Surgical Management and Outcomes of Hirschsprung Disease in Pediatric Age Group in Ain Shams University Hospital

Thesis

Submitted for Partial Fulfillment of the Master Degree in General Surgery

Presented By

Mohamed Hassan Ahmed Mohamed
MBBCH 2013

Under Supervision of

Prof. Dr. Ahmed El Sayed Mourad
Professor of General Surgery
Faculty of Medicine, Ain Shams University

Dr. Ayman Mostafa Allam
Lecturer of Pediatric Surgery
Faculty of Medicine, Ain Shams University

Dr. Mohamed Gamal El Sayed
Lecturer of General Surgery
Faculty of Medicine, Ain Shams University

Faculty of Medicine
Ain Shams University
2018

سورة البقرة الآية: ٣٢

Acknowledgment

First and foremost, I feel always indebted to **ALLAH,** *the Most Kind and Most Merciful.*

I'd like to express my respectful thanks and profound gratitude to **Prof. Ahmed El Sayed Mourad,** *Assisstant Professor of General Surgery, Faculty of Medicine, Ain Shams University for his keen guidance, kind supervision, valuable advice and continuous encouragement, which made possible the completion of this work.*

I am also delighted to express my deepest gratitude and thanks to **Dr. Ayman Mostafa Allam,** *Lecturer of Pediatric Surgery, Faculty of Medicine, Ain Shams University, for his kind care, continuous supervision, valuable instructions, constant help and great assistance throughout this work.*

I am deeply thankful to **Dr. Mohamed Gamal El Sayed,** *Lecturer of General Surgery, Faculty of Medicine, Ain Shams University, for his great help, outstanding support, active participation and guidance.*

I would like to express my hearty thanks to all **my family** *for their support till this work was completed.*

Last but not least my sincere thanks and appreciation to all patients participated in this study.

Mohamed Hassan Ahmed Mohamed

List of Contents

Title	Page No.
List of Tables	i
List of Figures	ii
List of Abbreviations	iii
Introduction	1
Aim of the Work	3
Review of Literature	
▪ Anatomy, Histology and Pathophysiology	4
▪ Presentation and Clinical Findings	16
▪ Diagnosis	19
▪ Other Causes of Colonic Motility Disorder	26
▪ Operations and Surgical Intervention	35
▪ Comparison between Different Surgical Techniques	52
Materials and Methods	62
Results	65
Discussion	71
Conclusion	81
Limitations	82
Summary	83
References	85
Arabic Summary	

List of Tables

Table No.	Title	Page No.
Table (1):	Descriptive data of patients who underwent Soave and Swenson techniques in relation to sex, age at operation, weight at operation	66
Table (2):	Operative details of Swenson and soave patients in relation to total length of resected colon, incision above dentate line	67
Table (3):	Operative data in both groups soave and Swenson patients in relation to post operative start of intestinal function and total hospital stay	68
Table (4):	Comparison between Swenson and Soave pull Through operations as regard postoperative complications of anastmotic leakage, early postoperative HAEC, postoperative intestinal obstruction (IO), wound infection and stenosis at site of anastmosis.	69

List of Figures

Fig. No.	Title	Page No.
Figure (1):	Blood supply of colon.	7
Figure (2):	Innervation of colon.	10
Figure (3):	Innervation of colon showing Auerbach's plexus and Meissner's plexus.	13
Figure (4):	Pathogenesis of HD showing narrow part (aganglionic part) and wide part (ganglionic part)	14
Figure (5):	Abdominal distension.	17
Figure (6):	Narrow distal segment.	19
Figure (7):	GG enema show level marked by an arrow.	20
Figure (8):	The level between rectum and sigmoid is marked by arrow.	21
Figure (9):	Anorectal manometry.	22
Figure (10):	(A) Normal rectoanal inhibitory reflex. (B) Abnormal increased contraction of anal canal and no relaxation of internal sphincter with rectal distension	23
Figure (11):	Histologic features of HD.	25
Figure (12):	Different techniques of repair.	38
Figure (13):	Transanal endo rectal pullthrough.	40
Figure (14):	Baby with colostomy.	42
Figure (15):	Laparscopic assisted pull through.	43
Figure (16):	Transanal endorectal pull through.	45
Figure (17):	Algorithm for having recurrent attacks of postoperative HAEC.	49
Figure (18):	Percentage of complications in comparison between Swenson and soave patients.	70

List of Abbreviations

Abb.	Full term

ACE.....*Antegrade colonic enema*
AChE .....*Acetyl-Choline Esterase Enzyme*
ARM.....*Anorectal manometry*
AXR.....*Abdominal x-ray*
Ba.....*Barium*
BT .....*Botox*
CIPO .....*Chronic Intestinal Pseudo-Obstruction*
Cm.....*Centimeter*
CM .....*Colon manometry*
CNS .....*Central Nervous System*
DL .....*Dentate Line*
ENSC.....*Enteric Nervous System stem cells*
EUPSA.....*European Paediatric Surgeons Association*
FT.....*Full Term*
FTB.....*Full thickness biopsy*
GG.....*Gastrograffin*
H .....*Hour/s*
H&E.....*Haematoxlin and Eosin*
HAEC.....*Hirschsprung's Associated Entero-Colitis*
HAPC.....*high-amplitude peristaltic contractions*
HD.....*Hirschsprung' s Disease*
IAS.....*Internal anal sphincter*
IASA .....*Internal Anal Sphincter Achalasia*
IC .....*Intractable constipation*
IMA.....*Inferior mesenteric artery*
IO.....*Intestinal obstruction*
IS.....*Internal Sphincter*
Kg.....*Kilogram/s*
Ml.....*Millimeter*

List of Abbreviations cont...

Abb.	Full term
MSPT	*Multi-stage pull through*
NEC	*Necrotizing Entero-Colitis*
No.	*Number*
NPO	*Nothing Per Oral*
PA	*Perineal Approach*
PN	*Parenteral nutrition*
PO	*Postoperative*
PT	*Pull-Through*
QOL	*Quality Of Life*
RMD	*Rectal mucosal dissection*
ROM	*Radio-opaque marker*
RSB	*Rectal suction biopsy*
SM Cuff	*Seromuscular Cuff*
SMA	*Superior mesenteric artery*
SSPT	*single stage pull-through*
TERPT	*Transanal endorectal Pull-through*

ABSTRACT

Background: Hirschsprung disease (HD) as described by Hirschsprung is a cause of constipation in early infancy, it is a complex disorder resulting from absence of ganglion cells in the bowel wall leading to functional obstruction and bowel dilatation proximal to the affected segment. Hirschsprung disease (HSD) occurs in approximately 1 in 5000 live births with an overall male: female ratio of 3: 1 to 4: 1.

Purpose: To review our outcome in managing cases of Hirschsprung disease in pediatric surgery department Ain Shams University children Hospital during the period from January 2014 to December 2016.

Patients and Methods: During the period from January 2014 to December 2016, patients with Hirschsprung's disease presented at Pediatric Surgery Department, Ain Shams University Hospitals age from 2 months to 12 years were retrospectively analyzed and included. Patients with redo surgeries were excluded.

Results: Twenty five pediatric patients with HD [21 boys (84%) and 4 girls (16%)] were retrospectively analyzed and included in the period of the study at Pediatric Surgery Department, Ain Shams University Hospitals from January 2014 to December 2016.

Conclusion: Single staged pull through is more common than multiple staged pull through with percentage 64% and 34% respectively. Soave is done in 52% of cases while swenson done 48% with no statistically difference between them as regard outcomes and post operative complication.

Keywords: *Hirschsprung Disease - Single Stage Pull-Through - Transanal Endorectal Pull-Through*

INTRODUCTION

Hirschsprung disease (HD) as described by Hirschsprung is a cause of constipation in early infancy, it is a complex disorder resulting from absence of ganglion cells in the bowel wall leading to functional obstruction and bowel dilatation proximal to the affected segment. [1] Hirschsprung disease (HSD) occurs in approximately 1 in 5000 live births with an overall male: female ratio of 3: 1 to 4: 1 [2, 3].

Children with HD come to medical attention with the following symptoms: Delayed passage of meconium (>24 h after birth), Abdominal distension that is relieved by rectal stimulation or enemas, Vomiting, Neonatal enterocolitis, severe constipation, Chronic abdominal distension, Failure to thrive. The diagnosis of HD is based on a combination of symptoms, radiological study as plain abdomen x-ray, Gastrografin enema, rectal manometry, and rectal biopsy [2, 3]. Full thickness Rectal biopsy is the gold standard for the definitive diagnosis of aganglionosis [4].

The most commonly performed repairs are the Swenson, Duhamel, Soave procedures. Recently, minimally invasive laparoscopic procedures have been introduced for the treatment of HD. Long-term complications of pull-through procedures may include intermittent enterocolitis, severe stool retention, as well as intestinal obstruction. [5]

Despite significant developments in the understanding of the pathologic anatomy and physiology of HD, the results of surgical therapy need more evaluation for assessment of the outcomes. It is therefore essential to assess clinical outcome in patients surgically treated for HD as any abnormality in this aspect would adversely affect the quality of life and growth.[5]

Aim of the Work

To review our outcome in managing cases of Hirschsprung disease in pediatric surgery department Ain Shams University children Hospital during the period from January 2014 to December 2016.

Chapter 1

ANATOMY, HISTOLOGY AND PATHOPHYSIOLOGY

Anatomy:

The colon is a 5-6–ft long, inverted, U-shaped part of the large intestine (lower gastrointestinal tract). The large intestine is within the alimentary tract where water is absorbed from indigestible contents. The large intestine includes the cecum, appendix, entire colon, rectum, and anal canal. It begins at the terminal ileum with the cecum. Unlike the small intestine, it has a shorter length but a much larger lumen. It is distinguished further from the small intestine by the presence of omental appendices, haustra, and teniae coli By definition, the cecum (and appendix) and ano-rectum, which are parts of the large intestine, are not included in the colon.

Embryologically, the colon develops partly from the midgut (ascending colon to proximal transverse colon) and partly from the hind gut (distal transverse colon to sigmoid colon). [6]

On plain abdominal radiographs, the colon is seen to be filled with air and some fecal material. The colon is identified with haustra (irregular incomplete sacculations confer regular complete valvulae conniventes in jejunum).

Ascending colon

The ascending (right) colon lies vertically in the most lateral right part of the abdominal cavity, occupying the right iliac fossa, right lumbar region and right hypochondrium. The proximal blind end (pouch) of the ascending colon is called the cecum. The ascending colon takes a right-angled turn just below the liver (right colic or hepatic flexure) and becomes the transverse colon, which has a horizontal course from right to left, occupying the right hypochondrium, epigastrium, and left hypochondrium.[7]

Transverse colon

The transverse colon again takes a right-angled turn just below the spleen (left colic or splenic flexure, which is attached to the diaphragm by the phrenocolic ligament) and becomes the descending (left) colon, which lies vertically in the most lateral left part of the abdominal cavity, occupying the left hypochondrium, left lumbar region, and left iliac fossa. Splenic flexure is higher (cranial) to hepatic flexure. The descending colon leads to the inverted V-shaped sigmoid colon, which then becomes the rectum at the S3 level; the sigmoid colon is so called because of its S-shape. [6, 7, 8, 9, 10]

Paracolic gutters

Lateral to ascending and descending colon are the right and left paracolic gutters of the peritoneal cavity, through which

fluid/pus in the upper abdomen can trickle down into the pelvic cavity. The ascending and descending colon are related to the kidney, ureter, and gonadal vessels of the corresponding side that lie behind them in the retroperitoneum; the ascending colon is also related to the C loop (second part) of the duodenum.[8]

Transverse colon and sigmoid colon

The transverse colon and the sigmoid colon have a mesentery (ie, transverse mesocolon and sigmoid mesocolon, respectively), but the ascending colon and descending colon are retroperitoneal, while the cecum is intraperitoneal but uses the mesentery of the ileum. The base of the transverse mesocolon lies horizontally across the duodenum and pancreas. The greater omentum has several parts, including the 4-layered omental apron hanging down off of the transverse colon and the 2-layered gastrocolic ligament connecting the greater curvature of the stomach and the transverse colon.

The rectum occupies the concavity of the sacrococcygeal cavity. It is fixed, primarily retroperitoneal, and subperitoneal in location. It transitions to the anal canal at the level of the puborectal sling which is formed by the fibers of the levator ani muscles. The rectum has an expanded middle segment called the ampulla. Anterior to the rectum are the rectovesical pouch, prostate, bladder, urethra, and seminal vesicles in males. In females, it is the recto-uterine pouch, cervix, uterus, and vagina.