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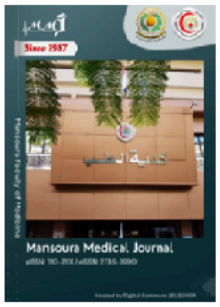

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POSTERIOR SAGITTAL PULL THROUGH PROCEDURE FOR HIRSCHSPRUNG'S DISEASE EVALUATION OF A NEW APPROACH

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POSTERIOR SAGITTAL PULL THROUGH PROCEDURE FOR HIRSCHSPRUNG'S DISEASE : EVALUATION OF A NEW APPROACH

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ABSTRACT

The standard surgical management of infants with Hirschsprung's disease (H.D) traditionally has been one of the pull through procedures developed by Swenson, Duhamel and Soave with a variable rate of complications. So, a new pull through procedure for the treatment of H.D. via posterior sagittal approach without laparotomy or laparoscopy was evaluated in this study. The approach has been performed for 21 patients having H.D. admitted at Pediatric Surgery Unit, Mansoura University Hospital. Theiqr ages ranged from 8 months to 5 years. Patients were followed up postoperatively for a period ranging from 6 months to 18 months. The results were satisfactory with complications occuring only in six cases. Thus, the study reveals that this approach can be performed successfully and safely for definitive treatment of patients having H.D.

INTRODUCTION

Hirschsprung's disease (H.D) is known to be a congenital disorder characterized by absence of the enteric ganglion cells. Recently, gene mutations associated with H.D affecting migration and differentiation of neural crest-derived neuroblasts have been clearly identified (Azuma and Okada, 1997). Abdominoperineal pull through operations as a definitive treatment for H.D have been performed for many years using one of the techniques developed by Swenson, Duhamel and Soave (Langer et al., 1999). Although the advantages

and disadvantages of each technique have been widely discussed, the selection of the surgical procedure appears to be the surgeon's choice (Morikawa et al., 1998). Different modifications of pull through techniques for H.D have been designed and near all of them require laparotomy or laparoscopy with a considerable rate of intraabdominal complications (Mondragon and Salgado, 1998). The posterior sagittal anorectoplasty (PSARP) developed by Pena and Devries in 1982 is widely recognized as the best technique available today for the surgical treatment of anorectal malformations (Martucciello et al., 1999). However, the use of this approach for treatment of H.D that was initially reported by Hedlund, 1997 is still not widely evaluated. Thus, a new pull through procedure for the treatment of H.D via posterior sagittal approach without laparotomy or laparoscopy will be evaluated in this study.

MATERIAL AND METHODS

The present series included 21 patients having congenital magacolon admitted at the Pediatric Surgery Unit, Mansoura University Hospital during the period from 1997 to 1999. Their ages ranged from 8 months to 5 years with a mean 20.6 ± 14.2 months. They included 15 males and 6 females. The preoperative diagnosis of H.D was based upon clinical examination, contrast enema which also indicated the length of the aganglionic segment, anorectal manometry in addition to punch biopsy of the mucosa and submucosa. Table I shows the ages at operation and the approximate length of the aganglionosis in each case by radiology. Three patients had been performed sphinctromyectomy but with no significant clinical improvement with recurrent constipation and episodes of enterocolitis. For all patients, pull through operation through posterior sagittal approach has been performed. The operation was performed with the patient in a prone jack knife position. The skin incision was made from the level of sacrococcygeal junction to approximately 2 cm from the mucocutaneous junction. The coccyx, the levator muscle and the upper portion of the striated muscle complex were divided in the midline. Care was taken to expose the rectal wall properly by opening the fascia of Waldeyer (Fig. I). Two traction sutures were placed on the posterior aspect of the rectum and the loop of the rectum was gradually mobilized by division of vessels and

bands posteriorly and laterally. The placement of a Nelaton catheter or a rubber drain all around the rectum would also facilitate the mobilization (Fig. II). New traction sutures were placed at each 3 to 5 cm of mobilized bowel. The peritoneal reflection was reached and the vas deference was identified and preserved. The dissection was completed higher up till the transition zone was reached where frozen section biopsy specimens were obtained to confirm the proper ganglionic level. If the ganglionic bowel was very wide, the dissection was continued proximally to reach a normal calibre ganglionic colon (Fig. III). At this stage, resection and an oblique coloanal anastomosis was performed manually. The manual anastomosis was constructed in a single layer with interrupted through and through 4/0 vicryl sutures (Fig. IV, V). After completing the anastomosis, construction of the sphincter muscles was carefully performed in the midline with 3/0 or 2/0 vicryl sutures. The wound was closed with a pelvic suction drain (Fig. VI). The length of the resected aganglionic segment ranged from 8 to 22 cm with a mean 12.3 ± 4.2cm. Patients were on total parenteral nutrition (T.P.N) for 5 days postoperalively. Patients were followed up for a period varying from 6 months to 18 months postoperatively by clinical evaluation and anorectal manometry. Anal resting pressure and anal squeeze pressure were recorded.

RESULTS

Posterior sagittal pull through operation was performed for 21 children having H.D. Uneventful clinical and functional results were obtained in 15 patients without early or late postoperative complications. Patients were fully continent without fecal or urinary problems. The anal resting pressure and anal squeeze pressure were significantly lowered postoperatively reaching approximately the normal levels (Fig. VII, VIII).

Complications occurred in six cases of the study (28%) (Table II). Intraoperative iatrogonic urethral injury occured in one male patient and was treated by urethral repair and suprapubic diversion for 2 weeks. Two patients developed anastomotic leakage with subsequent deep abscess and rectocutaneous fistula, for whom right. sided double barrel colostomy was performed in addition to high potency antibiotics and supportive therapy. The fistula closed spontaneously after one month. Two patients developed

extensive wound infection and dehisence till the muscle plane. They were treated with open drainage and debridement in addition to the proper antibiotic regimen based upon culture and sensitivity findings. Wounds were closed later on within 1.5 months by secondary sutures. One patient- with H.D. that was affecting the upper sigmoid region- developed enterocolitis three months after the pull through operation and he was in need for emergent colostomy diversion in addition to the other items of treatment of enterocolitis. Pathological reevaluatinn of the ganglionic level of the colon revealed that there was a residual hypoganglionic segment for which subsequent resection was performed 6 months after the first operation through abdominal route.

Table (I) : Patient ages and approximate length of the aganglionic segment (by radiology).

No	Age	Length of aganglolnc segment
1.	9 months	Lower sigmoid
2.	12 months	Lower sigmoid
3.	1.5 years	Upper sigmoid
4.	3 years	Upper rectum
5.	10 months	Upper sigmoid
6.	3.5 years	Lower rectum (previous myectomy)
7.	2 years	Lower sigmoid
8.	8 months	Upper rectum
9.	9 months	Lower sigmoid
10.	11 months	Mid sigmoid
11.	1 year	Upper rectum
12.	2.5 years	Lower rectum (previous myectomy)
13.	1.5 years	Mid sigmoid
14.	15 months	Upper rectum
15.	14 months	Lower sigmoid
16.	4 years	Lower sigmoid
17.	2 years	Upper rectum
18.	5 years'	Lower rectum (previous myectomy)
19.	1 year	Upper rectum
20.	10 months	Lower sigmoid
21.	11 months	Upper rectum

Lower rectum : 3 : 14.2%.
Upper rectum : 7 : 33.4%.
Lower sigmoid : 7 : 33.4%.
Mid sigmoid : 2 : 9. 5%.
Upper sigmoid : 2 : 9. 5%.s

Table (II) : Complications with posterior sagittal pull through operatior (follow up period from 3 months to 18 months) .

Complication	No
I- Intraoperative complications:	
❒ Iatrogenic urethral injury	1
II- Early complications :	
❒ Wound dehisence	2
❒ Anastomotic leakage	2
III- Late complications :	
❒ Soiling	0
❒ Incontinence	0
❒ Recurrent constipation ± Fecaloma	0
❒ Enterocolitis	1
❒ Urinary problems	0

Total 6 cases (28%) .

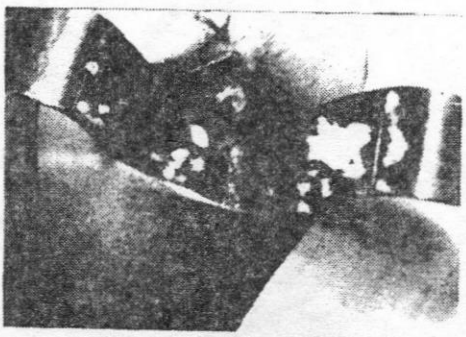

Fig. I : Exposure of the rectum.

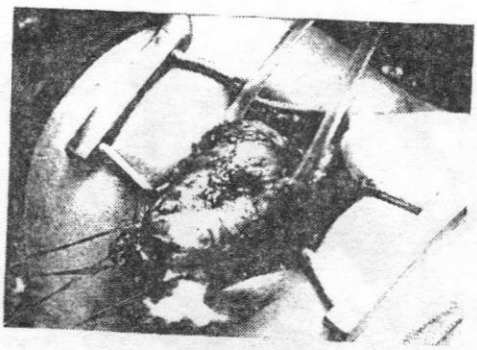

Fig. II : Delivery of the rectum by traction sutures and Nelaton catheter .

Fig. III : Complete mobilization of the rectum and sigmoid till the desired level.

Fig. V : Completing the anastomosis and starting to construct the sphincter .

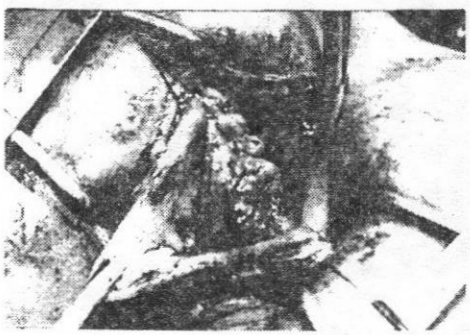

Fig. IV : Fashioning of a coloanal anastomosis .

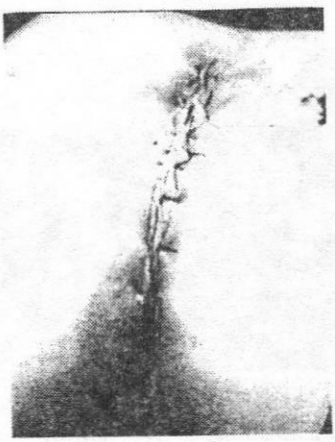

Fig. VI : Closure of the wound with a pelvic suction drain.

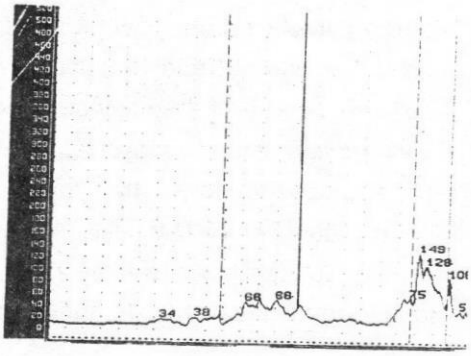

Fig. VII : Preoperative manometry showing the anal resting and squeeze pressures .

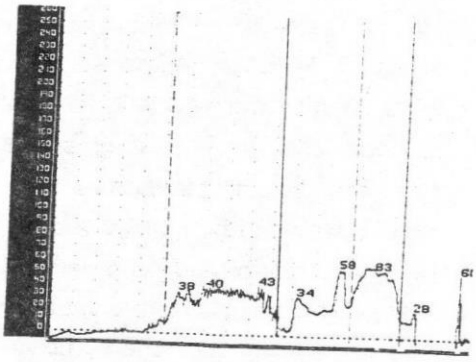

Fig. VIII : Postopreative manometry showing the anal resting and squeeze pressures.

DISCUSSION

The posterior sagittal approach- that is used widely for treatment of anorectal malformations- has been evaluated in this study for the pull through procedure for treatment of H.D. The approach has been performed for 21 patients having H.D and they were followed up postoperatively for periods ranging from 6 months to 18 months. Complications in our series occurred in six cases and fortunately enough, they were treated safely without residual inability. In the case presenting by enterocolitis with a residual hypoganglionic segment, redo operation has been performed through abdominal route for extensive mobilization of the colon and safe vascular ligation. Results in our series are encouraging and agreeing with the results of Hedlund, 1997. However he performed this approach for rectal aganglionosis only but in our series the approach has been performed for H.D. affecting the colon up to the upper sigmoid region . Also, our results are in agreement with the results of Niedzielski, 1999 who stated that this approach provided an excellent exposure of the opreative field allowing to perform the lowest possible re-

section and subsequent anastomosis. Meanwhile this approach is more feasible than the transanal pull through procedure for rectosigmoid HD performed by Mondragon and Salgado, 1998 and Albanese et al., 1999 as they were in need for laparoscopy assistance in their series. Also, it is more simple than the technique developed by Morikawa et al., 1998 who performed laparoscopic-assisted endorectal pull through using prolapsing technique. The complication rate in the present series (28%) was relatively lower than the series of Weber et al., 1999 who reported a complication rate up to 35% with re-operation procedures for H.D via intraabdominal approach. Subsequently another advantage of the posterior sagittal approach is that it solves the problem of patients with failed sphincteromyectomy thus avoiding them the risk of laparotomy. Thus, the present results show that this approach can be used with feasibility and safety for definitive treatment of H.D affecting up to the upper sigmoid zone (the length of the resected aganglionic segment reached 22 cm in some cases). However, the major difficulty of this approach lies in the proper understanding of the anorectal musculature and sphincteric mechanism that allows a good sphincteric reconstruction. Thus we can conclude that the posterior sagittal approach can be used for pull through operation as a definitive treatment of H.D with promising clinical and functional results provided that the sphincteric reconstruction should always remain to be a golden landmark that the surgeon should completely fulfill. However, it is still too early to evaluate the long term results on a wide number of patients.

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تقييم عملية سحب وإستئصال القولون لعلاج مرض تضخم القولون الخلقى عن طريق الفتح السهمى الخلفى

د.طارق بدراوى ، د. خالد قنديل ، د.محمد الغزالى والى

يعتبر علاج الأطفال المصابين بتضخم القولون الخلقى (مرض هيرشسبرنج) بواسطة عملية سحب وإستئصال القولون من العمليات المتعارف عليها وهى تجرى عن طريق إحدى الطرق المسماه بسوينسون ودوهامل أو سواف مع معدل متفاوت فى المضاعفات فى كل طريقة. وقد تم فى هذا البحث تقييم طريقة جديدة لإجراء عملية سحب وإستئصال القولون عن طريق الفتح السهمى الخلفى بدون إجراء فتح للبطن أو منظار للبطن. وقد تم إجراء هذه الدراسة على واحد وعشرون مريضاً يعانون من مرض تضخم القولون الخلقى وكانت تتراوح أعمارهم مابين ثمانية أشهر وخمسة سنوات وقد تم متابعة الحالات بعد العملية لمدة تتراوح مابين ثلاثة شهور إلى ثمانية عشر شهراً وكانت النتيجة مرضية فى خمسة عشر مريضاً مما يتبين أن نسبة المضاعفات قليلة نسبياً. ومن ذلك يتبين أن هذه الطريقة من الممكن أن تستخدم بنجاح وبأمان فى علاج هؤلاء المرضى وبنسبة مضاعفات أقل نسبياً فى العمليات التى تجرى عن طريق فتح البطن .