INTUSSUSCEPTION

A COMPARATIVE STUDY BETWEEN EARLY & LATE CASES OF INTUSSUSCEPTION

A THESIS
SUBMITTED TO THE IRAQI BOARD FOR MEDICAL SPECIALIZATION IN PARTIAL FULFILMENT OF THE REQUIREMENT FOR THE DEGREE OF FELLOWSHIP IN PAEDIATRIC SURGERY

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وَفَوقَ كُلِ ذِي عِلمٍ عَلِيم

صدق الله العظيم

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DEDICATION

I feel very much grateful and indebted to my family for their support, patience and love.

Acknowledgment

I would like to express my sincere thanks and great gratitude to Dr. MUNTHIR AL OBAIDI for his supervision, kind guidance and support for the completion of this study.

And I sincerely acknowledge the invaluable contributions of my colleagues, nursing staff of the Pediatric Surgical Ward of CWTH, and the operation theatre staff for making this work possible.

I certify that this thesis was prepared under my supervision at the scientific council of pediatric surgery in partial fulfillment of the requirement for the degree of fellowship of the Iraqi Board for Medical Specializations in Pediatric surgery.

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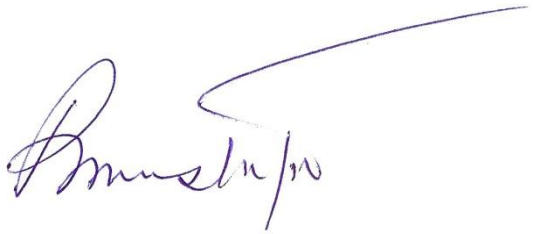

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LISTS OF ABBREVIATIONS

CWTH	Children welfare teaching hospital
RTI	Respiratory tract infections
U/S	Ultrasound
LUTH	Lagos University Teaching Hospital
EEA	End-to-End Anastomosis.
BPR	Bleeding Per Rectum.
AXR	Abdominal X-Ray.

تداخل الامعاء

دراسة مقارنة بين الحالات المبكرة والمتاخرة

دراسة مقدمة الى المجلس العلمي لجراحة الاطفال كجزء من متطلبات

نيل درجة زمالة المجلس العراقي للاختصاصات الطبية في جراحة الاطفال

من قبل

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الملخــص

الهدف :

دراسة حالات تداخل الامعاء حول الاعراض والعلامات السريرية للاطفال ودراسة دور التشخيص بواسطة الفحص الشعاعي والامواج فوق الصوتية مع بيان كيفية معالجتها من خلال التداخل الجراحي اللائق وما يترتب عليها من نتائج ومشاكل جانبية مع بيان كيفية معالجتها. كما تناولت الدراسة مقارنة بين التشخيص المبكروالتشخيص المتاخرلتداخل الامعاء من حيث الاسباب والنتائج.

التصميم :

دراسة مستقبلية لمئة واثنى عشر حالة مصابة بتداخل الامعاء للفترة مابين تشرين الاول 2006 ولغاية تشرين الاول 2008.

موقع الدراسة :

تم معالجة كل المرضى قيد الدراسة في مستشفى حماية الاطفال التعليمي-مدينة الطب في بغداد.

الوسائل المتبعة :

بالاضافة إلى الاعراض والعلامات السريرية تم الاعماد في التشخيص على فحص الرقائق الشعاعية في وضع الوقوف للبطن. بالاضافة الى استخدام الموجات فوق الصوتية في المساعدة بتشخيص الحالات الصعبة.

النتائج :

يعتبر تداخل الامعاء من الاسباب الشائعة لانسداد الامعاء في سن الطفولة وخاصة بين عمر 5 شهر و 2 سنة. كما قد يحدث في الاطفال اقل من 5 شهر او اكثر من 2 سنة ولكن بنسبة اقل نسبيا.

ان نسبة التشخيص المتاخر لتداخل الامعاء تعتبر نسبة عالية (43%).

تمت المقارنة بين الحالات المشخصة بصورة مبكرة (64 حالة) والحالات المشخصة بصورة متاخرة (48 حالة) من حيث اسباب التشخيص المتاخر والنتائج المصاحبة لذلك.

تراوحت اعمار المرضى بين 17 يوم و7 سنوات. كما كانت نسبة المرضى المصابين بتداخل الامعاء بين عمر 5 شهر و2 سنة حوالي 76,8%. ان تاخر التشخيص قد يكون مصاحبا للعمر اقل من 5 شهر او اكثر من سنتين.

يصورة عامة, ان تداخل الامعاء اكثر شيوعا في الذكور وتعتبر الاناث احد الاسباب المؤدية الى تاخر التشخيص.

ان الفترة بين بدء الاعراض والوصول ال المستشفى تراوحت بين 9 ساعة و 12 يوم.

عانى جميع المرضى من الام البطن, كما عانى المرضى من الاعراض التالية وبنسب متفاوتة: تقئ المادة الصفراء, كتلة البطن, نزف دموي عن طريق المستقيم, وغيرها.

ان تزامن الاعراض التالية (الام البطن, نزف عبر المستقيم, وكتلة البطن) له اهمية قصوى في التشخيص حيث حدث في 67,9% من الحالات المبكرة و 77,1% من الحالات المتاخرة.

ان تاخر تشخيص المرض اكثر من يومين ادى الى زيادة نسبة استاصال الامعاء مع الربط الاولي الى 16.1% كما ادى الى تفويه الصائم في 3.5% من المرضى.

يعتبر التشخيص السريري العنصر الاساسي في تشخيص تداخل الامعاء. كما بمكن الاستعانة بالامواج فوق الصوتية في تشخيص الحالات الصعبة.

ان نسبة المضاعفات اناتجة عن تداخل الامعاء كانت 20.5%, 4,8% للحالات المبكرة و 41,8 للحالات المتاخرة. علما ان تسرب الربط الجراحي, انسداد الامعاء الالتصاقي, وحالتي الوفات قد حدثت في الحالات المتاخرة فقط.

تراوحت فترة دخول المستشفى بين 2,5 يوم و 12,9 يوم. معدل البقاء في المشتشفى كان 3,4 يوم للحالات المبكرة و 6,1 يوم للحالات المتاخرة.

وكانت الاسبات وراء تاخر التشخيص كمايلي: -

1. العمر الشاذ.
2. الجنس انثى.
3. شذوذ الاعراض.
4. نوع تداخل الامعاء.
5. نقص في الوسائل التشخيصية.
6. اهل المريض.
7. الكادر الطبي والصحي.

الاستنتاجات والتوصيات :

1. يعتبر تداخل الامعاء احد الاسباب المهمة والشائعة لانسداد الامعاء في سن الطفولة وخاصة بين 5 شهر و سنتين.
2. ان تداخل الامعاء من الامراض المهددة للحياة مالم يتم تشخيصه بصورة سريعة ومعالجته بطريقة مناسبة.
3. ا ان تاخر تشخيص المرض من قبل الكادر الطبي والصحي غير المختص وما يتبعه من تاخر احالة المريض ال مراكز جراحة الاطفال التخصصية هو السبب الرئيسي وراء تاخر تقديم العلاج اللازم للمريض وبالتالي تدهور الحالة المرضية وزيادة نسبة الوفاة.
4. ان نسبة الوفاة وكذلك المضاعفات المصاحبة لتداخل الامعاء ناتجة عن التشخيص المتاخر للمرض والذي يؤدي الى تسمم الدم الناتج عن التهاب البريتون الجرثومي والجفاف الشديد. علما ان السبب الرئيسي لتاخر التشخيص هو الكادر الطبي والصحي. بالاضافة الى ذوي المريض والاعراض الشاذة احيانا.
5. يعتبر الفحص السريري الوسيلة الاساسية لتشخيص تداخل الامعاء. كما يمكن الاستعانة بفحص الامواج فوق الصوتية او الاشعة الملونة في تشخيص الحالات المشتبه بها.
6. لابد من استخدام الوسائل غير الجراحية لعلاج تداخل الامعاء. والاستعانة بالتداخل الجراحي اذا تطلب الامر.
7. تعتبر التوعية الصحية من الاركان الاساسية لتقليل المضاعفات الناتجة عن تداخل الامعاء وذلك عن طريق النشرات الصحية وحملات التوعية الصحية بالاضافة الى وسائل الاعلام.

Summary

Objectives

This study deals with the methods of treatment of congenital megacolon (Hirschsprung's disease) and evaluate the results of the multiple stages that used in the surgical treatment.

Design

This is a prospective study of (84) cases of Hirschsprung's disease treated by modified Soaves procedure (Boley's procedure) .

Site

The study consider all children presented to the children welfare teaching hospital in Baghdad with signs and symptoms of Hirschsprung's disease over 3 years from October 2005 to October 2008.

Results

The total number of cases were 84 patients with an age ranging from two days to six years and the mean age was (8 months); the commonest age group at presentation were those between (1-12) months(58.33%) of the cases; the male: female ratio was (3.6:1).

Sixty eight patients (81%) have positive history of delayed passing meconium more than 48 hours, and 54 patients (64.28%) presented with constipation and progressive abdominal distention. 13 patients (15.47%) presented with classical picture of neonatal intestinal obstruction, and other cases presented with perforation (7.14%), enterocolitis (9.52%), and fecal impaction (3.57%).

Rectal biopsy was the confirmatory method of diagnosis of HD in our study , all rectal biopsies were negative for ganglion cells.

Two methods of treatment are used according to the number of stages of surgery:

- **Two stages management**: Twenty nine patients managed firstly by diversion colostomy, and the second stage Boley's procedures were done without protective proximal stoma. Twelve patients (41%) had excellent results and the postoperative period was free of any complication.

- **Three stages management**: Thirty one patients managed firstly by diversion stoma, and the second stage Boley's procedures were with protective proximal stoma with less complication; 15 patients (48.4%) were free from complications, but 12 patients (46%) from 26 patients developed complications after closure of the protective stoma.

Conclusions

1) Boley's technique is a good procedure in the management of Hirschsprung's disease, which is one of the classical problems in pediatric surgery procedures,
   It provides good continence (no pelvic dissection).
2) Enterocolitis is a constant complication in both 2 stages and 3 stages methods.
3) Three stages technique is a safe way to avoid major complications.
4) There is no big difference between the two methods in postoperative complications, in addition one can avoid the complication of the colostomy closure by the 2 stages method.

Introduction

Hirschsprung's disease is defined as a lack of propagation of propulsive waves and an abnormal or absent relaxation of the internal anal sphincter due to aganglionosis, hypoganglionosis, or dysganglionosis.

Historical review:

- In 1691 , Fredrick Ruysch reported the autopsy finding ,of a child who died with what appear to be a congenital megacolon.[1]
- In 1886 , Hirschsprung's , a Danish pediatrician , delivered a lecture in which he described two boys who presented with a characteristic clinical picture: severe difficulty of defecation from birth associated with increasing abdominal distension and a deterioration in health, leading to death at the age of 7 and 11 months respectively. The autopsies revealed severe dilatation, bowel wall hypertrophy and mucosal ulceration of the colon.[2]
- The first reference relating the absence of ganglion cells to the underlying disturbance is that of Tittle in 1901.[3]
- Robertson and Kernohan in 1938, and Zuelzer and Wilson in 1948 were able to correlate the functional disturbances of the distal colon with aganglionosis.[4,5]
- Swenson and colleagues, in 1949 described the main principles for the radiological diagnosis of this condition.[6]
- The first rational surgical approach for management of congenital megacolon was reported by Swenson and Bill in 1948.[7]
- Rehbien in 1953, performed low anterior resection of aganglionic intestine, which extend distally well beyond the peritoneal reflection.[8]
- Duhamel [9] and Soave [10] subsequently revealed the two technique of retro-rectal and endorectal pull-through that are most commonly used during the last forty years.

- Further refinement of these techniques (modified Soave) was performed by Boley. [11] Recently laparoscopic management of Hirschsprung's disease is used in some centers.[12,13]
- Another approach called transanal resection is applied in the last few years in some centers.[14,15] and gaining acceptance by pediatric surgeons without exploring the abdomen.

EMBRYOLOGY AND ETIOLOGY

In normal embryological development, neuroenteric cells migrate from the neural crest to the upper end of the alimentary tract and then proceed in distal direction.[16]
The first nerve cell arrives in the esophagus by the fifth week of gestation, migration to the distal colon is achieved by the 12th week. The neuroenteric cells are guided in their migration by various neuronal glycoproteins.[17]
In Hirschsprung's disease, failure of the neuroblast to achieve the most distal migration is the causative factor.[18]. some time, actual cellular migration may occur, however, these neuroblast are then placed in a hostile or non-supportive environment that cause failure of ganglion development.Kuroda, Doody, and Donahoe [19] have demonstrated increased expression of class 2 antigens in the mucosa and submucosa of patients with Hirschsprung's disease.
There might be an autosomal dominant mode of inheritance with incomplete penetrance leading to long – segment aganglionosis , and an autosomal recessive or multifactorial mode of inheritance responsible for short-segment Hirschsprung's disease

PATHOLOGY

The gross features of Hirschsprung's disease vary with the duration of untreated disease . In the neonatal period, the intestine may appear fairly normal, as the child ages, the proximal aganglionic intestine hypertrophies and become thicker and longer than normal ,the taenia disappear. The transitional zone from ganglionic to aganglionic segment may be funnel like and vary in length.
The absence of ganglionic cells in the distal intestine is the hallmark of the disease. The ganglionic cells are absent in both the submucosal (Meissner) as well as the intermuscular plexus (Auerbach) . A marked increase in nerve fibers, which extend into the submucosa associated with absent ganglionic cells.

Aganglionosis typically extend to the rectosigmoid region in approximately 80% of cases.[20,21] . The incidence of various levels of aganglionsis is shown in table 1.

Table 1 : Extent of aganglionosis in Hirschsprung's disease

Site of the Transitional zone	POLLEY [21]	KLEIN & PHILIPART [22]	VANE & GROSFIELD [23]
Rectosigmoid colon	74%	58%	81%
Long segment of intestine	11%	26%	10%
Total colon	12%	12%	9%
Small intestine	3%	4%	0%

PATHOPHYSIOLOGY

The intrinsic autonomic nervous system of the bowel consist of:

1. Three distinct plexuses of ganglion cells with their neural connection :
   a. Auerbach's plexus, lies between the circular and longitudinal muscle.
   b. Henles or the deep submucosal plexus is along the margin of circular muscularis propria.
   c. Meissners plexus, lies immediately beneath the muscularis mucosa.

2. Cholinergic fibers from the vagal and the pelvic out flow also terminate in the intrinsic plexus, primarily in the distal rectum and internal sphincter.[24]

These plexuses contain an integrated neuronal network that act to control all function of the gut (absorption, secretion, blood flow and motility) with relatively little control from the body's CNS.[25]

Normal intestinal motility is primarily controlled by these intrinsic nervous system in each ganglion, loss of extrinsic control still allow for adequate function of

intestine.[26] These ganglion cells can cause contraction or relaxation of smooth muscles, however, relaxation seems to predominate under normal circumstances.

Extrinsic control of the intestine is through both preganglionic cholinergic fibers and postganglionic adrenergic fibers.

The cholinergic fibers result in contraction through the transmitter Ach. The adrenergic fibers are predominantly inhibitory but also contain some excitatory pathway and use norepinephrine to mediate their function.

The intestine also contains an intrinsic nervous system with a neurotransmitter that is inhibitory in nature, this system is referred to as noncholinergic nonadrenergic nervous system (NCNA) and in the 1990 Bult et al [27] identified nitric oxide (NO) as the most likely mediator of this system.

In classical Hirschsprung's disease the ganglionic cells are absent from all three plexuses. The space normally occupied by ganglionic cells is taken up by large longitudinally oriented nerve fibers.[28]

Bowel dysfunction in Hirschsprung's disease is the result of complex malformations of the intrinsic nervous system of the bowel, which include the absence of the ganglion,(NANC) nervous system interneuron, different peptidergic nerve fibers, and probably connective tissue structure of the bowel wall. [29]

HYPOGANGLIONOSIS

Proximal to the aganglionic segment of Hirschsprung's disease, there is generally a zone of hypoganglionosis.[30]

Hypoganglionosis can also represent an isolated disease entity, which is defined as the state in which the number of ganglion cells is reduced by a factor of 10 and the density of nerve fibers by a factor of 5.[30,31]

The number of nerve cells in the myenteric plexus is 50% of that of the normal innervated colon, and the distance between the ganglions is doubled.

Hypoganglionosis some time involves only a short segment of the colon and occasionally may involve the whole colon.

IMMATURITY OF THE GANGLION CELLS

Immature ganglion can be seen with intestinal neuronal dysplasia (IND) or hypoganglionosis and the condition may eventually cause bowel obstruction.[42]
Immature ganglion cells with monopolar small dendrite can be identified by lactate dehydrogenase staining.

Nerve cell in the immature ganglion have not developed a dehydrogenase containing cytoplasm, therefore; a differentiation between Schwann's cells and nerve cells cannot be made.

INCIDENCE

The incidence of Hirschsprung's disease range from 1 in 4400 to 1 in 7000 live births. [33] . The male to female ratio in patients with classic Hirschsprung's disease is generally reported as 4:1 in favors of males.[34]

In long segment disease, the ratio approaches 1:1 and may actually become reversed.[35]. The incidence of Hirschsprung's disease increases in familial cases to approximately 6% .

Bandar et al [36] calculated the risk for transmission of Hirschsprung's disease to relatives. In his study brothers of patient with short segment Hirschsprung's disease have a higher risk 4% than sister 1% .

In long segment Hirschsprung's disease brothers and sons of affected female have the greatest risk for being affected (24% and 29% respectively).

ASSOSCIATED MALFORMATIONS [37]

Associated malformations are present in 11% to 30% of the children with Hirschsprung's disease . The most common disorders were of the:

1. Urogenital tract 11%, voiding problems frequently occur in patients with an enlarged rectum , which compresses the bladder neck, with subsequent bladder neck obstruction and megacystis.
2. Cardiovascular system 6% .
3. Gastrointestinal system 6% ; congenital atresias of the small and large bowel meconium ileus , meconium plug and imperforate anus . Hypoganglionosis and often (IND) are frequently associated with (HD).
4. Various other malformations like cataract , coloboma, cleft palate and extremities or cerebral defect at 8% .
5. Prematurity is reported as many as 10% of the children with (HD).
6. Down's syndrome 4.2%-6%

DIAGNOSIS

The signs and symptoms of (HD) are highly variable, on one hand it may appear in the neonatal period with acute presentation of abdominal distention, vomiting, and failure to pass meconium. On the other hand, the disease may persist untreated into adult life.[28].

Ninety five percent of full term infants will pass their first stool with in the first 24 hours of life, and the remainder will pass their first stool with in the first 48 hours of life .[38]. Ninety four percent of infants with (HD) are seen to have had delayed passage of meconium. This is the cardinal symptom that ,if appreciated , allows the diagnosis in the neonatal period.[38]

Other presenting signs and symptoms include:

Abdominal distention, poor feeding and emesis with constipation. This constipation will either progress to severe obstruction or infant will develop enterocolitis (vomiting, progressive abdominal distention, and paradoxically explosive diarrhea).

As the disease progress peristalsis diminishes and then stops. At this point of intestinal decompensation, there is massive abdominal distention.
In the usual case abdominal distention wax and wane in response to formula changes, enema and laxatives . The mother may hear loud borborygmi.
In the older children there is typically severe abdominal distention with dilated loops of intestine out lines through thin abdominal wall.
Differentiation of Hirschsprung's disease from other forms of constipation can be difficult.
Fecal soiling is generally absent in older children but may occur in 4% of children especially with short segment disease .

The physical examination of baby often shows a full term healthy newborn baby with a distended abdomen, when the little finger is gently introduced into the anus, the rectal wall always feel tight and resist further probing.

In infants a visible and palpable transverse colon with an explosive passage of flatus and foul smelling liquid stool in response to rectal examination suggest Hirschsprung's disease .
In older children there is an increase of anteroposterior diameter of the chest and there are palpable fecal impaction in the abdomen. Classically the rectum will be empty , small amount of stool is found in up to 40% , and 15% have rectal impaction.[28]

Hirschsprung's disease is suspected on the basis of history and clinical findings . Radiological examination , anorectal manometry, and histochemical analysis of biopsy specimens establish the diagnosis.

Radiological diagnosis

The first diagnostic test in most children should be plain supine and upright films of abdomen, which show air fluid level in the colon; and one often can identify a dilated segment of transverse colon crossing the upper abdomen. The classical finding in the barium enema is that of a normal – caliber rectum or narrow distal segment, a funnel – shaped dilatation at the level of transition zone , and a marked dilation of the proximal colon.[28] barium enema is a diagnostic tool , but its limitation must be appreciated.

Error in interpretation of the barium enema rest on two factors:

- Age : in neonates the proximal bowel has not yet dilated in contrast to the aganglionic zone. The x-ray diagnosis in the neonate is improved by obtaining a film 24 hours later that shows retained barium.
- The length of the aganglionic segment:
    - In a total colon aganglionosis ; because the entire colon is small but is not a true microcolon
    - In short segment; because the column of barium may obscure a narrow rectal segment.

Contrast enema examination had been reported to be inconclusive in 10% of children with confirmed Hirschsprung's disease and in (29%) of cases with (HD) associated intestinal neuronal dysplasia (IND) type B.[39]

Children with enterocolitis may show thickening of the bowel wall with mucosal irregularity and grossly distended bowel loops on plain film. In these cases, a characteristic transitional zone may not be present due to inflammatory impairment of muscular function in the normally innervated colon.

Anorectal electromanomertry: (ARM)

The diagnostic accuracy of ARM for Hirschsprung's disease has been reported to be as high as 85% [39,40] . Normally , distention of the rectum using a balloon results in relaxation of the internal sphincter.

Contractions or waves were pathognomonic .[40] . However, ARM can be misleading in newborns, as the normal rectosphincteric reflex is completed by 12th day of life . Although the accuracy of manometry increases with patient's age , it is never

sufficiently accurate to make a definitive diagnosis of HD without confirmatory biopsy evidence. Consequently, manometry is at best only a rough screening test.[41]

In a highly sophisticated study conducted in a special manometric laboratory , there were 10 false positive and 8 false negative results in 229 examinations. There was a 26% error rate in infants under 1 months of age.[42,44]

Rectal biopsy

The confirmation of the diagnosis is based on the absence of ganglion cells and the presence of an excess of nonmyelinated nerves in adequate rectal biopsy. [42]

The specimen must be taken at least 1.5 cm above the pectinate line. [42]

The traditional full-thickness rectal biopsy has obvious diagnostic value. However, the tissue is difficult to obtain since good rectal exposure is required and under general anesthesia.

Since the introduction of suction biopsy technique [43], the procedure become less traumatic and can be performed without anesthesia. Suction biopsy specimens are taken at 2cm,3cm,5cm and if possible higher up above the dentate line. The optimal size of biopsy is approximately 3.5mm in diameter to include submucosa.

In children with suspected hypoganglionosis or heterotopias of the myenteric plexus, full-thickness biopsies are recommended.

In suction biopsy, as many as 20 to 50 sections may be required to confirm the presence of ganglion cells, which are smaller and more difficult to identify than in the Auerbachs plexus. No errors of interpretation were found in a series of 42 patients when two adequate specimens from each patient were examined.[43]

Histochemichal technique

Normal patients show barely detectable acetyl cholinesterase activity in the intestinal mucosa, when ganglion cells are absent , there is an overabundance of acetylcholine, and consequently, of the corresponding enzyme acetyl cholinesterase. When acetyl cholinesterase stains are used; it is necessary to look at only one section to make a definitive diagnosis.

TREATMENT

The enteric nervous system ENS is the brain of the intestine. Similar to the brain, it functions automatically to some degree, even when some neural mechanisms are deficient.[27] . Therefore affected newborn may be admitted with intestinal obstruction where as others present later in childhood or adulthood with chronic constipation.[27]

Once symptomatic, most patient, require decompression as the first step of management. The choice of treatment depends on the child's age and general condition. However, rather repeated rectal irrigation or a colostomy will be indicated before the definitive operations.

Rectal irrigation are indicated to:

a) Decompress the neonate while awaiting a definitive diagnosis.
b) Treat enterocolitis.
c) Remove fecal impaction in older children.

Irrigation is not an enema! Saline is instilled into the rectum through a rubber catheter with a syringe and then allowed to run out through the catheter by gravity. The procedure is repeated several times a day until the infant's bowel is decompressed. During this time, oral mineral oil and mineral oil retention enemas are given to soften the impaction if present. After the impaction has been broken up once daily irrigation is sufficient to keep the bowel empty. During this time, the mother is taught how to give the irrigation.[29]

A colostomy is indicated in :

1. Any child who has had enterocolitis.
2. In younger children when irrigation fail.
3. Older children who are malnourished and have hugely distended colon.

The colostomy should be placed just proximal to the transition zone in the most distal portion of the bowel with ganglion cells. Biopsy must be taken from the bowel wall to accurately determine the proper colostomy site.

A blindly placed transverse colostomy should be avoided because in 10% of patients the aganglionosis extends across the ascending colon. Furthermore, a transverse colostomy attaches the colon to the anterior abdominal wall, and there may be insufficient length of bowel to bring it to the perineum during the reconstruction. Even if the bowel distal to the stoma is long enough, it would be small in diameter, and when anastomosed to

the rectal stump, it is more likely to get stricture. Finally if the colostomy is left in place, a third operation will be necessary for its closure.

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We usually create a loop colostomy. Care should be taken not to narrow the proximal stoma, which might create a partial obstruction.

Suturing the afferent and efferent segment of the loop together can prevent a prolapsed of the bowel.

Definitive procedures:

Swenson and **Bill** performed the first resection of an aganglionic segment in 1948, [27] since then 3 other basic techniques have been developed.

Swenson Technique

The patient is positioned to provide surgical access to the abdomen and perineum at the same time. The proximal colon and mesentery are dissected to achieve a sufficient length for reconstruction. The peritoneal reflection at the rectosigmoid is then incised and the deep pelvic dissection commenced. The dissection is performed close to the rectal wall to protect the pelvic autonomic nervous system. Division of rectosigmoid is accomplished by a stapling device at a convenient level. The rectosigmoidal stump then inverted through the anus. The mucocutaneous line should be clearly visible.

An oblique incision is made through the anterior half of prolapsed rectum and a clamp is inserted into the pelvis to grasp and pull the proximal ganglionic segment through the anus then an extra anal anastomosis is performed with interrupted suture material.

Duhamel procedure

The principle of this procedure is preservation of the internal anal sphincter, opening of the retrorectal space only, followed by retrorectal pull through of the ganglionic part of the colon, and elimination of colorectal septum.

Today the retrorectal anastomosis is most often performed using a stapling devise [45].

Anterior resection according to Rehbein

Rehbein technique differs from Swenson's procedure in that, the anastomosis is a low, anterior colorectal anastomosis . The pelvirectal dissection is completed leaving the aganglionic terminal 2 to 3cm of the rectum in infants and 4 to 5cm in older children with vigorous sphincter dilation with Hegar bougies.

The anastomosis may be performed using a circular end to end anastomosis stapling instrument introduced through the anus ,[46] or by direct suture deep in the pelvis.

Soave procedure "Modified Soave"

The Soave procedure is an ingenious and appealing operation. Because the aganglionic rectosigmoid is removed by an endorectal dissection, theoretically minimizing the risk the risk of the pelvic injury associated with the Swenson procedure. The normally innervated colon is passed through anorectosigmoid muscular cuff. There is no aganglionic segment of rectum left, such as that, which occurs with the Duhamel procedure. This operation was originally performed without colostomy, leaving a portion of the pulled through colon protruding well beyond the anal skin margin, this was then excised at a second operation two weeks later.

This two- stage procedure was modified by Boley into a one- stage operation by effecting a primary anastomosis to the anal verge.

The endorectal dissection is initiated usually 1 or 2cm above the peritoneal reflection . It is recommended that the endorectal dissection be carried down to approximately 1 or 2cm above the dentate line in order to preserve the sensitive anal mucosa.

Laparoscopic pull through technique

Advancement in minimally invasive surgery and instrumentation had resulted in pull through procedure being performed using laparoscopic technique. Most surgeons who treat HD laparoscopicaly have used a modified Swenson pull through technique. [48] But the Duhamel method and the Soave technique have been performed laparoscopicaly as well. Operative times required for the laparoscopic Swenson procedure have been reported to be similar to those for open technique.[47,48] .The duration of the Duhamel procedure was somewhat longer[49] .The experience with the technique remains limited to special centers.

Primary pull-through

Primary pull through in the newborn was introduced mostly by surgeons who prefer Boley's procedure.[53,54]

They postulate that endorectal dissection is more difficult to perform in older children than in infants because of tenacious adhesions in the submucosal plane caused by chronic proctitis and daily enemas.

During the first 3 months of life and especially in the newborn, the rectum shows less inflammation and the dissection between the submucosal layer and muscular cylinder is easy to perform. Primary endorectal pull through usually does not require a protective colostomy. The reported results of primary pull through are not better than those of staged treatment. In a reported series of 24 patients, 1 died, 1 developed bowel volvulus , 9 (39%) suffered from recurrent enterocolitis and 12 (42%) were constipated.[55]

Primary transanal endorectal pull through is a single stage transanal modified pull through done without abdominal incision, which is an another approach applied in the last few years in some centers.[14,15]

Treatment of Total Colonic Aganglionosis (TCA)

For the treatment of TCA, Martin [50,51] introduced a long side to side anastomosis between the normal ileum and the aganglionic descending colon and the rectum. The operation should be performed at the age of 1 year, where the performing of the rectal and pelvic anastomosis is easier.

Unfortunately, Martin modification of Duhamel procedure has not completely eliminated the complications, such as frequent and liquid stools, excoriated perineum, enterocolitis and nighttime incontinence, which occur in as many as 66% of patients.[52] Therefore, we prefer the Rehbein deep anterior resection in patients with total colonic aganglionosis.[52]

The persistent rectal achalasia increase transit time and the resorption capability of the ileum without increasing the frequency of enterocolitis.

Aganglionic patch (Kimura procedure) [6]

Another technique that has recently become popularized is Kimuras utilization of a parasitized cecal patch:

1. The ganglionic ileum is first decompressed with an end ileostomy.
2. Several weeks later an extensive longitudinally ileocolostomy approximately 10-25 cm in length is created.
3. After 6-12 months, the ileocolostomy is used for the pull through with either a Swenson or endorectal technique.

Postoperative complications [29,56]

Early complications

Complications that become manifested with in the first 4 weeks after operation are usually the result of technical error or infection.[56]

1. Anastomotic leak : (7%) , Factors that increase the risk of leak include: tension on the anastomosis, ischemia of the rectal cuff or distal colonic segment, inadequate suturing, and distal obstruction. Always lead to stricture. Need emergency enterostomy and usually heal within 4-5 months.
2. Cuff abscess and retraction of the pull-through segment are the most serious complications after endorectal pull-through. Contamination of the cuff, incomplete removal of the mucosa, hemorrhage or insufficient drainage lead to cuff abscess or mucocele in 5% of patients.
3. Voiding dysfunction: more in older children, more after Swenson procedure followed by Duhamel procedure ; because of nerve damage during surgery and also may result from compression by the enlarged rectum before operation.
4. Anastomotic stricture.
5. Wound infection and intra-abdominal adhesion.
6. Early mortality only with total colonic aganglionosis.

Late complications [29]

More frequent during the first postoperative months and decrease within the following years.

1. Chronic constipation: mostly due to anal sphincter achalsia, incomplete resection , stricture, and fecaloma in case of Duhamel procedure. Stricture result from anastomotic leak, narrow cuff, or damage to the blood supply.
   Incomplete resection may need 2nd pull through.
2. Fecal incontinence (encopresis, soiling): more with Swenson's operation and least with Rehbein operation.
3. Enterocolitis
4. Long term voiding dysfunction: rare , more in older children with long lasting chronic constipation.
5. Sexual dysfunction: dyspareunia in female patients , primary infertility and poor erection in male patients.
6. Late mortality: 2% in Soaves , 5% in Swenson's operation.

AIM OF THE STUDAY

To evaluate the Boley's procedure as a definitive pull through procedure for Hirschsprung's disease .

PATIENTS AND METHODS

This is a prospective study of 84 cases of Hirschsprung's disease admitted to the children welfare teaching hospital in Baghdad over 3 years from October 2005 to October 2008.

Physical examination and investigations were done (plain abdominal X-ray, Barium enema,and rectal biopsy). All cases were confirmed by rectal biopsy.
Modified Soave "Boley" technique was done for (60) patients, pull through, by pulling the ganglionic part outside the anus and everting the submucosal-mucosal tube into the perineum to facilitate the performance of the anastomosis with the proximal ganglionic part just (1-2 cm) above the dentate line.

The presentation, technique of pull through, and complications following surgery were all analyzed and discussed.

Operative technique

The operation was performed by using a combined abdomino-perineal approach.

- The abdomen opened through a transverse infra-umbilical incision.
- Identification (and resection) of the aganglionic segment with the cone segment, the resection extended few centimeters into the normal colon, then its opening is closed with a silk sutures.
- The dissection started above the peritoneal reflection to separate the seromuscular layer from the mucosa and submucosa, which extended down to the anus.
- The cuff of the seromuscular layer is left as funnel shape, and fixed from the upper 4 angles by stay sutures.
- The perineal portion of the operation then started by inserting a long curved artery forceps through the anus and pushed to the site near by the closed lumen of remaining colon. This segment is pulled through the anal canal outside, so all the aganglionic segment can be resected outside the abdomen through the rectum.
- Then the anastomosis is carried down between the mucosa of the anal canal (above the dentate line) and the normal ganglionic colon.
- Or the aganglionic segment can be resected within the abdominal cavity.

RESULTS

SEX :

Eighty four patients (66 males and 18 females), the male to female ratio is (3.6 : 1)

Table 2: Male to Female ratio

Total number	Male	Female	M/F ratio
84	66	18	3.6 : 1

Age

Their age range from 2 days to 6 years (17 patients at the neonatal age),(49 patients at the infantile age), and (18 patients older than 1 year).

Table 3 : Age distribution

Age	No.	%
Neonatal	**17**	**20.23**
Infantile	**49**	**58,33**
Older	**18**	**21.42**

Presentation

Seventy one patients had positive history of delayed passing meconium more than 48 hours, (13) patients their parents couldn't remember that.
Thirteen patients presented with classical neonatal intestinal obstruction (failure to pass meconium, abdominal distention, and repeated bilious vomiting).

Fifty four patients presented with constipation and progressive abdominal distention.
Three patients presented with fecal impaction; all were older than one year age.

Six patients presented with perforation ;(4) patients at the neonatal age (3) of them with cecal perforation and the forth one the perforation was in the terminal ileum, the remaining (2) patients at the infantile age (1) with cecal perforation at 3 months age , and the other with transverse colon perforation at 4 months age.

Eight patients presented with enterocolitis , and enterocolitis occurs in other (11) patients during the period of conservative management.

Table 4: The presentation

Result	Cases no.	Percentage
Neonatal intestinal obstruction	13	15.47
Constipation and progressive abdominal distention	54	64.28
Enterocolitis	8	9.52
Fecal impaction	3	3.57
Perforation	6	7.14
Total	84	100%

Investigations

Plain x-ray film :(in erect and supine position)
Revealed distended bowel loops with air-fluid levels in the colon and paucity of air in the rectum.

Barium enema: Barium enema reveal the classical finding of normal caliber (or narrow) rectum or distal segment, a funnel shaped dilatation at the transitional zone, and a marked dilatation of the proximal colon in 51 patients (78.4 %) from 65 patients. .

Rectal biopsy:

Rectal biopsy (full thickness) was the confirmatory method of diagnosis of HD in our study , all rectal biopsies were negative for ganglion cells.

Management

Left infra-umbilical divided colostomy after excision of the transitional zone has been done for 39 patients and the histopathological examination revealed absence of ganglion cells in the distal narrow end of the zone and presence of ganglion cells in the proximal dilated end of the zone, In addition; rectal biopsies also revealed absence of ganglion cells.

Multiple biopsies has been taken from different site of the bowel in all cases required surgery in the neonatal period or presented with perforation , the sites of biopsies were The rectum, transverse colon, appendix, and the site of the ileostomy.

The aganglionic segment was of short segment type (rectosigmoid) in 61 patients, long segment type in 17 patients, and total colonic aganglionosis in 6 patients were presented in the neonatal period, one of them presented with ileal perforation.

Table 5: the length of aganglionic segment

Aganglionic segment	No. of cases	%
Short segment (rectosigmoid)	61	72.61
Long segment (extended to the descending and transeverse colon)	17	20.23
Total colonic aganglionosis	6	7.14

Colostomy performed for 73 patients ,and ileostomy performed for 11 patients { those with total colonic aganglionosis and those with perforation },two of those presented with perforation and managed with ileostomy died post operatively. Left infra-umbilical divided colostomy after excision of the transitional zone has been done for 39 patients, one patient died post operatively due to preoperative enterocolitis and sepsis. Transverse colostomy has been done for 34 patients, one patient died during postoperative period due to sepsis, and one escaped from the management.

Pull-through operation with protective stoma has been done for 31 patients (25 with protective colostomy and 6 with protective ileostomy 3 of them already with ileostomy). Complications:

- High postoperative fever in 4 patients (12.9%)
- Severe wound infection in 4 patients (12.9%)
- Anastomotic leak in one patient (3.2%)
- Anastomotic stricture in 7 patients (22.5%), one of them required stricturoplasty

After closure of the protective stoma for 26 patients; the complications were closure leak in 1 patients (3.8%), severe wound infection in 3 patients (11.5%), burst abdomen in 2 patients (7.6%), enterocolitis in 4 patients (15.3%) one of them had recurrent enterocolitis and died, adhesive intestinal obstruction in 2 patients (7.6%)

Table 6 : complications following 3 stages management

Complications after Boley's procedure for 31 patients	No. of cases	%
High postoperative fever	4	12.9
Severe wound infection	4	12.9
Anastomotic leak	1	3.2
Anastomotic stricture	7	22.5
Total	16	51.6%
Complications after stoma closure for 26 patients	**No. of cases**	**%**
Closure leak	1	3.8
Severe wound infection	3	11.5
Burst abdomen	2	7.6
Enterocolitis	4	15.3
Adhesive intestinal obstruction	2	7.6
Total	12	46.15%

Pull-through operation without protective stoma has been done for 29 patients ,23 of them with previous pelvic colostomy. The complications were severe wound infection in 3 patients (10.3%), anastomotic leak in one patient (3.4%)(colostomy performed), fecal fistula in one patient (3.4%) (colostomy performed), anastomotic stricture in 8 patients (27.5%) (one required surgery), enterocolitis in 3 patients (10%), and adhesive intestinal obstruction in one patient (3.4%).

Table 7: complications following 2 stages management

Complications after Boley's procedure for 29 patients	No. of cases	%
Severe wound infection	3	10.3
Anastomotic leak	1	3.4
Fecal fistula	1	3.4
Anastomotic stricture	8	27.5
Enterocolitis	3	10.3
Adhesive intestinal obstruction	1	3.4
Total	17	65.38

Discussion

Sex

The male to female ratio is generally reported (4:1)[33]. In our study it's nearly the same (3.6 : 1). *Klein an et al* [57] ,reported male to female ratio (3.3 : 1)

Age

The mean age of diagnosis in the past few decades is decreasing. During 1960s the mean age was (18.8) months and it decreased to (2.6) months in 1980s *Klein an et al* [57] . In our study the mean age was (8) months.

The majority of our patients was presented below the age of one year (58.33%), (20. 23%) below one month and (21.42%) were presented above one year of age.

Presentation

Regarding the presenting signs and symptoms, 13 cases (15,47 %) presented with classical signs and symptoms of neonatal intestinal obstruction; which occur in 25% as mentioned by Orr J D[35]**.**

Ninety five percent of full term normal infants will defecate with in the first 24 hours of life, and the remainder will pass their first stool by 48 hours (Clark DA) [38]. In our study 71 patients (84%) had history of delayed passage of meconium.

We have 54 patients (64.28 %) presented with constipation and progressive abdominal distention, which is similar to that mentioned by Orr J D[35] **(69%)**

Six patients (7.14 %) presented with perforation ; nearly similar to that mentioned by Orr J D (5%); (4) patients at the neonatal age (3) of them with cecal perforation and the forth one the perforation was in the terminal ileum, the remaining (2) patients at the

infantile age (1) with cecal perforation at 3 months age , and the other with transverse colon perforation at 4 months age.

Eight patients (9.52%) presented with enterocolitis , and enterocolitis occurs in other (11) patients during the period of conservative management. So enterocolitis developed in 19 patients (22.6 %), which is reported to occur in (12-58 %) of patients with HD (Elhaapy AL, Carneiro PMR, Surana R).[58,59,60]

Management

The definitive pull-through procedure was modified Soave (Boley's) procedure .

Two stages management

Twenty nine patients managed firstly by diversion colostomy (23 patients with pelvic colostomy after transitional zone excision) and the second stage Boley's procedures were without protective proximal stoma i.e. pulling the ganglionic colon at the level of the previous stoma.

Twelve patients (41%) had excellent results and the postoperative period was free of any complication. One patient (3.4%)developed anastomotic leak treated by colostomy and closure of colostomy 6 months later on. One patients (3.45) developed fecal fistula also treated by colostomy and the parents of the child decided to treat him outside the country.\

In a study done in the C.S Mott children's Hospital in the university of Michigan medical center, between 1974 – 1995 they have performed 260 endorectal pull through. They performed 62 cases with this two stages technique, the results is shown in table 8.

Table 8: A comparative study in the university of Michigan

Postoperative Complications	Comparative study (62 patients)		Our study (29 patients)	
	No. of cases	%	No. of cases	%
Severe wound infection	2	3.2	3	10.3
Anastomotic leak	1	1.6	1	3.4
Fecal fistula	0	0	1	3.4
Anastomotic stricture	2	3.2	8	27.5
Enterocolitis	8	12.9	3	10.3
Adhesive intestinal obstruction	6	9.7	1	3.4
Total	19	30.6	17	58.6

In comparison to our study it was nearly the same results regarding the incidence of enterocolitis (12.9%). We have higher incidence of severe wound infection (10.3%) versus (3.2%) in the mentioned study.

We have much higher incidence of anastomotic stricture (27.5%) versus (3.2%) in the mentioned study; this high incidence may be due to over reporting of even mild degree of stricture.

Three stages management

Thirty one patients had (3) stages Boley's pull through.

The incidence of anastomotic leak (3.2%) and anastomotic stricture (22.5%) is nearly the same in the (2) stages management.

Wound infection was slightly higher (12.9%) may be due to contamination from the colostomy, and wound infection occurred in 3 patients(11.5%) after closure of the protective stoma.

After closure of the protective stoma; the incidence of enterocolitis was (15.3%); slightly higher than in those treated by 2 stages method; but higher incidence of adhesive intestinal obstruction developed (7.6%).

Anastomotic leak after colostomy closure occurred in one patient (3.8%) of the patients, and burst abdomen in 2 patients (7.6%).

The incidence of anastomotic leak (3.2%) is nearly the same as that mentioned by *Holschneider AM* [56] (4%) , but higher incidence of anastomotic stricture (22.5%) and wound infection (12.9%) than those reported in the same study (7%), (5%) respectively.

Conclusions

1. Boley's technique is a good procedure in the management of Hirschsprung's disease, which is one of the classical problems in pediatric surgery procedures, It provides good continence (no pelvic dissection)
2. Enterocolitis is a constant complication in both 2 stages and 3 stages methods.
3. Three stages technique is a safe way to avoid major complications.
4. There is no big difference between the two methods in postoperative complications, in addition one can avoid the complications of the colostomy closure by the 2 stages method.

Recommendations

- To achieve good continence and avoid voiding dysfunction, we recommend Boley's procedure because it doesn't require pelvic dissection which may cause injury to pelvic nerve plexus.

- Early diagnosis and management is important to avoid complicatios as enterocolitis and to avoid severe colonic distension which affect the pull-through procedure.

- Adequate determination of the level of aganglionosis is important to avoid incomplete resection that result in postoperative constipation.

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