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ORIGINAL ARTICLE

The Clinical Impact of Delayed Management of Hirschsprungs Disease

Khaled Safwat El Sayed Fahmi¹, Ali Helmy Abdelwahed Elshewy¹, Ahmed Ezzat Rozeik² and Emad Ali Ali Alshatti³

¹ General Surgery Department , Faculty of Medicine , Zagazig University, Zagazig ,Egypt

² Pediatric Surgery Department, Faculty of Medicine , Zagazig University, Zagazig, Egypt

³ General Surgery Department, Faculty of Medicine ,Zawia University, Libya

Corresponding Author:

Name: Emad Ali Ali Alshatti*

Email:

imadashatti88.com@gmail.com

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ABSTRACT

Background: Hirschsprung's disease (HD) is a developmental disorder of the enteric nervous system that is characterized by the absence of ganglion cells in the myenteric and submucosal plexus from the anorectum for a variable colonic or intestinal distance. HD is a congenital bowel motility disorder which occurs in approximately one of every 5000 live births. This results in a lack of peristalsis leading to a huge dilatation of the colon proximal to the aganglionic bowel. **Aim of work:** The purpose of the present study is to investigate the clinical impact of delayed management of children with HD. and how it would affect their outcomes? **Methods:** The present study was performed in the period from July 2018 to august 2019 in Pediatric Surgery Department of Zagazig University Hospital. 18 patients enrolled in prospective clinical study **Results:** The delayed diagnosis of HD is associated with a different pattern of presentation compared with neonatal HD. Most of the cases were presented by chronic constipation, abdominal distension and bilious vomiting. Moreover, outcome is worse in older children who are diagnosed with this disease. **Conclusions:** HD in older children associated with long hospital stay, increased cost of treatment compared with the younger children. Postoperative outcome in older children associated not only with increased risk of serious morbidities but also indirectly affect their long-term outcome.

Keywords: Hirschsprung's disease, Aganglionic, Developmental, Outcomes .

INTRODUCTION

Hirschsprung's disease (HD) is a developmental disorder of the enteric nervous system that is characterized by the absence of ganglion cells in the myenteric and submucosal plexus from the anorectum for a variable colonic or intestinal distance. HD is a congenital bowel motility disorder which occurs in approximately one of every 5000 live births^[1]. This results in a lack of peristalsis leading to a huge dilatation of the colon proximal to the aganglionic bowel^[2].

Short-segment disease is most common and is confined to the rectosigmoid region of the colon. Long-segment disease extends past this region and can affect the entire colon. Rarely, the small and large intestines are involved^[3]. Late diagnosed HD is a challenge in developing countries because

it may have a negative impact on the management of HD and rule out one stage operative procedure. The bowel in this patients is chronically dilated especially the colon proximal to the aganglionic bowel tends to be severely dilated^[4].

Late diagnosed HD requires a challenging management by adequate bowel and general preparation. Rectal wash out is the best way to prepare the colon even it is a demanding procedure, but when it becomes ineffective, transverse colostomy can be an option. One stage transanal endorectal pull-through (OTEPT) is a safe option with less morbidity. Plication procedure avoids anastomosis incongruence^[5].

Lee and his colleagues,^[6] reported that, delayed diagnosis of HD in children not only increases serious morbidities in the neonatal period, but also indirectly affects their long-term outcome.

Clinicians need to be vigilant in the recognition of HD in children and provide appropriate care accordingly, to prevent the detrimental complications.

Early postoperative complications, especially anastomotic leakage, occurred frequently in children with late-diagnosed HD. Therefore, a diverting stoma should be considered in these patients. The long-term functional results were comparable with those seen in children operated upon as neonates^[4].

Previous studies have suggested that Hirschsprung's-associated enterocolitis (HAEC) occurs more often in patients whose diagnosis of HD is delayed.

Haricharan and Georgeson^[7] reported that delayed diagnosis is also associated with poorer long-term outcomes^{[8], [9]}. However, these studies involved only older children.

The purpose of the present study is to investigate the clinical impact of delayed management of children with HD, and how it would affect their outcomes?

METHODS

The present study was performed in the period from July 2018 to August 2019 in Pediatric and general Surgical Department of Zagazig University Hospitals. 18 patients enrolled in prospective clinical study. **Inclusion criteria:** Patients who diagnosed at 3 years or more by contrast enema and rectal biopsy, HD with rectosigmoid type. **Exclusion criteria:** Patients below 3 years, Patients have additional gastrointestinal anomalies, Unfit for surgery, Trisomy 21 or HD longer than rectosigmoid.

All patients in the study were subjected to; **Clinical assessment** and **Laboratory investigations** such as complete blood picture (CBC) that measured by automated blood counter, Liver function tests: serum bilirubin (total and direct), serum albumin, serum alanine transaminase and aspartate transferase measured by kinetic method. Kidney function tests that includes serum creatinine, serum urea and serum uric acid by colorimetric assay. Bleeding profile; INR, PTT, PT also estimation of Serum Ferritin and estimation of Serum electrolyte when needed.

Radiological investigations as Plain abdominal x-rays done for diagnosis of fecal stasis, air fluid level, cut off sign, dilatation of proximal bowel and Contrast enema for demonstration of dilated colon, transitional zone.

Rectal biopsy was used to give definitive diagnosis.

Preoperative colonic preparation; Serial rectal washouts (enemas) (10ml/kg) and digital

dilatation of the rectum was performed before the surgery is begun. One percent Neomycin should be added to the final rectal irrigation. Intravenous antibiotics are given within 1 hour preoperatively.

Operative plan: Some patients were subjected to transanal endorectal pull through and some to trans-abdominal in pediatric surgery Department in Zagazig University Hospitals.

A-Steps of operation: The patient was placed in a prone position with the pelvis elevated. As a first step to the transanal mucosectomy of the rectum, the anal canal is exposed, and a circumferential incision is made 1 cm above the dentate line in the rectal mucosa. Using blunt dissection, a submucosal plane is developed using multiple 5/0 silk traction sutures in the mucosa to facilitate its separation from the muscular wall. This submucosal plane is extended 6 cm. The next step is to prepare the muscular sleeve through which the normoganglionic colon can be pulled. At the same site at which mucosectomy was finished, a complete incision on the rectal muscle is made to reach into the perirectal tissue. To liberate the muscular sleeve, perirectal tissue is dissected and smooth muscle fibers of the rectum are divided circumferentially. Then, the muscular sleeve is returned to its original position. Finally, a posterior myotomy of the sleeve is made above the place, where the anastomotic line would be created. Then, the rectum is pulled down and the mesenteric vessels are dissected, tied and divided. Thus, the colon is free and could be pulled through. Then, the macroscopic transitional zone could be seen. When the normoganglionic colon is reached and freed without tension up to the proposed anastomotic line. To securely fix the normally ganglionic pulled through colon, two absorbable sutures are placed in the lateral walls of the muscular sleeve. The anastomosis to the rectal mucosa is sutured with absorbable material.

B-Postoperative: The bladder catheter was removed on the 1st postoperative day. Postoperative bowel peristalsis was audible within 24-48 hours. Oral feeding started within 5 days postoperatively when audible intestinal sound, followed by soft diet 24-48 hours later. Third generation cephalosporin was administered at induction of anesthesia and continued for 7-10 days after operation.

Follow up: Outpatient visits were arranged weekly for one month and every 2 weeks for 3 months and then monthly thereafter. Rectal examination was performed two weeks after operation with the finger or Hegar dilator for assessment of the anastomosis. Anal dilatation is started after 2 weeks, at first, 3 times dilatation per week using

Hegar dilators then decrease of frequency of dilatation till 3 months.

Ethical Clearance: Written Informed consent was taken from the patient parents to participate in the study. Approval for performing the study was obtained from Pediatric Surgery Departments, Zagazig University Hospitals after taking Institutional Review Board (IRB) approval. The work has been carried out in accordance with the code of ethics of the world medical association (Declaration of Helsinki) for studies involving humans.

STATISTICAL ANALYSIS

All data were collected, tabulated and statistically analyzed using SPSS 22.0 for windows (SPSS Inc., Chicago, IL, USA). Continuous data are expressed as the mean ± SD & median (range), and the categorical data are expressed as a number (percentage). Continuous variables were checked for normality by using Shapiro-Wilk test. Mann Whitney U test was used to compare two groups of non-normally distributed data. Categorical data were compared using Chi-square test or Fisher's exact test when appropriate. All tests were two tailed. p-value < 0.05 was considered statistically significant (S), p-value < 0.001 was considered highly statistically significant (HS), and p-value > 0.05 was considered statistically insignificant (NS).

RESULTS

The table shows most of the cases were presented by chronic constipation , abdominal distension and history of delayed passage of meconium with 11 cases were presented with bilious vomiting . Table (1):This table showed operative data of cases with mean of operative time was 145.55 minutes and mean of postoperative hospital stay was 14.54 days.

Table (2):The table shows 8 cases did trans anal pull through, 6 of them was done colostomy (4 sigmoid , 2 transverse). four cases did trans abdominal and all of them done transverse colostomy, Also table show 6 cases did combined procedures, 4 of them done colostomy (2 sigmoid – 2 transverse). Table (3): The table shows Postoperative complications with 3 cases had bowel obstruction, 7 cases had enterocolitis which treated by nothing per mouth bowel rest, IV fluid , nasogastric tube, and triple antibiotic. Three cases had abdominal wound infection which treated by culture and sensitivity, repeated dressing and antibiotic. Seven cases had perianal excoriation which treated by use of barrier cream and antibiotic. Six cases had anastomotic leakage two of them responded well to conservative treatment in the form of NPO, IV fluid and antibiotic, and the other 4 did diverting ileostomy. Six cases had anal stenosis and 3 cases had constipation presented by difficult defecation which treated conservatively , soft diet ,use of laxative and regular dilatation. Table (4):This table showed the duration of follow up range from 6_10 months. The incontinence scores in present study were from 18 to 21 , the incontinence score were within the age-specific related 95% confidence interval (CI) for eight patients and superior to upper CI for tow patients and 3 patients had scored constipation below their age-specific lower limit 95% CI which indicates the presence of incomplete continence and constipation. Table (5) : This table showed decrease of squeezing contraction in majority of cases (13 patients) 72.2% and present in the 5 patients and also the resting tone decreased in most of the patients (11 patients) and normal in seven cases . Table (6):

Table (1): Clinical presentation of the studied patients (N=18)

Clinical presentation	The studied patients (N=18)	
	No.	%
Delayed passage of meconium	14	77.8%
Abdominal distension	15	83.3%
Bilious vomiting	11	61.1%
Chronic constipation	17	94.4%

Table (2): Operative data of the studied patients (N=18)

Operative data	The studied patients (N=18)				
	Mean	±	SD	Median	(Range)
Operative time (minutes)	145.55	±	26.61	150	(120 – 190)
Hospital stay	14.54	±	3.47	14.25	(10 – 18)

(days)				
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Table (3) Operative procedures of the studied patients

Operative procedurs	The studied patients (N=18)	
	No	%
Trans ananl pull through	8	44.4%
Trans abdominal	4	22.2%
Combined (trans anal – trans abdominal)	6	33.3%
With colostomy	14	77.78%
Without colostomy	4	22.2%

Table (4): Postoperative complications and mortality of the studied patients.

Postoperative complications	The studied patients (N=18)	
	No.	%
Bowel obstruction	3	16.6%
Enterocolitis	7	38.89%
Constipation	3	16.6%
Anal stenosis	6	33.3%
Anal excoriation	7	38.9%
Anastomotic stricture	4	22.2%
Anastomotic leakage	6	33.3%
Wound infection	3	16.67%
Death	0	0%

Table (5): Postoperative clinical assessment of the studied patients (N=18).

Postoperative clinical assessment	The studied patients (N=18)				
	Mean	±	SD	Median	(Range)
Follow-up duration (months)	7.16	±	1.38	6.50	(6 – 10)
Incontinence score(n=8)	19.72	±	1.48	20.18	(18 – 21)
incontinence score normal	24.1	±	2.57		(22-26)
Constipation score(n=10)	19.83	±	1.09	20	(17 – 23)
normal constipation score	21.1	±	2.13		(20-22.4)

Table (6): Postoperative EMG of the studied patients (N=18)

Postoperative EMG	The studied patients (N=18)	
	No.	%
Resting tone		
Normal	7	38.89%
Subnormal	11	61.1%
More than normal	0	%0
Squeezing contraction		

Normal	5	27.78%
Subnormal	13	72.2%
More than normal	0	0%

DISCUSSION

Most of the cases were presented by chronic constipation, abdominal distension, bilious vomiting and history of delayed passage of meconium. This result was in agreement with Sarin et al., [10] who reported 75% of study presented with typical feature of intestinal obstruction. Diagnosis depend on clinical examination and all of the patients did gastrographin enema which show picture suggesting dilated colon , level of transitional zone and confirmed by rectal biopsy .

Eight patients underwent transanal endorectal pull through, 6 of them did colostomy (4 sigmoid , 2 transverse). Four cases did trans abdominal and all of them done transverse colostomy, Also table show 3 cases did combined procedures, 4 of them done colostomy (2 sigmoid – 2 transverse). Operative data of cases with mean of operative time was 145.55 min and mean of postoperative hospital stay was 14 days. This is in agreement with Qi Li et al., [11] which was mean operative time 160± 35.0 min (range 150–250) and mean postoperative hospital stay 15 ± 3.7 days (range 11–21 days) after pull-through procedures.

Hirschsprung's disease in older children associated with long hospital stay, increased cost of treatment compared with the younger children (less than 3 years). Although, the decreased incidence of HD in older children but the risk of postoperative complications higher than younger children and require more than one stage of pull through procedure unlike the younger children which may need one stage .

In the present study, the postoperative complications were, 3 cases (16.6%) had bowel obstruction, 7 cases (38.89%) had enterocolitis which conservatively treated by nothing per mouth , nasogastric tube, bowel rest, IV fluid , and triple antibiotic, six cases had anastomotic leakage two of them responded well to conservative treatment in the form of NPO, IV fluid and antibiotic, and the other 4 did diverting ileostomy .

Six (33.3%) cases had anal stenosis and 3 cases had constipation presented by difficult defecation which treated conservatively by soft diet, use of laxative and regular dilatation, Three cases (16.67%) had wound infection which treated by culture and sensitivity, repeated dressing and antibiotic. Seven cases (38.9%) had perianal excoriation which treated by use of barrier cream

and antibiotic. This is in line with Choe et al., [12] which reported 20% had bowel obstruction and in line with Hashish et al., [13] which demonstrated that study with 24 patients with 3 patients (13.6%) had anastomotic leak, 10 patients (41.6%) had perianal excoriation, Six patients (25%) had enterocolitis and seven patients (29.6%) had wound infection.

Hirschsprung's disease in the older child portends a worse outcome compared with younger children. Specifically, there was a difference in the complications as constipation, small bowel obstruction, enterocolitis anastomotic leak. On other hand, the incidence of morbidity and mortality increased in older children were directly attributable to risk of postoperative complications. In these study we assessed ano-rectal function by using Pediatric incontinence/constipation scoring system (PICSS Score).The incontinence scores in present study were from 18 to 21 , the incontinence score were within the age-specific related 95% confidence interval (CI) for eight patients and superior to upper CI for tow patients and 3 patients had scored constipation below their age-specific lower limit 95% CI, which indicates the presence of incomplete continence and constipation .

Electromyogram used in our study postoperatively to test ano- rectal function which showed decrease of squeezing contraction in majority of cases (13 patients) 72.2% and present in the five patients and also the resting tone decreased in most of the patients (11 patients) and normal in seven cases . Also showed neuropathic changes in the majority of the cases in the form of large amplitude, prolonged duration and wide polyphasic motor unit.

CONCLUSION:

Postoperative outcome in older children associated not only with increased risk of serious morbidities but also indirectly affect their long term outcome. HD in older children associated with long hospital stay, increased cost of treatment compared with the younger children. The stooling function is satisfactory in most of the patients after operation and the trans-anal endorectal pull-through operation is a safe and reasonable procedure for delayed management of hirschsprung's disease. Continence in older children is one of the serious complications that affect there quality of life. Growth and development is adequate however, nutritional

elements as hemoglobin, ferritin, albumin, are affected and become at low normal level so close monitoring of patient and supplementation is mandatory. Longer period and with greater sample size are needed to confirm the findings in this study.

conflicts of interest. None

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