



Evidence-Based Resource

What Is The Optimal Strategy In The Management Of Obstructive Symptoms And Enterocolitis (HAEC) After Pull-Through For Hirschsprung's Disease?

Evidence Level II

The management of obstructive symptoms and re-occurring episodes of Hirschsprung associated enterocolitis (HAEC) after surgery poses a significant obstacle for children with Hirschsprung's disease (HD). Changes in the intestinal barrier and disrupted gastrointestinal mucosal immunity have been suggested to result in the development of HAEC after surgery; as such, numerous experimental treatments have been suggested to control obstructive symptoms and prevent the occurrence and/or re-occurrence of HAEC. However, there remains a paucity of evidence surrounding which postoperative strategy is optimal in decreasing the incidence of obstructive symptoms (e.g., consistent abdominal distension or severe constipation) and HAEC (i.e., lethargy, fever, electrolyte disruption, diarrhea, heightened inflammatory markers, and abdominal bloating).

Soh and colleagues performed a systematic review and meta-analysis to summarize the best presently available evidence regarding the recommended treatment strategies (e.g., routine postoperative dilatations, postoperative rectal irrigations, posterior myotomy/myectomy (POMM), botulinum toxin (BT) injection, topical nitric oxide (NO), probiotic prophylaxis, and anti-inflammatory drugs). A total of 29 relevant studies published between 1992 to 2017 were identified of which (1) 2 case-control and 5 case-series in a total of 504 patients reported on the use of routine postoperative anal dilatations; (2) 2 case-control studies in a total of 172 patients focused on the use of routine postoperative rectal irrigations; (3) 4 case-series in a total of 50 patients reported on the use of POMM to manage postoperative obstructive symptoms and/or re-occurring HAEC; (4) 9 case-series reported on the use of BT injection after pull-through; (5) 3 case-series in a total of 13 patients reported on the use of topical isosorbide dinitrate paste or glyceryl trinitrate ointment; (6) 3 randomized controlled trials focused on the use of prophylactic probiotics (i.e., lactobacillus, Bifidobacterium, Streptococcus, and Enterococcus) postoperatively; and (7) 1 case-series in a total of 8 patients reported on the use of an anti-inflammatory drug to prevent HAEC. The outcome measures included: patient demographics, frequency of improvement or entire relief of symptoms, the need for repeated or more surgery, postoperative interventions, and the type of pull-through.

Proportion meta-analysis and random-effect was conducted to calculate the proportion attributable to noncomparative studies. A random-effect model was used to generate risk ratio (RR) for categorical variables and mean difference (MD) for continuous variables including 95% confidence intervals (CI). The Egger's regression test assessed publication bias when 3 or more studies were included. $P < 0.05$ was considered significant. An $I^2 > 50\%$ indicated gross statistical heterogeneity. The results included:

- No difference in the incidence of postoperative stricture with or without the use of routine dilations [RR 0.3 (0.02, 5.7); $p = 0.4$]; $I^2 = 72.2\%$; $p = 0.05$
- Significant reduction in the incidence of postoperative HAEC with rectal irrigations facilitated for a minimum of 6 months [RR 0.2 (0.1, 0.5); $p = 0.0001$]; $I^2 = 0\%$; $p = 0.9$
- 79% (60.6, 93.5); ($I^2 = 39.3\%$ $p = 0.17$) resolution of obstructive symptoms and 80% (64.1, 92.1); ($I^2 = 0$; $p = 0.5$) in the resolution of HAEC using POMM
- An overall short term-response of 77.3% (68.2, 85.2); ($I^2 = 38.2\%$; $p = 0.13$) and overall long-term response of 43.0% (26.9, 59.9); ($I^2 = 78.4\%$; $p = 0.0001$) in the use of BT injection following pull-through
- Very limited present evidence to support the use of NO donors to treat obstructive symptoms after pull-through for HD
- No differences in the incidence of postoperative HAEC with or without the use of prophylactic probiotics [RR 0.6 (0.2, 1.7); $p = 0.3$]; $I^2 = 54.7\%$; $p = 0.1$
- No further evidence on the use of anti-inflammatory drugs for prevention of HAEC

Conclusions:

Rectal irrigations seem to be effective at preventing HAEC; however, given that the studies pertaining to this strategy refer to different surgical procedures and one study implemented an additional intervention (i.e., antibiotic prophylaxis), the results from the meta-analysis are inconclusive. There was a possibility of publication bias and statistical heterogeneity for treating obstructive symptoms with routine postoperative dilatations; thus, the authors have a challenging time recommending it to all patients after pull-through. In the short-term, BT injection appears to be a valid alternative to POMM. There is a need for further investigation to support the use of NO donors after pull-through, prophylactic probiotics, and anti-inflammatory drugs to prevent HAEC due to a lack of convincing evidence. Findings from this review and meta-analysis indicate that there is a lack of high quality evidence characterized by significant heterogeneity and potential publication biases; hence, there is limited support for any of these strategies.

A limitation of this study is that the differences in outcomes could not be compared or contrasted according to the surgical procedures performed since the majority of the studies included did not stratify their results by procedure type.

Systematic Reviews

[Soh HJ, Nataraja RM, Pacilli M. Prevention and management of recurrent postoperative Hirschsprung's disease obstructive symptoms and enterocolitis: Systematic review and meta-analysis. J Pediatr Surg. 2018;53\(12\):2423-9.](#)

