on day four (n = 10, 50%) and full feeds were mostly given on day five (n = 17, 85%) Table 3.

Table 3

| Days   | NPO | Clear fluids | Breastmilk | Soft feeds | Full feeds |
|--------|-----|--------------|------------|------------|------------|
|        | Frequency | %   | Frequency | %   | Frequency | % | Frequency | % | Frequency | % | Frequency | % |
| ≤ 2 days | 12    | 60  | 9         | 45  | 0         | 0 | 0         | 0 | 0         | 0 | 0         | 0 |
| 3–5 days | 8     | 40  | 10        | 50  | 5         | 25| 10        | 50| 17        | 85 |
| 6–10 days | 0     | 0   | 1         | 5   | 1         | 5 | 10        | 50| 3         | 15 |
| ≥ 11 days | 0     | 0   | 0         | 0   | 0         | 0 | 0         | 0 | 0         | 0 | 0         | 0 |
| Total   | 20    | 100 | 20        | 100 | 6         | 30| 20        | 100| 20        | 100 |

During follow-up all children had first anal dilatation at 3 weeks follow-up. Two children got frequent bowel movements and soiling (n = 2, 10%); one child had stricture (n = 1, 5%) and was managed with daily anal dilatations for four weeks as an outpatient and reviewed weekly. One child experienced perineal skin excoriation (n = 1, 5%) at three weeks. At weeks 7 and 12, two (2) children got enterocolitis (n = 2, 10%) and were treated with rectal washouts, intravenous antibiotics, and intravenous fluids. Table 4
Table 4

| Complication                     | Week 3 |       | Week 7 |       | Week 12 |       |
|----------------------------------|--------|-------|--------|-------|---------|-------|
|                                  | frequency | %   | Frequency | %     | Frequency | %   |
| Anal stricture                   | 1      | 5    | 0      | 0     | 0       | 0    |
| Perineal excoriation             | 1      | 5    | 0      | 0     | 0       | 0    |
| Soiling                          | 2      | 10   | 0      | 0     | 0       | 0    |
| Enterocolitis                    | 0      | 0    | 2      | 10    | 2       | 10   |
| Ongoing obstructive symptoms    | 0      | 0    | 0      | 0     | 0       | 0    |

Discussion

The goals of surgical management for Hirschsprung disease are to remove aganglionic bowel and reconstruct ganglionic bowel down to the anus while preserving normal sphincter function [34]. We prospectively followed 20 patients with simultaneous stoma closure and pull-through for immediate and short-term complications. Overall this study found that patients did well with simultaneous stoma closure and pull-through and few early complications were observed.

Existing literature has shown that early complications such as anastomotic leakage, anal strictures, and perineal excoriations have been observed [1][34]. In this study anastomotic leak was observed in one patient. The anastomotic leak was recognized on the fifth post-operative day, and proximal diversion was performed. Leaks have been shown to occur due to inadequate blood supply of the pulled-through bowel and can be avoided by minimizing tension on the anastomosis. The sigmoid colon is usually mobilized by dividing sigmoid vessels and retaining marginal vessels. Often it is necessary to mobilize the splenic flexure to obtain adequate length to avoid tension on anastomosis [1][3][34]. The patient that got burst abdomen had significant spillage intraoperatively while pulling bowel through. Bowel preparation could have helped in this case. This caused intra-abdominal sepsis hence suture breakdown of the fascial closure. We lavaged the abdomen at re-exploration and closed and patient recovery was uneventful.

One patient had a stricture in this study. Similar literature has shown that strictures and retraction of the pull-through may occur as a result of poor blood supply and tension [34]. To minimize this complication, the authors kept the dissection in the correct plane along the rectal wall to avoid injury to the deep pelvic nerves, vessels and other structures such as the vagina, prostate, vas deferens, and seminal vesicles [1][3] as well as maintaining the blood supply.

Late complications may include ongoing obstructive symptoms, frequent bowel movements, incontinence, soiling, and enterocolitis. During follow-up period the authors observed frequent bowel movements in two children (n = 2, 10%) at three weeks, and two children experienced enterocolitis (n = 2,
20%) at 7 and 12 weeks follow-up. Patients who developed enterocolitis were admitted and treated with rectal washouts, intravenous antibiotics, and intravenous fluids.

Longer term complications that will require future follow-up will include bowel function and enterocolitis as well as the socioeconomic impact of these conditions [1][34].

The main limitation of our study was a short follow-up period and relatively small number of patients. In spite of these limitations, we are encouraged by the success of this procedure and recommend it for our setting where stoma care has many challenges, reducing number of surgeries from three to two has significant positive impact in our patients’ lives.

We did not measure the economic impact for the patients which we believe would be substantial given the costs associated with a single surgical episode – up to 1/3 can become impoverished, many come and are turned away due to no theater space but have already spent money on transport and missed [37][38]. These benefits could be measured in future studies.

Like patients with anorectal malformations (ARM), many patients with HD live with stomas and the significant impacts on their quality of life (QoL). Therefore, community awareness through support groups and stoma care groups might help improve presentation, and as a result, the outcome. We created community led support group for ARM in the Mbarara, South-Western Ugandan which could be expanded to HD. This has substantially improved awareness with more patients presenting with colostomies for definitive repair.

**Conclusion**

In this small case series, we have demonstrated that pull through and simultaneous stoma closure can be safely performed in resource constrained settings such as ours. Further studies are needed to understand the quality of life and economic impact of changing the management of HD patients from three operations to two. Additional work is also needed to encourage early presentation of patients with HD such that the definitive pull through can be accomplished prior to school years.

**Declarations**

**Authors Contribution:**

Study conception and design: Felix Oyania, Stella Alice Nimanya, Martin Situma

Acquisition of data: Felix Oyania, Anne Wesonga, Martin Situma, Meera Kotagal

Analysis and interpretation of data: Felix Oyania, Doruk Ozgediz, Meera Kotagal

Drafting of manuscript: Felix Oyania, Martin Situma, Meera Kotagal,
Acknowledgement:

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Compliance with Ethical Standards:

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Conflict of interest statement:

all authors declare that they have no conflict of interest

Ethical approval:

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Ethical approval was obtained from Mbarara University research ethics committee # 02/05-19.

Informed consent:

Informed written consent was obtained from all parents/guardians of the featured patients included in the study.

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Figures
**Figure 1**

Abdominal phase: definitive repair with spontaneous stoma takedown

**Figure 2**

Perineal phase: End-end full rectal wall thickness anastomosis performed using interrupted vicryl 4/0 before stitches were cut.