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Use of an enterocolitis triage and treatment protocol in children with Hirschsprung disease reduces hospital admissions☆

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ABSTRACT

Background: While a consensus for the definition of Hirschsprung associated enterocolitis (HAEC) is lacking, the mainstay of treatment includes rectal irrigations with or without antibiotics. This treatment is often effective when initiated as an outpatient. Our institution implemented a triage algorithm in an effort to standardize care thus providing more timely treatment and preventing unnecessary hospital admissions. We sought to review our short-term experience.
Methods: A retrospective review was performed of all Hirschsprung (HD) patients <6 years old over two distinct time periods from May 2016–2017 (pre-protocol, group A) and June 2017–2018 (post-protocol, group B). Patients with a colostomy were excluded. Primary end point was hospital admission. Presenting symptoms were categorized as moderate or severe, with patient triage based on number and quality of symptoms.
Results: Eighty-seven total patients were included. Rectosigmoid transition zone was most common (75%) and 20% of patients had trisomy 21. HAEC occurred in 22% of patients in the preprotocol group (group A, n = 78, 27 episodes) and 20% of patients in the post-protocol group (group B, n = 87, 32 episodes). In group A, 78% of episodes required an unplanned visit and 74% resulted in admission. In group B, 81% of episodes required an unplanned visit and 50% resulted in admission (33% reduction in hospital admission, p = 0.06). Irrigations only, without antibiotics, were used in 30% of episodes in group A versus 41% in group B. Of patients who initially contacted the office by phone (group A = 7 episodes, group B = 6 episodes), outpatient management was successful in 43% versus 100% respectively (p = 0.07). No patient experienced increased morbidity in group B.
Discussion: Implementation of a HAEC treatment algorithm shows promise in improving the management and resource utilization of this complex patient population. It is anticipated that continued education of caregivers and the treatment team will result in a greater effect. A multi-institutional implementation of this algorithm is needed to characterize risk factors associated with failure of outpatient management.
Level of Evidence: III, Treatment Study.

Hirschsprung-associated enterocolitis (HAEC) is associated with high morbidity and continues to be the most common cause of death in children with Hirschsprung disease (HD) [1–3]. Discrepancies exist regarding the exact clinical presentation of HAEC, and recent literature seeks to standardize the definition in hopes of creating more consistent outcome measures for future studies [1,2]. There is a general consensus as to the importance of the following symptoms in diagnosing HAEC: fever, diarrhea, explosive stools, abdominal distension, vomiting, and evidence of obstruction [1,2,4–6]. Because of the non-specific clinical presentation and the potential for significant morbidity, many Hirschsprung's disease (HD) patients are over-treated, resulting in increased rates of hospitalization and unnecessary use of antibiotics [1,2]. While most providers with experience in the management of HD would agree that treatment of HAEC involves rectal irrigations, general guidelines regarding the use and duration of antibiotics, frequency and duration of rectal irrigations, and outpatient versus inpatient treatment are lacking.

We implemented an algorithmic HAEC triage protocol at our institution in an effort to standardize care among providers, identify patients appropriate for outpatient management, and prevent unnecessary hospital admissions. Here we review our short-term institutional experience with this triage protocol.

1. Methods

Following IRB approval, a retrospective analysis was performed examining all HD patients treated at our tertiary care children's hospital from May 2016 to June 2018. Our protocol was initiated in June 2017. The entire cohort was evaluated across two distinct time periods, the pre-protocol period (May 2016–2017, group A) and post-protocol

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period (June 2017–2018, group B). This study was conducted as a retrospective pre-post intervention, cohort study with new patients included as they entered our practice. Patient demographics and associated clinical data were obtained from a review of our electronic medical record system.

Inclusion criteria for our study consisted of patients with HD less than 6 years of age prior to May 2016. Of this population (n = 87), patients were further classified according to whether or not they experienced an episode of enterocolitis. As demonstrated by Kwendakwema et al., a majority of HAEC episodes at our institution have been found to occur within the first 2 years following pull-through surgery [7]. Given this, patients >6 years of age were excluded from our study, as it was felt this age cut-off would adequately capture the patient population of interest. Furthermore, patients with a colostomy at the time of HAEC were also excluded. Patients with colostomies at time of HAEC episode represented a cohort of patients with complex comorbidities, and thus it was felt that exclusion of this subgroup would provide a more homogenous cohort for analysis. The primary end point for this study was hospital admission. Unplanned hospital visit was defined as initial presentation to clinic or the emergency department without preceding telephone call.

Patients were considered to have HAEC if rectal irrigations, with or without antibiotics, were initiated based on presenting symptoms. While the definition of HAEC at our institution remained consistent through both the pre- and post-protocol time periods, HAEC treatment in the early era was initiated solely based on provider discretion. Following protocol implementation, HAEC was explicitly defined as having at least 2 moderate criteria as determined by our institutional algorithm (Fig. 1). Treatment in this cohort was standardized by protocol, as described below.

Following development, the protocol was introduced at both faculty and staff meetings, with an electronic copy made available to all providers on a shared access drive for continued reference as needed. A dedicated colorectal nurse practitioner monitored all Hirschsprung-related admissions and protocol compliance with subsequent feedback and education provided.

A checklist of symptoms was created using those from the HAEC score Delphi analysis and the symptoms categorized as either moderate or severe [2]. Using published recommendations from the American Pediatric Surgical Association Hirschsprung Disease Interest Group a triage and treatment protocol was created [1]. Triage was based on quality and quantity of the symptoms elicited (Fig. 1). At our specialty Colorectal Center, all caregivers receive extensive counseling of signs and symptoms of HAEC, are taught how to perform rectal irrigations and are provided with necessary equipment to perform rectal irrigations at home. For newborns undergoing a delayed one-stage pull-through procedure, this teaching is performed and irrigations initiated following diagnosis by suction rectal biopsy prior to hospital discharge. Patients initially managed with a leveling colostomy and a staged endorectal pull-through receive rectal irrigation teaching at their clinic visit following the pullthrough. Likewise, our team reviews the signs and symptoms of HAEC at each follow-up visit, and subsequently verifies that the patient and their caregiver has a rectal irrigation kit at home.

When HAEC symptoms occurred and clinic contact was initiated by the caregiver via telephone, the triage protocol was used to determine if the patient was an appropriate candidate for outpatient management, or if an in-person evaluation with possible hospital admission was needed. This protocol was also used to guide outpatient versus inpatient treatment when initial contact occurred in the clinic or emergency department.

A subset of patients demonstrating severe symptoms was advised for immediate admission to the hospital. When an in-person visit was required, a stool sample was collected and antibiotics were initiated. Antibiotics were often discontinued after 48 h of negative stool cultures, but were occasionally continued for a complete 7-day course based on treating surgeon discretion. Likewise, if stool cultures were positive for pathogenic bacteria, antibiotics were continued for 7 days. Patients evaluated in-person also had a rectal irrigation performed by the surgical provider. Those who saw clinical improvement following irrigation were discharged home on rectal irrigations per our algorithm. Patients with no improvement following rectal irrigation were admitted to the hospital for intravenous hydration, antibiotics and rectal irrigations. Patients treated in the outpatient setting who saw no symptomatic improvement following rectal irrigation alone were advised to be seen in-person.

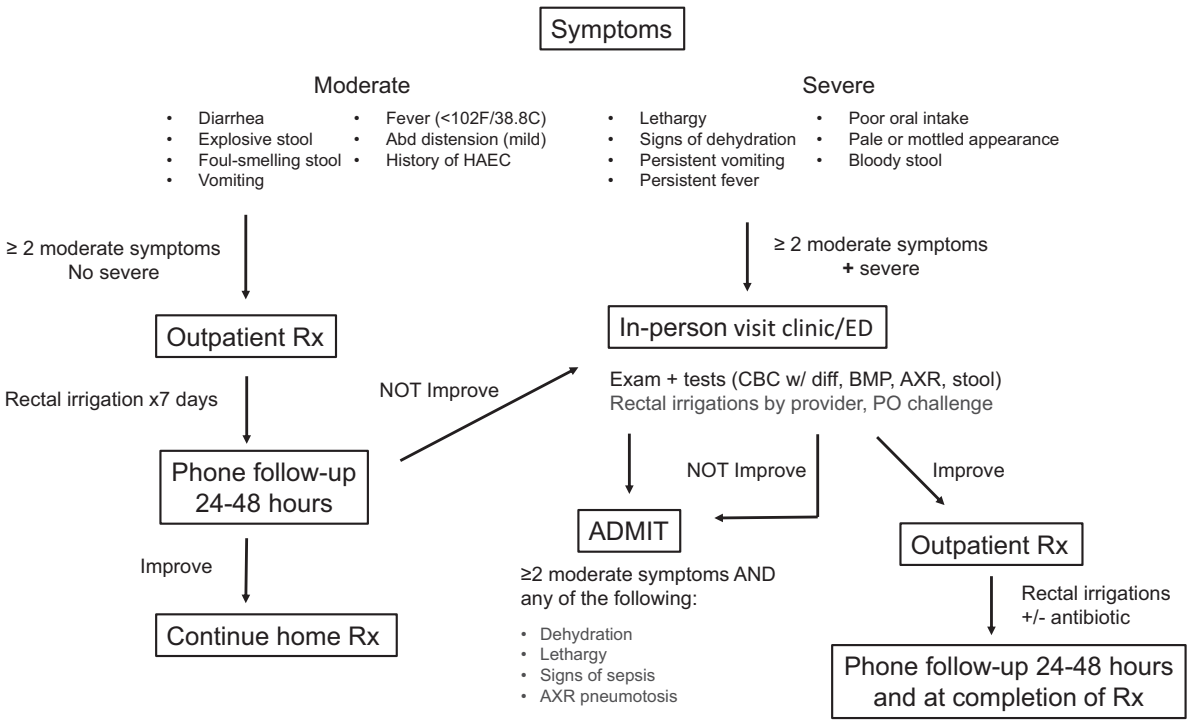

Fig. 1. HAEC triage algorithm.

Table 1
Patient demographics.

Variable	Total patients (n = 87)	Patients with HAEC (n = 27)
Gender (n,%)		
Male	63 (72.4%)	19 (70.4%)
Female	24 (27.6%)	8 (29.6%)
Ethnicity		
White	65 (74.7%)	22 (81.5%)
Hispanic/Latino	13 (14.9%)	2 (7.4%)
Black/African American	4 (4.6%)	2 (7.4%)
Asian/Pacific Islander	3 (3.4%)	1 (3.7%)
Not listed	2 (2.3%)	0 (0%)
Trisomy 21	17 (19.5%)	3 (11.1%)
Surgery type		
Primary	42 (48.3%)	15 (55.6%)
Staged	43 (49.4%)	12 (44.4%)
No surgery	2 (2.3%)	0 (0%)
Transition zone*		
Short segment	65 (74.7%)	19 (70.4%)
Long segment	16 (18.4%)	4 (14.8%)
Total colon	6 (6.9%)	4 (14.8%)

* Short segment = rectosigmoid transition zone, long segment = transition zone proximal to rectosigmoid with ganglion cells present in cecum, total colon = aganglionosis of entire colon with transition zone in distal ileum.

Values were analyzed using either Pearson's chi-squared tests for categorical variables, and independent sample t-tests for continuous variables. Clinical significance was defined as a p-value ≤0.05.

2. Results

A total of 87 patients were identified according to inclusion criteria. A majority of these patients were male (72%) with Caucasian ethnicity (75%). Median age at time of HAEC episode was 1.9 years. Sixty-five patients had a rectosigmoid transition zone (75%), and 20% of our patient population had a diagnosis of Trisomy 21 (Table 1). Group A and B were comprised of 78 and 87 patients, respectively. Twenty-two percent of patients in Group A (27 episodes) and 20% of patients in Group B (32 episodes) were treated for HAEC (Table 2). Six patients in Group A (7.7%) and 10 patients in group B (11.5%) experienced ≥2 episodes of HAEC. Thirty-eight HAEC episodes (64%) were found to occur within the first 2 years following pull-through surgery. Forty-eight percent of patients underwent a primary endorectal pullthrough (majority were laparoscopic-assisted Soave-type endorectal pull-through), and 49% underwent a staged operation (neonatal leveling colostomy followed by a laparoscopic-assisted Soave-type endorectal pull-through). HAEC did not appear to correlate with staged versus non-staged management as the incidence was 23.7% and 26.2% in post-pullthrough staged and non-staged patients, respectively (p = 0.80).

A total of 7 patients had episodes of enterocolitis both before and after implementation of the triage protocol, and thus were included in the analysis of the corresponding time period. These 7 patients comprised a total of 13 and 9 HAEC episodes in the pre- and post-protocol periods, respectively. Of the 13 HAEC episodes prior to protocol implementation, 11 required hospital admission. Likewise, 4 of the 9 HAEC episodes in the post-protocol period required admission.

In group A 74% of episodes and in group B 50% of episodes resulted in admission (Table 3). This represents a 33% reduction in hospital admission following implementation of an HAEC triage protocol (p = 0.06). Mean length of stay for patients requiring hospitalization was 3 days and 6.3 days for Group A and Group B, respectively (p = 0.22). A majority of patients in both groups initially presented to the emergency department (ED) for care (67% vs 72%, p = 0.67). Of patients who initially contacted the clinic by telephone (group A = 7 episodes, Group B = 6 episodes), 43% of Group A and 100% of Group B were successfully managed in the outpatient setting with no further escalation of care (p = 0.07). Treatment with rectal irrigation only was used in 30% of HAEC episodes in Group A, versus 41% of episodes in Group B (p = 0.38). Total antibiotic use was noted to have a 12% decrease following protocol implementation (67% vs 59%, p = 0.56). There was no increased morbidity among patients in Group B.

3. Discussion

The reported incidence of HAEC is highly variable due to lack of a uniform definition and the non-specific nature of presenting symptoms. The overall occurrence of HAEC in our study was 31% which is consistent with previous reports which range from 2 to 50% [2,3,6,8]. Pastor et al. developed a scoring system in efforts to standardize the definition of HAEC and establish a consistent, reproducible measure for future investigations [2]. However, this scoring system was not intended to guide clinicians in diagnosis and management of HAEC. Guidelines for the diagnosis and management of HAEC have been published by the APSA Hirschsprung Disease interest group; however, treatment of HAEC is inconsistent among providers and institutions.

HAEC may rapidly progress to sepsis and even death, which leads to conservative treatment of admission to the hospital for rectal irrigations, intravenous antibiotics and intravenous fluid resuscitation. Assuming that the patient is appropriately hydrated, the mainstay of treatment remains rectal irrigation with or without antibiotics, which can safely be initiated in the outpatient setting for many patients [1]. We initiated a HAEC triage protocol based on the guidelines mentioned above in an effort to standardize diagnosis and treatment among providers at our institution. The goal was to diagnose HAEC early and initiate outpatient treatment thereby avoiding hospital admission. As demonstrated in our algorithm (Fig. 1), an essential component to safe and successful outpatient treatment is caregiver compliance and short-term follow up.

Table 2
HAEC episode presentation and treatment characteristics

Variable	Group A (n = 78)	Group B (n = 87)	Total	P
Patients (n,%)				
With HAEC episode(s)	17/78 (21.8%)	17/87 (19.5%)	27/87 (31.0%)	0.72
With ≥2 HAEC episodes	6/78 (7.7%)	10/87 (11.5%)	17/87 (19.5%)	
Total HAEC episodes	27	32	59	
Initial presentation/episode (n,%)				
Telephone	7 (25.9%)	6 (18.8%)	13 (22.0%)	0.51
Clinic	2 (7.4%)	3 (9.4%)	5 (8.5%)	0.79
ED	18 (66.7%)	23 (71.9%)	41 (69.5%)	0.67
Treatment initiated/episode (n,%)				
Irrigations only	8 (29.6%)	13 (40.6%)	21 (35.6%)	0.38
Antibiotics only	2 (7.4%)	1 (3.1%)	3 (5.1%)	0.46
Irrigation and antibiotics	16 (59.3%)	18 (56.3%)	34 (57.6%)	0.82
None	1 (3.7%)	0 (0%)	1 (1.7%)	

Table 3
Clinical outcomes.

Variable	Group A	Group B	Total	P
Episodes requiring hospital visit* (n,%)	21/27 (77.8%)	26/32 (81.3%)	47/59 (79.7%)	0.74
Episodes requiring admission (n,%)	20/27 (74.1%)	16/32 (50%)	36/59 (61%)	0.06
Days in hospital for patients requiring admission (mean)	3	6.8	4.7	0.22
Unplanned visit(s) which met protocol criteria for outpatient management (n,%)	7/20 (35%)	10/26 (38.5%)	17/46 (37%)	
Episodes successfully treated with outpatient management (n,%)	3/7 (42.9%)	6/6 (100%)	9/13 (69.2%)	0.07

* Hospital visit defined as unplanned presentation to either clinic or ED.

An unexpected finding following initiation of our protocol was that unplanned clinic/ED visits were unchanged. In part, it is suspected this is a result of early provider discomfort with managing HAEC symptoms by telephone conversation in combination with protocol novelty. Nevertheless, a 33% reduction in hospital admission was achieved. Though not statistically significant, we feel this reduction is clinically important. A total sample size of 116 patients (58 pre- and 58 post-protocol) would be necessary to see a statistically significant effect between groups (assuming a traditional alpha of 0.05, beta of 0.2, and power of 0.8).

We have a large team of advanced practice clinicians, rotating surgical residents and a number of surgeons that receive phone calls from caregivers after hours. We anticipate that unplanned visits will continue to decline as patient caregivers and the surgical team become more familiar with our protocol. This point is demonstrated by the fact that 7 HAEC episodes in Group A (35% of unplanned visits) and 10 in Group B (39% of unplanned visits) met criteria for outpatient management based on symptoms, but instead presented to the clinic/ED for in-person evaluation. Six of the 7 patients in the pre-protocol cohort whose symptoms met criteria for outpatient management were subsequently admitted to the hospital for treatment. Conversely, only 2 of the 10 patients in the post-protocol cohort who met outpatient criteria were admitted for hospitalization.

With regard to HAEC management, we found that numerically more patients in Group B were successfully treated as an outpatient, with no further escalation of care required (43% vs 100%, $p = 0.07$). Likewise, in this small subset of patients we noted a 12% overall decrease in antibiotic use for treatment of HAEC (67% vs 59%, $p = 0.56$). We did not appreciate an increased need for ongoing care in patients who underwent rectal irrigation without supplemental antibiotic use. These findings suggest that our triage algorithm may appropriately identify HAEC and appropriately target patients who are candidates for safe outpatient management. Interestingly, it was noted that patients requiring hospitalization in Group B had a longer average length of stay when compared to Group A (3 days vs 6.3 days, $p = 0.22$). Although not statistically significant, we speculate that this clinically significant difference is related to appropriate identification of patients with more severe disease for inpatient treatment.

Education is paramount when attempting to employ a patient treatment algorithm such as the one proposed. We emphasize early recognition of symptoms to caregivers and conduct extensive teaching on how to perform rectal irrigations. This teaching includes an instructional video and observance of the caregiver performing a rectal irrigation by our staff. Also, our post-operative course includes routine rectal irrigations starting at the first post-operative visit (twice a day for 1 month then daily for 1 month). This post-operative care is based on findings from a prior case series from our institution by Marty et al. [9] and has become standard care for all HD patients post-pullthrough. Furthermore, early caregiver contact with our clinic is repeatedly encouraged if the patient experiences any symptoms which could be consistent with HAEC.

Our study is subject to several limitations. First, our population is comprised of patients treated at a large children's hospital and specialty clinic and therefore may not be generalizable to all settings. Second, patients at our institution undergo a two-month course of routine home rectal irrigations following pull-through surgery. Thus, families are familiar with how to perform rectal irrigations and have supplies to do so at home. Additionally, given the retrospective pre-post intervention, cohort design of our study there exists the possibility that results may be due to unaccounted for changes other than the implemented protocol. Such potential confounders include but are not limited to variation in treatment among providers prior to establishment of uniform protocol, caregiver compliance, and acquisition of new hospital practitioners. Results could also be explained by the fact that simply having a protocol allows for providers to be more attuned to the diagnosis of HAEC, possibly prompting earlier treatment. Finally, given the retrospective nature of our study, data collection was limited to that information previously documented in the electronic medical record (EMR) system.

4. Conclusion

Use of a HAEC triage and treatment algorithm shows promise as a method to decrease health care resource utilization without increasing morbidity. It is anticipated that with continued education of patient caregivers and the surgical team our program will realize an even greater effect. A multi-institutional implementation of this algorithm is needed to further characterize risk factors associated with failure of outpatient management and to determine if the algorithm is appropriate for broad use.

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