The Reconstructive Pelvic Medicine Clinic at Seattle Children's Hospital and Quality Improvement Efforts for Patients with Hirschsprung Disease

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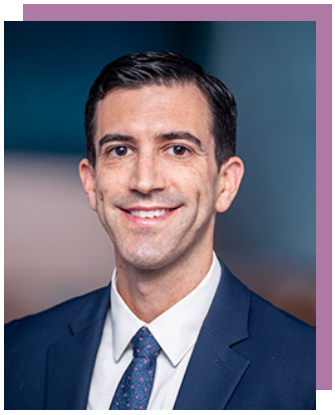

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Drs. Caitlin Smith, Assistant Professor, and Samuel Rice-Townsend, Assistant Professor, Division of Pediatric General Surgery, are pediatric colorectal surgeons at Seattle Children's Hospital (SCH), and are actively involved in the multidisciplinary Reconstructive Pelvic Medicine (RPM) Program, which Dr. Smith also directs. The RPM Program represents one of the few multidisciplinary pediatric colorectal and reconstructive pelvic surgery programs in the United States, offering a comprehensive approach to patients with complex surgical problems such as anorectal malformations, cloaca, and Hirschsprung disease (HD). The clinical and translational research they perform reflects the RPM Program's mission, aiming to improve the quality of life for patients affected by these conditions.

These conditions are rare and their management specialized, and for these reasons, their research often leverages collaboration with the Pediatric Colorectal and Pelvic Learning Consortium (PCPLC), an international consortium of surgeons and other specialists to facilitate multi-institutional studies. SCH was one of the founding centers for this collaborative.

In an example of this work, they recently examined practice patterns of botulinum toxin (Botox-BT) use in patients with Hirschsprung disease. Infants and children with Hirschsprung disease may struggle with obstructive symptoms after pullthrough procedure in up to 30% of cases. While injection of BT is widely accepted as a therapeutic option, its described use and patient selection varies. Using the PCPLC multi-institutional registry, Drs. Smith and Rice-Townsend sought to characterize patterns of BT use across these referral centers as a step toward the development of consensus guidelines.

A total of 494 pediatric patients with Hirschsprung disease were included in the study between 2017 and 2021. About a quarter of patients received BT during the time period. Most patients underwent a pullthrough procedure alone while about 10% underwent a primary and redo pullthrough procedure.

Patients who had a redo pullthrough were more likely to receive BT and also received it more frequently. This was expected. Interestingly, however, patients who underwent primary pullthrough at a younger age were less likely to receive BT. While most of the patients who received BT were male and white, patients with Hispanic ethnicity were statistically less likely to receive BT when compared to non-Hispanic patients.

No significant difference was seen otherwise for the patients who received BT compared to those that did not in terms of sex, race, ethnic status, insurance status, or a diagnosis of Trisomy 21.

Looking at variability of BT use across centers, a wide range was seen. Percentage of HD patients receiving any BT varied significantly among hospital sites ranging from 8%-50% of HD patients. Frequency of BT also varied significantly by hospital site.

In summary, Drs. Smith and Rice-Townsend found that at referral centers, patients with Hirschsprung disease who required revisional surgery received more BT, while Hispanic patients received less. Additionally, there was a significant association between timing of pullthrough procedure and use of BT. These findings warrant further investigation. The wide variation in practice patterns across institutions also justifies ongoing efforts within the consortium to clarify best practice protocols for BT administration in patients with Hirschsprung disease.

Age and Timing of Botox Use in Hirschsprung Patients

	Patient ever received a Botox injection			
	Yes (N = 118)	No (N = 376)	Overall (N = 494)	P-value
Age in years at data lock (May 3rd, 2021)				0.406[1]
< 5	48 (40.7%)	177 (47.1%)	225 (45.5%)	
≥ 5 and < 12	55 (46.6%)	150 (39.9%)	205 (41.5%)	
≥ 12	15 (12.7%)	49 (13.0%)	64 (13.0%)	
Endorectal pullthrough history				<.001[1]
Only a primary pullthrough on record	83 (70.3%)	282 (75.0%)	365 (73.9%)	
A primary pullthrough and a revision on record	26 (22.0%)	23 (6.1%)	49 (9.9%)	
Age at primary endorectal pullthrough (months)				0.021[2]
N	109	305	414	
Median (Q1, Q3)	3.8 (1.5, 11.4)	2.9 (0.5, 7.8)	3.4 (0.7, 8.5)	
Age at primary endorectal pullthrough				0.019[1]
< 1 month	22 (18.6%)	98 (26.1%)	120 (24.3%)	
1 month to 3 months	19 (16.1%)	58 (15.4%)	77 (15.6%)	
> 3 months to 6 months	28 (23.7%)	48 (12.8%)	76 (15.4%)	
> 6 months to 1 year	13 (11.0%)	49 (13.0%)	62 (12.6%)	
> 1 year	27 (22.9%)	52 (13.8%)	79 (16.0%)	