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Redo pullthrough for Hirschsprung disease: A single surgical group's experience ☆

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ABSTRACT

Introduction: This study presents our surgical experience for redo-pullthrough (RedoPT) for Hirschsprung disease (HD). It reviews the patient's clinical outcomes and assesses stooling patterns after RedoPT.
Methods: A retrospective review of our institution's RedoPTs as well as one author's overseas cases was performed. Stooling scores were tabulated using an established survey tool and compared to primary PT matched patients.
Results: Between 1974 and 2012, 46 individuals (52% males) underwent RedoPT, representing 3 percent of all HD pullthroughs. Median age at primary PT and RedoPT was 1 year (range 1 week–18 years) and 3.5 years (range 8 weeks–41 years), respectively. Indications for RedoPT were predominately for aganglionosis/transition zone pathology (71%); followed by stricture or an obstructing Duhamel pouch (19%), tight cuff (8%) and a twisted PT (4%). None were performed for an isolated clinical diagnosis of repeated bouts of enterocolitis. RedoPT surgical approach depended upon the initial pullthrough technique and any previous complications. Stooling scores were significantly ($P < 0.05$) worse in the RedoPT patients compared to the historically-matched group of children undergoing a primary PT for HD (5.5 ± 1.2 vs. 12.2 ± 1.4, primary PT versus RedoPT, respectively). When breaking down this total score into individual parameters, stooling pattern scores (1.0 ± 0.2 vs. 4.1 ± 0.4, $P = 0.001$) and enterocolitis scores (2.0 ± 0.4 vs. 4.2 ± 0.4, $P = 0.001$) were statistically worse in the RedoPT group. Patients in both groups had similar overall continence rates.
Conclusion: Appropriately selected children undergoing a RedoPT can achieve good results, with comparable continence rates to those undergoing a primary PT.

Successful operative management for Hirschsprung disease (HD) has been performed for over 65 years [1]. Repair usually results in satisfactory results. However, a small but significant proportion of children will experience long-term postoperative complications [2]. Medical management and/or minor surgical procedures correct the complications in most situations, but some patients will require a redo-pullthrough (RedoPT) for the definitive treatment of severe or persistent symptoms [3,4]. Because of the uncommon frequency of RedoPT, relatively few series have evaluated patient's outcomes following such RedoPTs in HD. The objectives of this paper are to understand the indications for RedoPT surgery, analyze the surgical procedure used for RedoPT and evaluate stooling outcomes of patients after RedoPT through an extensive review of our own experience. The present report represents one of the largest series of RedoPT in HD. In addition to this retrospective review, we also examined a subset of patients in which we were able to conduct a survey to assess current stooling patterns.

1. Methods

The study was approved by the University of Michigan IRB (HUM00001989). The cohort included 32 RedoPT patients operated on at the University of Michigan, CS Mott Children's Hospital and 14 patients operated on at other institutions by the same surgeon (AGC). Demographic data, pathology, operative and clinical notes were reviewed. Additionally, timing of and type of primary PT and RedoPT, length of aganglionic segment, and early complications (anastomotic leak, twisted PT, abscess, wound infection, perforation, Hirschsprung associated enterocolitis (HAEC), and incontinence) and late complications (constipation, stricture, fistula, fecal incontinence, osteomyelitis, and HAEC) of both operations were recorded. Patient age, indication, involved segment length, timing between and type of RedoPT were recorded from cases performed by AGC at an outside institution. However, neither review of pathology nor stooling outcomes are reported from this smaller group of cases performed outside our university. This latter group was primarily included to

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look at length of time in between operations, type of RedoPT used as well as indications for RedoPT.

Twenty-one of the 32 patients remained current in our system allowing long-term follow up. A Stooling Survey from El-Sawaf et al, was completed through a telephone interview [5]. Stooling scores comprised a composite evaluation of continence, stooling pattern (e.g., excessively loose or explosive stooling) and evidence of enterocolitis. For unreachable families, the most recent clinic notes were reviewed to complete the Stooling Survey questions. Pathology slides from primary PT and RedoPT were also reviewed, when available, by a single pediatric expert pathologist (RR) to gain a better understanding of the pathogenesis of disease. Nerves were defined as hypertrophic if greater than 40 microns in diameter.

1.1. Statistical analysis

The results from our institution's RedoPT group were compared to a dataset from a group of published dataset primary-PT patients after matching for sex, mental developmental delay and length of aganglionosis (Table 1) [6]. These primary-PT controls were compared to the RedoPT group using a two-tailed t-test and Chi squared where appropriate. Results were also stratified by the length of time from the primary PT to the RedoPT. Immediate RedoPT was defined as less than 6 months from primary PT. Early and late RedoPTs were defined as between 6 months and 3 years and after 3 years, respectively from the time of primary PT. Results are expressed at the mean ± SD. Statistical significance was set at $P < 0.05$.

2. Results

2.1. Demographics

Between 1974 and 2012, 46 individuals (52% male) underwent RedoPT. Median age at primary PT and RedoPT was 1 year (mean ± SD 2.2 ± 4.2, range 1 week–18 years) and 3.5 years (mean ± SD 6.9 ± 9.2, range 8 weeks–41 years), respectively. Median time between primary PT and RedoPT was 11 months, however, the range was quite wide (mean ± SD, 39.3 ± 61.3) at our facility; and somewhat longer at outside facilities: 30.5 months (mean ± SD, 59.2 ± 105.6). Table 1 summarizes the patients who underwent RedoPT and compares them to a matched group of primary PT. The child's primary PT was performed at our institution in 28% of cases; with 59% of these having rectosigmoid disease, 13% left-sided and 9% mid-colon or right-sided (including 1 patient with total colonic) disease. For 19% of patients, no recorded length of involved aganglionosis was documented. Open endorectal PT (ERPT) was the most frequent primary PT (41%), followed by transanal ERPT (22%), Swenson (17%), Duhamel (11%), and unknown operation (9%).

2.2. Outcomes after the primary pull though

Early complications after the patient's primary PT occurred in 44% of all cases that eventually underwent a RedoPT. This early complication rate was significantly higher than the historical cohort of patients analyzed for stooling outcomes who did not require a RedoPT (16% $P = 0.01$) [5]. These early complications included anastomotic leak (19%), obstruction (9%), twisted PT (4%) and enterocolitis (6%). Late complications following the primary PT in those children eventually requiring a RedoPT included obstructive symptoms (87%) of which functional constipation made up the majority (54%). Obstruction was caused by stricture in 22% and 9% had a specific technical cause for constipation, consisting of an obstructing Duhamel pouch ($n = 3$) or twisted PT ($n = 1$). Other late complications included fistula (4%, one rectocutaneous, one colocutaneous) and one child (2%) each with fecal incontinence, osteomyelitis, enterocolitis and persistent distention.

Table 1
Summary of RedoPT group and primary-PT controls.

	RedoPT	Primary PT	P value
N	32	89	
Male	52%	82%	0.17
MDD	6.5%	7%	0.75
Segment length			
Rectosigmoid	57%	80%	0.3
Left	11%	13%	0.91
Mid or right	9%	7%	0.86
Unknown	23%		

Primary-PT data from Kim et al [5]. MDD = Mental Developmental Delay.

2.3. Work-up prior to redo pull through

Nineteen of 46 (41%) patients underwent an ostomy creation prior to RedoPT. Indications for the ostomy included dilated colon ($n = 10$), stricture ($n = 3$), anastomotic leak ($n = 3$) and miscellaneous reasons ($n = 3$); one of whom had a retained segment from an incorrect pathology interpretation at the time of primary PT. Twenty-eight percent of patients at our institution underwent intervention in an attempt to alleviate obstructive symptoms prior to RedoPT as per our previously published algorithm [3]. This included Botox® (Allergan, Inc. Irvine, CA) injection into the anal sphincters and anal dilations (2 patients) or a posterior myotomy or myectomy (POMM; 7 patients). The indications for POMM or dilations were stricture or tight cuff. However, 5 of 7 were subsequently found to have retained aganglionosis either by biopsy or upon final review of pathology after RedoPT. An additional child was found to have a twisted segment during RedoPT.

2.4. Redo pull through: indications and operative approach

Indications for RedoPT were not documented in 8 patients. The remaining indications for RedoPT were predominately for a retained aganglionosis/transition zone pathology (RA/TZP; 71% overall and 56% from children who had a RedoPT at our institution). Other indications included stricture/obstructing Duhamel pouch (19%), excessively tight cuff (8%), twisted PT (4%), but none for recurrent enterocolitis (0%). The time between primary PT and RedoPT was suggestive of the indication for operation. The immediate reoperative group (those performed within 6 months after the primary PT) all had gross anatomical indications (twisted segment, obstructing Duhamel pouch or stricture) for their RedoPT. Whereas, the early (<3 years) and late (>3 years) RedoPT groups showed 40% and 70% of patients having RA/TZP, respectively.

Operative approach varied widely (Fig. 1), and depended greatly on the type of primary PT as well as the type of complication leading to the RedoPT. An open ERPT was the most commonly performed RedoPT procedure (38%). This was followed by a Swenson (25%), Duhamel (13%), and transanal ERPT (7%). For an additional 13% of patients who suffered from a frozen pelvis secondary to several previous attempts at RedoPT not performed by our group further distal dissection could not proceed in the distal rectum. To address these latter patients, an end-to-end anastomosis (EEA) was performed using a circular EEA stapling device.

2.5. Pathology

Pathology specimen from the primary PT was available from the 9 children who had both their primary and RedoPT at our hospital. All slides from the RedoPTs viewed confirmed the documented indication for RedoPT. In one case a segment of aganglionic bowel was incorrectly used in the PT as a result of a misread frozen section. During the final pathologic reading, this patient's proximal surgical