OUTCOME OF ONE STAGE TRANSANAL ENDORECTAL PULL-THROUGH FOR MANGEMENT OF HIRSCHSPRUNG'S DISEASE

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ABSTRACT

Introduction: over the years, the surgical management of recto-sigmoid Hirschsprung's disease (HD) has evolved radically and at present a single stage transanal pull-through can be done in suitable cases, which obviates the need for multiple surgeries and blood transfusion. The aim of this study was to test the outcome and safety of single stage endorectal pull-through for management of (HD).

Material and methods: A retrospective analysis (between January 2007 to November 2008) was carried out on all cases of Hirschsprung's reporting to pediatric surgery unit ,Zagazig university hospital, that were managed by transanal pull-through as a definitive treatment. All selected patient including neonate had an aganglionic segment confined to the rectosigmoid area, confirmed by preoperative barium enema and postoperative pathology. All children had their operation done without construction of prior colostomy .we relied on the level indicated on a barium enema as well as clinical intra operative assessment since we lacking the facilities of frozen section .

Result: Transanal pull-through was performed in 14 children including 5 neonates. Mean operative time was 100 minutes (range 64 to 135 minutes). And mean hospital stay was 3-5 days. Oral feeding started 24-48 hours postoperatively. No patient required laparotomy because all patients including neonates had an aganglionic segment confined to the rectosigmoid area. Blood loss ranged between 20 to 85 ml with blood replacement required in 2 cases. Since all children were given an epidural caudal block, the requirement of analgesia in these cases was minimal. postoperative complications included perianal excoriation in 11 out of 14 (78%)patients lasting from 3 weeks to 6 months, which included all neonates (n=5). Complete anorectal continence was noted in 10 of 14 (71.3) children in follow up of 2 years, whereas 3 other patients showed a steady improvement in their continence status.

Conclusion: one stage transanal endorectal pull-through for selected cases is easy, bloodless, without visible scar and with short hospital stay. We believe that this procedure can be safely conducted based on the clinical experience which all pediatric surgeons possess since they regularly perform leveling colostomies in Hirschsprung disease.

Key words: single stage, transanal, pull-through procedure, Hirschsprung's disease, frozen section biopsy, perianal excoriations.

INTRODUCTION

Hirschsprung's disease is a fairly common developmental disorder that occurs in newborns as a result of absence of ganglion cells in the distal bowel resulting in functional intestinal obstruction. Traditionally Hirschsprung's disease has been managed by three staged procedures over 4 to 6 month period. This included a defunctioning colostomy and multiple biopsies from the colon, a definitive pull-through and finally a colostomy closure (1).most patient required a blood transfusion at some stage. Over the years, the surgical management of rectosigmoid Hirsrchsprung's disease (HD) has evolved and present, a single stage transanal pull-through can be done in suitable cases, which obviates the need for multiple surgeries and blood transfusion. The procedure leaves no scar, is associated with less postoperative pain and shorter hospital stay. Return of bowel function postoperatively is much earlier and permits initiation of oral feeds. However, it is primary endorectal pull-through in the newborn period was first described by So et al in 1980 (2), with (81%) of the patients were totally continent after 18 year follow up.

MATERIAL AND METHODS

A retrospective analysis was carried out on all cases of Hirschsprung's disease reporting to pediatric surgery unit, Zagazig university hospital that was managed by transanal pull-through as a definitive treatment. There were 14 cases, including 5 neonates. The diagnosis in the neonates was based on history of failure to pass meconium in the first 24 to 48 hours of life, clinical presentation with

abdominal distention and vomiting, plain radiograph which showed absence of gas shadow in the pelvis, barium enema in which there was abnormal retention of contrast in a 24 hour film and confirmed by full thickness rectal biopsy. Further, the level of contrast tapering was taken as an indicator of the level of aganglionosis. All selected patients including neonates had an aganglionic segment confined to the rectosigmoid area, confirmed by preoperative barium enema and postoperative histology. All children had their operations done without construction of prior colostomy . technique used involved eve rsion of the anal verge with silk sutures to expose the pectinate line (fig.1). Submucosal infiltration with adrenaline saline was done and submucosal dissection was performed up to the level of peritoneal reflection leaving a muscular cuff. The bowel is then pulled out through the anal opening (fig2) and level of resection decided based on the appearance and texture of the bowel. The resected edge is then sutured to the pectinate line (fig.3). Following the pull-through, the excised segment was sent for histopathological examination to confirm the diagnosis of Hirschsprung's disease and also the presence of ganglion cells in the pulled through end. Assessment was done on the following parameters, operation time, blood loss and need for replacement, initiation of feeding postoperatively, requirement of analgesia, duration of hospital stay and perianal skin excoriation.

RESULT

Transanal pull-through was performed in 14 children including 5 neonates. There were 3 females and 11 males with age ranging from 21 days to 3 years. Mean operating time was 100 minutes (range 64 to 135 minutes). No patient required laparotomy because all patients, including neonates had an aganglionic segment confined to the rectosigmoid area. Slight difficulty was experienced in entering the submucosal plane in the initial part of our learning curve. Blood loss ranged between 20 to 85 ml, with blood replacement required in 2 cases. Most children were very hungry within 48 hours and feeding was initiated at around 24 to 48 hours. postoperative complications included perianal excoriation in 11 out of 14 (78.5%) patient lasting from 3 weeks to 6 month, which included all neonates (n=5). Other complications included enterocolitis (n=1)(7.1%), they were treated with saline rectal irrigation, no oral feeding and 3rd generation cephalosporins. recurrent constipation(n=1)(7.1%) and rectal prolapse (n=1)(7.1%). Complete anorectal continence was noted in 10 of 14 (71.3%) children, whereas 3 other patients showed a steady improvement in their continence status. No death occurred in the series. The mean hospital stay was 3-5/days.

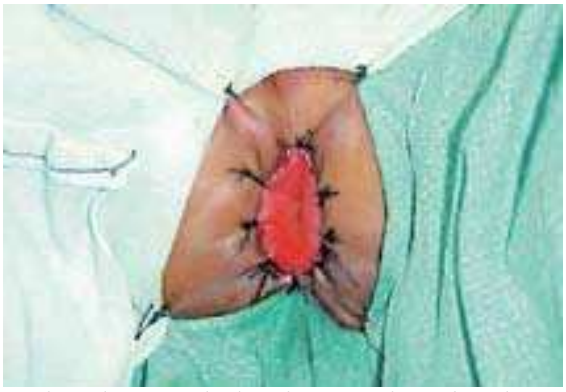

Figure 1. pre-operative photograph showing eversion of anal verge by silk suture.

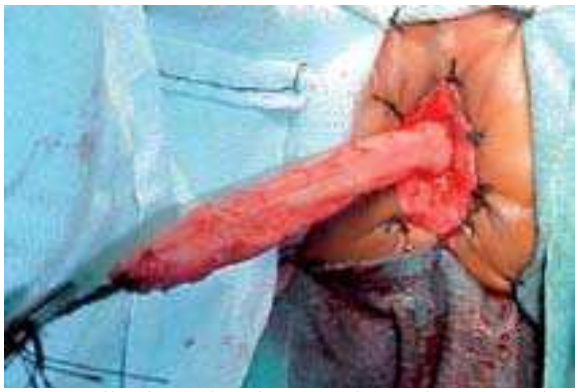

Figure 2. pulled out aganglionic colon from anal opening up to the level of resection.

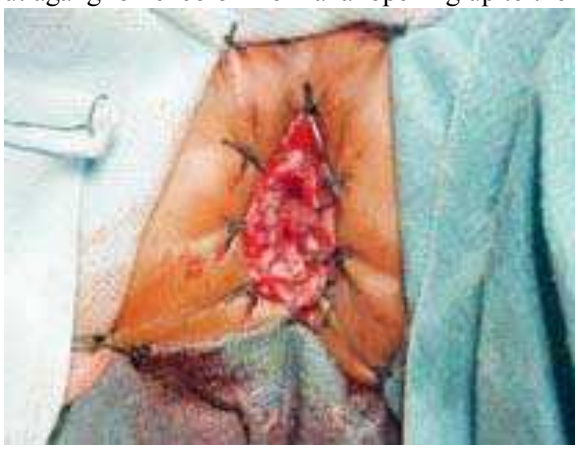

Figure 3 . completed colo anal anastomosis

DISCUSSION

Hirschsprung's disease is a common surgical problem in children. Majority of children have the classical disease in which there is a short aganalionic segment, which may extend up to the rectosigmoid junction. Traditional mangement of these cases by 3- stage procedure resulted in significant morbidity associated with a defunctioning colostomy and repeated surgeries. With introduction of a single stage trans-anal endorectal pull-through procedure, significant reduction in morbidity is possible as it avoids repeated staged surgeries. The use of a transanal endorectal pull-through is feasible in neonates as well as in older children Ellhalaby et al,(3). The mean operating time in our series was 100 minutes (ranging from 64 to 135 minutes). The longer operating time occurred in the early cases due to our learning curve. The mean operative time with Egyptian multicenter study of Ellhalaby et al (3) was (120minutes) and with that of Teeraratkul(4) was (140 minutes). This difference may be caused by younger age in our series. Blood loss varied between 20 and 85 minutes with replacement of blood reguired only in 2 cases (14). Perianal excoriation is a troublesome problem associated with trans-anal endorectal pull-through, which results in significant morbidity and more importantly parental anxiety as to whether the operation has been carried out successfully. In our series, 11 out of 14 (78.5%)children had peri-anal excoriation, which lasted for a period varying

from 3 weeks to 6 months. This occurs most probably due to overstretching of the anal sphincters and subsequent increased stool frequency. While Hadidi has also reported increased stool frequency in all cases lasting up to 6 weeks (5), Rintala reported perianal skin rash in 14 out of 26 (54%) cases (6). EnterocoLitis occur in our series in one patient postoperatively (7.1%), in other series of Gao et al (7) where the incidence of enterocolitis was (6%), this in contrast to the series conducted by Kamal A (8) where the incidence of enterocolitis was high 14.2% this high incidence of Kamal study attributed to immaturity of the immune system in neonates. In our series, laparotomy was not needed in all cases, due to selection of cases preoperative with short aganglionic segment, in Kamal A series (8) laparotomy was needed in 4.6%of cases due to long segment disease. This in reverse to Egyptian multicenter study (3) where the conversion rate was 2.7%due to tear of the mesenteric vessels and difficult mucosectomy. This difference may be attributed to easier mucosectomy and control of mesenteric vessels in neonatal period. Most authors have done the trans-anal pull-through aided by knowledge of the level of aganglionosis on the basis of a frozen section biopsy (4, 5). In our series, since we did not have the facility of a frozen section biopsy, we relied on the level indicated on a barium enema as well as clinical intra-operative assessment. The intra-operative assessment consisted of a visual and palpatory examination of the mobilized bowel. The visual examination assisted us by indicating the level of the transition zone and the proximal dilated bowel. Palpation of the mobilized bowel reveals a thicker bowel wall in the transition zone as compared to a thin walled, soft and compliant proximal bowel. To be on the safer side, we resected an extra 5 to 7 cm length of dilated proximal bowel than we would have done with tissue diagnosis. Histopathological examination of ganglion cells in the pulled through bowel in all cases justified our clinical assessment in the management of rectosigmoid Hirschsprung's disease by trans-anal endorectal pull-through. Zhang et al also performed the single stage trans-anal pull-through based on preoperative barium enema and postoperative histopathological confirmation (10). While many would object to the clinical assessment of the level of aganglionosis as compared to frozen section, our belief in the clinical assessment is strengthened by the fact that most pediatric surgeons used to perform a leveling colostomy at the level of perceived aganglionosis / transition zone. This colostomy was over the years also based on a visual and tactile assessment. The only difference was that the level of aganglionosis was subsequently confirmed histologically prior to the definitive pull-through. However, despite not having the facility of frozen section, we decided to use our experience and clinical acumen to offer a better surgical option with significantly reduced morbidity to these children. Histological confirmation vindicated our belief in ourselves.

CONCLUSION

Single stage trans-anal endorectal pull-through procedure for the management of rectosigmoid Hirschsprung's disease is now a well established and preferred approach. Parental satisfaction is immense due to the lack of scars on the abdomen. We believe that this procedure can be safely conducted based on the clinical expertise and experience which all pediatric surgeons possess since they regularly perform leveling colostomies in Hirschsprung's disease. This can be performed even in hospitals that don't have facilities of frozen section.

REFERENCES

1- Harjai MM. HIrschsprung's disease: Revisited. J postgrad Med; 46:52-4, 2000.

2- So HB,Schwartz DL, becker JM,et al. endorectal pull-through without preliminary colostomy in neonates with Hirschsprung's disease . J PEDIATR SURG 15:470-471, 1980.

3- Elhallaby EA,Hashish A, Elbarbary MM,et al. transanal one-stage endorectal pull-through for Hirschsprung's disease : A multicenter study. J pediatr surg 39:345-351,2004.

4- Teeraratkul S, transanal one stage endorectal pull-through for Hirschsprung's disease in infants and children. J pediatric surg 38:184-187, 2003.

5- Hadidi A. trananal endorectal pull-through for Hirschsprung's disease: experience with 68 patients. J pediatr surg 38(9): 1337-1340,2003.

6- Rintala RJ, Wester T. transanal endorectal pull through with short muscular cuff in the treatment ofHirschsprung's disease.preliminary study with 37 patients. Cir pediatr 16:161-165, 2003.

7- Gao Y,Zhang X,Xu Q et al. primary transanal rectosigmoidectomy forHirschsprung's disease: preliminary results in the initial 33 cases. J pediatr surg; 36:1816-1819, 2001.

8- Kamal abd Elah ALI. Trananal endorectal pull-through for Hirschsprung's disease during the first month of life. Annals of pediatric surgery, vol.6, No 2, pp 81-88,April 2010.

9- Langer JC, Durrant AC, Torr luis de la, T eitelbaum DH,Minkes RK,CatyMG,Wildhaber BE,Ortega, Hirose S, Albanse CT. one –stage transanal soave pull-through for Hirschsprung's disease. A multicenter experience with 4 children. Ann sur 569-83; 2003.

10- Zhang SC, Bai YZ, Wang WL. Clinical outcome in children after trananal one stage endorectal pull-through operation for Hirschsprung's disease. J pediatr surg; 40:1307-11.2005.

نتائج عملية علاج مرض هيرشسبرنج عن طريق سحب القولون واستئصاله من فتحة الشرج فى مرحلة واحدة

المقدمة :-

مرض هيرشسبرنج من العيوب الخلقية فى الأطفال نتيجة عدم وجود العقد العصبية فى نهاية القولون والشرج مما ينتج عنه تضخم شديد فى القولون وإمساك عند الأطفال . فى الماضى كانت تتم عملية علاج مرض هيرشسبرنج عن طريق ثلاثة مراحل .

المرحلة الأولى :- عمل فتحة فى القولون للاخراج من خلال البطن .

المرحلة الثانية :- سحب القولون واستئصاله من فتحة الشرج .

المرحلة الثالثة :- هى قفل فتحة القولون فى البطن .

ولكن حديثا يتم علاج مرض هيرشسبرنج فى مرحلة واحدة عن طريق سحب واستئصال القولون من فتحة الشرج . وكان أول من أجرى هذه العملية هو العالم (سو واخرين) عام 1980 وكانت النتائج جيدة .

المرضى والأدوات :-

هذا البحث تم بأثر رجعى على حالات مرض الهيرشسبرنج التى تم إجرائها فى وحدة جراحة الأطفال بجامعة الزقازيق . وتم علاجها بعملية سحب واستئصال القولون من فتحة الشرج فى مرحلة واحدة فى الفترة من يناير 2007 الى نوفمبر 2008 . ويشمل هذا البحث 14 حالة منهم 5 أطفال حديثى الولادة .

تم تشخيص الحالات بواسطة:-

1- التاريخ المرضى وتأخر خروج الميكونيم أكثر من 48 ساعة بعد الولادة .
2- انتفاخ شديد بالبطن مع قيء .
3- أشعة بالصبغة على القولون .
4- خزعة من المستقيم و تحليلها باثولوجى للتأكد من عدم وجود العقد العصبية فى القولون

الطريقة التى تم استخدامها فى كل الحالات هو سحب القولون واستئصاله عن طريق فتحة الشرج فى مرحلة واحدة وإرسال الخزعة وتحليلها باثولوجى للتأكد من التشخيص وكذلك لمعرفة المستوى الذى تم عنده استئصال القولون.

النتيجة:-

لقد تمت هذه الدراسة على 14 مريض منهم 5 حديثى الولادة 3 إناث و11 من الذكور

متوسط الوقت المستغرق فى العملية حوالى 100 دقيقة (من 64 الى 135 دقيقة)

لم يتم اللجوء الى فتح البطن فى كل الحالات اثناء العملية.

وجدت بعض الصعوبات فى سحب القولون من فتحة الشرج خاصة فى الحالات الأولى من التدريب .

متوسط الدم المفقود أثناء العملية من 20 ملم الى 25 ملم وقد تم نقل الدم لحالتين فقط اثناء العملية .

تم إعطاء سوائل بالفم لجيع الحالات فى الفترة من 24 الى 48 ساعة بعد العملية .

مضاعفات ما بعد العملية :-

1- التهابات فى فتحة الشرج فى 11 حالة و استمرت من 3 اسابيع الى 6 شهور.

2-التهاب تسممى فى القولون فى حالة واحدة وقد تم علاجها بواسطة المضادات الحيوية والمحاليل وعمل غسيل للقولون بالحقن الشرجية .

3-سقوط فى الشرج فى حالة واحدة .

4-استعادة التحكم فى الخروج تم فى 10 حالات وتحسن تدريجى فى باقى الحالات

5-لاتوجد حالات وفيات فى الدراسة .

الإستنتاج:-

إن عملية سحب واستئصال القولون عن طريق فتحة الشرج فى مرحلة واحدة لعلاج مرض الهيرشسبرنج أصبحت عملية سهلة وأمنة ويمكن كذلك اجراؤها فى الاماكن التى لايتوفر فيها استعدادات لفحص الباثولوجى اثناء العملية .