

## Application of Delphi Score in Patients with Hirschsprung Associated Enterocolitis (HAEC): Two Sudanese Pediatric Surgery Centers' Experience and Review of the Literature

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### Abstract

**Background:** Hirschsprung-associated enterocolitis (HAEC) represents one of the fatal complications related to Hirschsprung's disease. Early identification and proper management are crucial to prevent a great deal of morbidity, financial burden, and mortality related to this clinical condition.

**Patients and Methods:** The study is a Retro-prospective, descriptive-analytic, hospital-based study. The study was conducted in Soba university hospital and Ribat university hospital during the period from March/2015 to December/2016. Data were collected by a specially constructed questionnaire analyzed using the software Statistical Package for the Social Sciences (SPSS) Version 23.0.

**Results:** Fifty-two patients met the inclusion criteria, and 41 (78.8%) were males. Male to Female ratio was 3.7:1. The study population's age ranged from 1 week to 10 years, and enterocolitis was reported in 10 (19.2%) patients. On admission, the mean value of the Delphi Score was  $5.44 \pm 4.42$ . The patients attained more than 10 points on the Delphi scale, 8 of them were diagnosed with enterocolitis.

**Conclusion:** This study shows the efficacy of the Delphi score in the identification of HAEC and the pattern of Hirschsprung disease presentation among the Sudanese population.

**Keywords:** Hirschsprung-Associated Enterocolitis; HAEC; Hirschsprung Disease; HD; Delphi Score

### Introduction

Hirschsprung-associated enterocolitis represents a challenge in the diagnosis and management with great variability of reported incidence. This urged the need for an international standardized scale to categorize cases with Hirschsprung disease and facilitate the early recognition of seriously lethal complications if not treated properly [1].

Definitive operative management and the ratio of patients experiencing episodes of Enterocolitis were linked as operative correction doesn't eliminate the risk. Many researchers reported a correlation between the incidences of this complication and Down's syndrome [2,3].

### Patients and Methods

All patients admitted to Soba and Ribat University hospitals with a confirmed diagnosis of Hirschsprung disease, the age group

from 1 week up to 16 years with no gender preference were included in this study. Those patients were classified according to gender, relation to the definitive operative management, presence of Down's syndrome, family history of the disease, cause of admission and duration of admission in the hospital placing the treatment protocol used into consideration.

The study was conducted as a retro-prospective, descriptive-analytic, hospital-based multicenter study for 21 months. From March 2015 till December 2015 patients were considered the retrospective group, and from January to December 2016 was the prospective group. Data were collected by a specially constructed questionnaire, entered and analyzed using the software Statistical Package for the Social Sciences (SPSS) Version 23.0 according to the objectives using multivariate analysis, Chi-Square and regression analysis tests. P-value was regarded as significant if < 0.05 within confidence interval (CI 95%).

**Ethical approval:** Permission was taken from the hospital administration, registry, and pediatric surgery departments with written approval. Consent was taken verbally from the parents of the patients during the admission for the prospective group, and for the retrospective group, they were contacted by phone.

**Results**

Males represent 41 (78.8%) of the patients while females were 11 (21.2%); with a male to female ratio of 3.7:1. Most of the studied patients 22 (42.3%) aged between 1 - 5 years; the majority 33 (63.5%) from central Sudan residence; in most cases, tribes were not mentioned 33 (63.5%) (Table 1). The majority of patients 20 (38.5%) were diagnosed with HD at age of 1 year; two types of diagnosis were reported including short segment HD 43 (82.7%) and long-segment HD 9 (17.3%) (Table 2).

Down's syndrome was reported in only 4 (7.7%) of the patients. At admission to operation 26 (50%) of the patients were preoperative the other half postoperative [9 (17.3%) early and 17 (32.7%) late postoperative]]. Causes of admissions were preoperative preparation in 13 (28.8%), postoperative recovery in 13 (25%), chronic constipation 14 (26.9%), and enterocolitis 10 (19.2%).

The management protocols at the hospital were IV antibiotics 48 (92.3%) followed by IV fluids 47 (90.4%), rectal wash 38

		Number	Percent
History	Diarrhea with explosive stool	3	5.8
	Diarrhea with foul-smelling stool	6	11.5
	Diarrhea with bloody stool	0	0.0
	Previous history of enterocolitis	18	34.6
Physical Examination	Explosive discharge of gas and stool on rectal Exam	10	19.2
	Distended abdomen	41	78.8
	Decreased peripheral perfusion	5	9.6
	Lethargy	15	28.8
	Fever	14	26.9
Radiology	Multiple air fluid levels	14	26.9
	Dilated loops of bowel	38	73.1
	Saw-tooth appearance with the irregular mucosal lining	1	1.9
	Cutoff sign in rectosigmoid with the absence of distal air		40.4
	Pneumatosis intestinalis	3	5.8
Laboratory	Leukocytosis	25	48.1
	Shift to left	11	21.2

**Table 1:** Patient score at admission. The overall mean score (5.44 ± 4.42).

(73.1%), NPO 29 (55.8%), and nasogastric tube 18 (34.6%) (Table 3). Duration of admission was 7-14 days for 23 (44.2%) of the patients, followed by 1 - 7 days 22 (42.3%) and more than 14 days 7 (13.5%).

On admission, the mean value of the Delphi Score was 5.44 ± 4.42. The patients attained more than 10 points on the Delphi scale, of them 8 diagnosed with enterocolitis. The frequency of each item of the score (history, physical examination, radiology, and laboratory) are shown in table 1.

**Discussion**

The early diagnosis and recognition of Hirschsprung disease is crucial in the appropriate management of patients and can be considered as a critical factor in the prevention of enterocolitis [4]. Although most pediatric surgical centers strictly adhere to these

		1 - 7 days	7 - 14 days	> 14 days	P Value
Gender	Male	16	21	4	0.001
	Female	6	2	3	
Age of the patient	0-12 month	8	6	2	0.017
	1 yr - 5 yrs	9	9	4	
	6 yrs - 10 yrs	5	6	0	
	11 yrs - 16 yrs	0	2	1	
	0-12 month	8	6	2	
Patient residence	Central Sudan	15	14	4	0.012
	Eastern Sudan	2	2	0	
	Northern Sudan	2	1	0	
	Western Sudan	3	6	3	
Down's syndrome	Yes	0	4	0	0.028
	No	22	19	7	
Age of HD diagnosis	1 <sup>st</sup> month	6	6	2	0.011
	1 <sup>st</sup> year	8	8	4	
	Bet. 1 - 5 yrs.	7	7	0	
	Bet. 6 - 10 yrs.	1	0	1	
The state of admis. to operation	Preop.	16	9	1	0.013
	Early postop.	3	4	2	
	Late postop.	3	10	4	
Family history of HD	Yes	0	2	0	0.022
	No	22	21	7	

**Table 2:** The factors significantly related to Duration of admission.

guidelines, HAEC continues to be a serious, often life-threatening condition.

In our study, 52 pediatric patients were enrolled to determine the incidence and pattern of presentation of Hirschsprung-associated enterocolitis among Sudanese children presenting to Soba and Ribat university hospitals and the outcome of management.

		1 - 7 days	7 - 14 days	> 14 days	P Value
Cause of admission	Preop. preparation	10	5	0	0.018
	Postop. recovery	3	8	2	
	Enterocolitis	0	6	4	
Management at hospital	Chronic constipation	9	4	1	0.016
	NPO	7	16	6	
	NG tube	3	9	6	
	IV fluids	18	22	7	
Antibiotic selection and usage	IV antibiotics	19	22	7	0.023
	Rectal wash	12	19	7	
	No antibiotic	4	1	0	
	2 <sup>nd</sup> generation cephalosporin + metronidazole	9	8	0	
	3 <sup>rd</sup> generation cephalosporin + metronidazole	8	14	7	
Other	Other	1	0	0	0.023
	No	22	22	7	

**Table 3:** The factors significantly related to duration of admission.

Among the patients enrolled in this research 20 (38.5%) were diagnosed with HD in the first year of their childhood and 14 (26.9%) diagnosed during their neonatal period. Moreover, enterocolitis was reported in 10 (19.2%) of the studied patients. Similar to other studies, criteria that received the highest importance scores were diarrhea, explosive stools, abdominal distension, and radiologic evidence of bowel obstruction or mucosal edema [3,5-7].

Two categories were reported according to the diseased segment including short segment HD 43 (82.7%) and long-segment HD 9 (17.3%), comparable to Menezes., *et al.* case series of fifty-six patients (75.6%) with rectosigmoid disease and 18 (24.3%) with long-segment disease [8]. The incidence of the short-segment disease is four times greater in males than in females; equal numbers of males and females present with long-segment HD Badner., *et al.*

Elhalaby, *et al.* showed increased frequencies of HAEC in patients with the long-segment disease compared with those with short aganglionic Segment disease, 49% vs 31%, respectively [9]. Similar Data from Reding, *et al.* report a 56% incidence of HAEC in the preoperative period among children with the long-segment disease compared with a 16% incidence among children with short-segment disease ( $P < 0.01$ ) indicating that long-segment aganglionosis upsurges the risk of developing enterocolitis [10].

Down syndrome was reported in only 4 (7.7%) of our patients, HD and trisomy 21 have known association with 2.9% - 8.2% of HD patients also having trisomy 21 reported by Frykman, *et al.* while other studies by Teitelbaum, *et al.* and Coran, *et al.* respectively showed a range of 14 to 16% occurrence [3,11-13].

The patient's status to the definitive operation 26 (50%) of them was preoperative the other half were postoperative [9 (17.3%) early and 17 (32.7%) late postoperative]. Ten patients were diagnosed as HAEC according to the clinical presentation and Delphi scores seven of them (13.3%) of the total number were postoperative. some researchers described that patients who have HAEC preoperatively are prone to recurrent episodes in the postoperative period, Other studies by Harrison and Carneiro have shown no association between the occurrence of preoperative and postoperative HAEC [14,15].

HAEC was higher among males than females, however other authors in a previous study conducted by Elhalaby, *et al.* showed that the increased incidence of HAEC among female gender and although an increased incidence of HAEC has been reported in girls (42% vs 30%, ( $P= 0.16$ ) with no statistical significance [9].

The management protocols at the hospital were IV antibiotics 48 (92.3%) followed by IV fluids 47 (90.4%), rectal wash 38 (73.1%), NPO 29 (55.8%), and nasogastric tube 18 (34.6%). Antibiotic therapy is commenced with broad-spectrum intravenous antibiotics in severe diseases. C. difficile cover must be ensured [16] as aggressive therapy and resuscitation with monitoring, follow up and application of preventive strategies to ensure a clear protocol in handling patients of HD and suspected HAEC which was recommended by Frykman, *et al* [1].

## Conclusion

This study shows the efficacy of the Delphi score in the identification of HAEC and the pattern of Hirschsprung disease presentation among the Sudanese population.

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