ORIGINAL RESEARCH PAPER

General Surgery

PRESENTATION AND MANAGEMENT OF HIRSCHSPRUNG'S DISEASE IN INFANTS AT A TERTIARY CARE CENTRE AND OUTCOME OF PRIMARY PULL-THROUGH IN THESE PATIENTS.

KEY WORDS: HSD (Hirschsprung's disease), LATPT(laparoscopic assisted transanal pullthrotisugh, HAEC(Hirschsprung's associated enterocolitis)

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ABSTRACT

Hirschsprung's disease is a common cause of neonatal intestinal obstruction. The treatment of Hirschsprung's disease has evolved to a single stage pull-through. . The aim of study was to evaluate the feasibility of performing the primary pull-through surgery and to develop an optimum treatment protocol for patients of Hirschsprung's disease (HD) presenting to our institution.

It was a prospective cohort study of newborn and young infants with features of Hirschsprung's disease conducted in our department from Oct. 2014, till Oct. 2018. The diagnosis was based on history, clinical examination, water soluble contrast enema and a definitive rectal biopsy. Transition zone was confirmed by intra-operative frozen section. Patients with weight of more than 5kgs and absence of features of concurrent enterocolitis, were taken for surgery. Patients included in the study underwent primary pull-through by different methods were put on strict follow-up protocol for 24 months and results were evaluated.

Out of 40 patients of Hirschsprung's disease presented to SKIMS from Oct. 2014 till Oct. 2017, only 25 patients underwent primary pull-through. Mean age of patients at the time of pull-through was6.9±7.12 months and mean weight was 8.76 ±2.67kg. Delayed passage of meconium was seen in23 patients (92%). Preoperative rectal biopsy was done for confirmation of diagnosis and intra-operative frozen section for delineation level of aganglionosis. Laparoscopic transanal pull-through was most common operation done in 13 (52%) patients. Anastomotic leak was seen in 2(8%) patients, wound infection occurred 2(8%) patients, enterocolitis in1(4%) and constipation in 2 (8%) patients. One patient who had undergone trans-anal pull-through underwent a redo for a retained cuff.

Primary pull-through is feasible and safe option with excellent results in neonates and infants with Hirschsprungs disease.

INTRODUCTION

Hirschsprung's disease may present with neonatal intestinal obstruction, history of constipation dating back to the newborn period, or delayed passage of meconium . Among normal full-term infants, 98% pass meconium in the first 24 hours of life and the remainder will pass their first stool by 48 hours [1] It has always been said that over 90% of HD infants fail to pass meconium in the first 24 hours of life [2]. The diagnosis can be made with radiographic studies , anorectal monometry and a rectal biopsy [3]. Rectal biopsy is used to confirm the diagnosis

A number of different operations have been described for the treatment of HD. Swenson was first to advocate resection of this aganglionic segment rather than previous techniques concentrating on the proximal dilated colon.[4]. In the past a multi-stage surgical resection of the aganglionic bowel was done for Hirschsprung's disease which was highly morbid. In 1980, So and colleagues were the first to report a one-stage pull-through procedure in neonates with Hirschsprung's disease without a preliminary colostomy. Primary endorectal pull-through without enterostomy[5] has gained in popularity since first described in 1980, being further modified to include minimally invasive approaches.[6]

A single stage primary pullthrough surgery avoids massive disfiguring scars over the abdomen , repeated hospital admissions with associated social and psychological problems and multiple surgical procedures under anaesthesia. Most importantly this procedure avoids requirement for a colostomy with its associated morbidity[7]. In this article, we will demonstrate our experience with neonates presenting with HSD and feasibility of performing the single stage primary pull-through surgery and pitfalls in performing the study.

MATERIAL AND METHODS:

The study was prospective cohort study. It was conducted in the Department of Pediatric Surgery, SKIMS, Soura from Oct. 2014 to Oct. 2018. After proper approval by the institutional ethical committee, all the neonates and infants with suspected Hirschsprung's disease were admitted, registered and evaluated on inpatient basis.

Patients were diagnosed as Hirschsprung's disease based on clinical history, radiological studies and full-thickness rectal biopsy. Water-soluble contrast helped identify a transition zone between a narrowed aganglionic and dilated, otherwise normally innervated segments. A rectal biopsy was done in patients to confirm HSD . On-table rectal frozen was done to confirm level of aganglionosis.

Once diagnosed patients were put on a definite rectal wash-out protocol. Parents were taught how to give home based colonic washes with normal saline using a 16 french soft drain till the patient was optimised for a primary pull-through and attained a weight significant enough to withstand a major surgery. Decision regarding timing of surgery in infants

diagnosed as Hirschsprung's disease was taken depending on the absence of features of enterocolitis, significant weight gain of around 5kgs and age of baby feasible to withstand significant anesthetic and surgical risks. This decision was tailored to individual cases.

Patients underwent pull-through by different methods depending on patients age, segment involved, feasibility of performing laparoscopy, and clearity of transition zone. In case tasransition was not still seen levelling biopsies were taken to do a pull-through at a later stage.

Photograph Figure File Showing A Neoanate Presenting With Abdominal Distension Whose Contrast Study Reveals A Clear Transition Zone.

RESULTS

During this study period 40 children were diagnosed as having Hirschsprung's disease and only 25 patients underwent primary pull-through and were included in the study. The reason for abandoning primary procedure in these patients are cited in table 1.

Table 1; Table Showing Reason For Adopting Multistage Surgery.

Reason for multi-stage surgery	Frequency(n)	Percentage(%age)
Perforation peritonitis	3	20
Patients not deflating on wash-out protocol and Patient having poor weight gain on wash-out protocol	7	46.7
Non compliance of parents with *Recurrent HAEC(Hirshsprung's associated enterocolitis)*	*4*	*26.7*
Gross discrepancy between dilated and constricted segments	1	6.7
Total	15	

Male were three times the number of females with a ratio of 3.2:1. Majority i.e, 21 patients (84%) underwent pull through in age less than 1 yr. 14 patients underwent a primary pull through in age group of 3-6 months which is an appropriate age in our set up. Only 4 patients underwent pull-through after age of 12 months either due to late presentation or some comorbidity. Mean age at the time of pull-through was 6.9±7.12 months. Weight of patients ranged from 2.5- 3.5 kgs on presentation. Paients were put on a wash-out protocol till at least a target weight of upto 5kg was reached. The mean weight at the time of pull-throgh was8.76±2.67kg. Table 2 shows clinical symptoms and signs which strongly suggested presence of Hirschsprung,s disease in study patients.

Table 2:

Table 2 showing various clinical symptoms and signs which strongly suggested presence of Hirschsprung,s disease in study patients		
Symptoms	Frequency	Percentage
DPM	23	92
Features of failure to thrive	10	40
Abdominal distension and features of obstruction	20	80
Blast sign on DRE	7	28
Hirschsprung's associated enterocolitis	0	0
Constipation	20	80

Water soluble contrast enema was done in all the patients. The water soluble rectal contrast study had a sensitivity of 48% . Final diagnosis was confirmed by a rectal biopsy

Transition was not seen in 5 patients when they underwent diagnostic laparoscopy. In these patients levelling biopsies were taken and a single stage pull-through was done at a later date after levelling biopsy reports were available. Table 3: Showing surgical procedure done in HSD

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Type of surgery	Frequency	Percentage
LATPT(Soave-Boley type)	13	52
Primary Duhamels pull-through .	1	4
TAPT	6	24
Diagnostic LAP+Biopsy followed by a Primary pull-through later.	5	20

Standard length disease was seen in 18 patients. 6 patients had extended segment disease with transition in the descending colon and one patient had transition in transverse colon. Laparoscopic pull-through was the most common procedure done in our patients 52% followed by Transanal pull-through in 24%. Only one patient underwent Duhamels pull-through. 5 patients underwent diagnostic laparoscopy with levelling biopsies as transition zone was not clear .These 5 patients underwent a primary pull-through later when the biopsy reports were available. In our study 2 patients had an anastomotic leak. Both the patients underwent resugery with a ileostomy which was closed after six weeks. 2 patients had wound infection settled with daily wound dressings. One patient had prolonged ileus which resolved on conservative treatment. One patient who was operated in first month of life had a convulsion in post-operative period was shifted to paediatric medicine evaluated and diagnosed as intraventricular haemorrhage. Patient was managed conservatively , symptoms resolved and was discharged.

Two patients presened with constipation in the late postoperative period. One among these two patients was later admitted with enterocolitis settled with conservative treatment . The other patient had fecal soiling and recurrent enterocolitis. He was reinvestigated and diagnosed as retained cuff of aganglionic tissue around the pullthrough colon. The cuff was resected and patient is behaving well on follow-up.

DISCUSSION:

Diagnosing the Hirschsprung's disease (HD) early and repair make a better prognosis in these patients .Currently two basic approaches exist to treat such patients namely a multiple staged pull through and a single stage procedure each with its own advantages and disadvantages. Several investigators have reported good results after a one-stage procedure without a stoma for infants with Hirschsprung's disease. However several researchers have reported an increased rate of enterocolitis after primary pull-through. An earlier diagnosis, and single stage repair is an improved strategy, which will bring a better prognosis for HD patients.

During the study period 40 patients were diagnosed as having the Hirschsprung's disease. The excluded patients (37.5%) had massively dilated proximal colon, patients with enterocolitis or late presentation to hospital .This observation suggests a similarity of inclusion criteria as proposed by Singh SJ et [8] Teitelbaum DH [9]. This percentage is far more than published results can be ascribed to patients presenting late in obstruction who mandated a preliminary colostomy.

Male to female ratio was 3.2;1 in patients diagnosed with Hirschsprung's disease and included in our study .Kamal Abd El-Elah Ali [10] also noticed male predominance in their study.
In the present study primary pull through was planned in children age less than 12 months. Mean±SD age of patients at the time of pull-through was 6.9±7.12. 21 (84%) children

underwent pull through in age less than one year. Only 4 patients (16%) underwent pull-through after 12 months due to either a comorbid condition or late presentation.. This indicates that young infants are potential candidates for primary pull- through. Age distribution was similar in a study conducted by Langer JC [11,12] Age distribution in our study confirms the theoretical advantages of early primary pull-through which are less dilatation of proximal bowel, less chances of HAEC, small length of extraperitoneal colon and rectum with less endorectal dissection to be done. The mean weight of patients at the time of pull-through in our study was 8.76±2.67.kgs. No child below 5kgs was taken for surgery.A study by Langer JC[12] has demonstrated that primary pull-through should not be done if wt<4kg.

92% patients in our study presented in neonatal period mostly with a history of delayed passage of meconium. In a study by et al[8] reported neonatal presentation was seen in (90%) cases. Singh SJ Similarly Schäppi MG [13] in his study has advocated that > 90% affected children symptoms start in the neonatal period and less than 1% are diagnosed during adult life.

We demonstrated in our study that features of Hirschsprung's and clear transition zone were seen in only 48% patients on contrast studies. So water soluble contrast study has a low sensitivity. This finding is supported by the observation by Schäppi MG [13] et al who has documented a low sensitivity and specificity of contrast enema in comparison with biopsy. However he has advocated that contrast enema can be used as an additional investigation to assess the length of aganglionic segment before surgery.

We utilised the frozen section technique to delineate the segment involved intraoperatively. 5 patients underwent levelling biopsies and a single stage pull-through later. In a study on frozen section in HD. Rouzrokh M et al [14] has established that the role of FS in the diagnosis of HD cannot be denied, especially in ascertaining the presence of the ganglionic segment during surgery.

Six patients underwent primary transanal primary pull-through. In these patients transition zone was clear on pre-operative contrast study and was confirmed on intra-operative frozen section. Duhamel's pull-through was done in one patient. In 18 patients a diagnostic laparoscopy was done. Among these 18 patients 13 patients had a clear naked – eye transition which was again confirmed by on-table frozen section so we proceeded with a laparoscopic assisted pull-through . 5 patients on diagnostic laparoscopy did not have a clear transition zone on operation table so pull-through was abandoned and biopsies were taken and a primary pull-through was done at a later date. In these five patients later a pull-through was done after histopathology confirmed the diagnosis of Hirschsprung's disease with level of aganglionosis. So majority of patients underwent laparoscopic primary pull-through.

Georgeson KE et al [15] in his study advocated that Laparoscopic-assisted colon pull-through appears to reduce perioperative complications and postoperative recovery time dramatically. The technique is quickly learned and has been performed in multiple centers with consistently good results.

18 patients (72%) in our study had a standard length disease where the superior limit of transition was rectosigmoid. Similar observations were made by Amiel J [16]

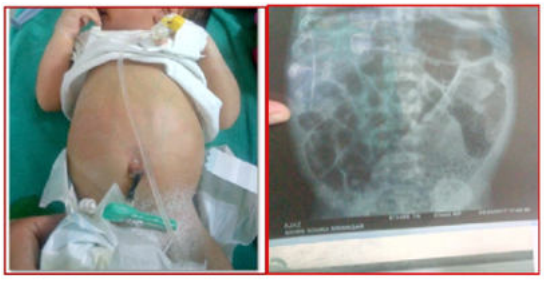
In our study only two patients had an anastomotic leak and were rexplored . A stoma was done and was later closed uneventfully. Only one patient in our study requird a retained cuff which had to be resected. So a total of 3 patients among 25 required second surgery after primary pull-through, i.e, 12%.

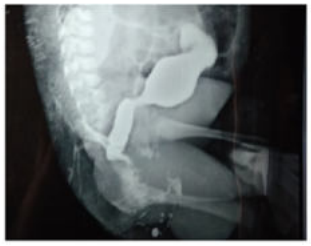
Long term complications were seen in two patients. Both patients initially presented with constipation. Later one patient had an episode of HAEC and underwent a sphincteromyotomy. The other patient having constipation presenting two years after pull-through. Patient was asymptomatic for 2 years after surgery. Patient is improving with bowel management and laxatives. It is well documented in literature that HSD patients can present with HAEC from infancy to adulthood.

K. Ongeti, et al. [17] In his study advocated that single-stage repair may expedite care and reduce the colostomy morbidity justifying primary pull-through in our patients. All the patients in our study are still on a strict follow-up

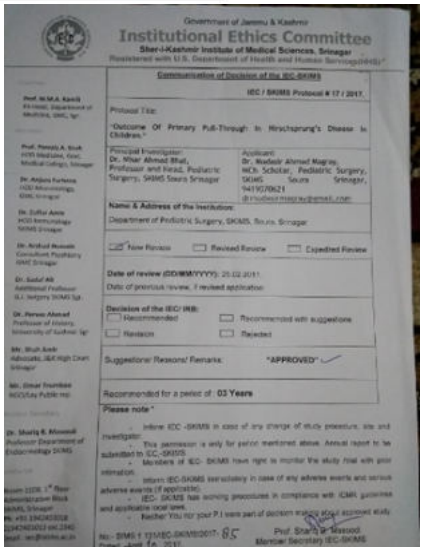
CONCLUSION

From our study we suggest primary pull-through is feasible, reproducible, easy to learn operation with good results but confirmation of our findings and observations needs a larger no of patient cohort.





The above photographs show a neonate presenting to our Department with delayed passage of meconium and plain X-ray and contrast study in the same neonate. Contrast study is clearly showing a transition zone



Government of Jammu & Kashmir
Institutional Ethics Committee
Sheri-i-Kashmir Institute of Medical Sciences, Srinagar
Communication of Decision of the IEC-SKIMS
IEC / SKIMS Protocol # 17 / 2017
Protocol Title:
"Outcome Of Primary Pull-Through In Hirschsprung's Disease In Children."
Principal Investigator: Dr. Nisar Ahmad Bhat, Professor and Head, Pediatric Surgery, SKIMS Soura Srinagar
Applicant: Dr. Rudaiz Ahmad Magray, MCh Scholar, Pediatric Surgery, SKIMS Soura Srinagar, 9419720621
Name & Address of the Institution: Department of Pediatric Surgery, SKIMS, Soura, Srinagar.
New Review / Revised Review / Expedited Review
Date of review (DD/MM/YYYY): 20.02.2017
Date of previous review, if revised application:
Decision of the IEC/ IRB: Recommended / Recommended with suggestions / Revision / Rejected
Suggestions/ Reasons/ Remarks: "APPROVED"
Recommended for a period of: 03 Years
Please note:
Prof. Shazia R. Masood, Member Secretary IEC-SKIMS

DECLARATIONS:

The study was approved by institutional ethical committee of Sheri Kashmir Institute of Medical Sciences, Soura, Srinagar, India. The photo of certificate I have pasted be low.

A proper informed consent was taken from the parents and/or legal guardians of all the patients as they were minors.

The procedures were carried out in accordance with relevant guidelines and regulations.

The study was an observational study on hospital patients and a proper consent was taken before any procedure was being contemplated as per ethics and rules framed by the thical committee.

The study was not funded as the procedure is well established in the department and it was an observational study.

There is no conflict of interest involved.

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