Original Article

Role of Ano Rectal Myomectomy in Children with Chronic Refractory Constipation

Rajeev Gurunath Redkar, Vinod Raj, Anant Bangar, Varun Hathiramani[1], Swathi Chigicherla, Shruti Tewari

Lilavati Hospital and Research Centre, Mumbai, Maharashtra, India, [1]Department of Pediatric Surgery, Kings College Hospital, NHS Foundation Trust, London, UK

Abstract

Background: Chronic refractory constipation (CRC) is an uncommon type of constipation. These children have persistent symptoms even after treatment with high dose laxatives, which may cause abdominal distension, vomiting, cramping and bloating. We conducted this study to assess the diagnostic and therapeutic role of anorectal myomectomy in children with CRC. **Materials and Methods:** This study includes 107 patients who fit the criteria of CRC. Complete bowel preparation with polyethylene glycol solution, enemas and antibiotics was carried out before surgery in all patients. The anorectal myomectomy was carried out under general anaesthesia with the patient in the high lithotomy position. The patients were followed up from 6 months to 13 years postoperatively. The success of myomectomy was based on the daily and complete passage of stools without the need for medication or enemas. **Results:** A total of 99 patients were included in the study, of which, 86 (86.86%) patients showed a good response to anorectal myomectomy. Of these, 32 patients had normal histology, 14 had histology suggestive of Hirschsprung's disease, 8 had hypoganglionosis, 10 had ultra-short segment Hirschsprung's disease and 22 had hypertrophic nerves with immature ganglia. Poor response was seen in 13 (13.13%) patients of whom 5 had normal histology, 5 had Hirschsprung's disease, 2 had hypoganglionosis and 1 had ultra-short segment Hirschsprung's disease. **Conclusion:** Anorectal myomectomy is an effective and technically simple procedure in selected patients with CRC for both diagnostic and therapeutic purposes.

Keywords: Anorectal myomectomy, chronic refractory constipation, Hirschsprung's disease

Introduction

Constipation is a common problem in the paediatric population, which can usually be treated with dietary modifications and laxative therapy. However, some children have refractory symptoms, and when subjected to high-dose laxative therapy can develop abdominal distension, vomiting, cramping and bloating.[1] Chronic refractory constipation (CRC) is defined as children who are unable to pass stools in spite of being on maximum laxative therapy and require daily rectal stimulation in the form of enemas or suppositories to pass stools for more than 6 months. There have been many reported operative strategies for the management of severe constipation. This article studies the role of anorectal myomectomy in the management of CRC.

Children with CRC tend to develop dilatation of the colon due to retention of stools which can progress to megacolon like a picture on the contrast enema. In patients with this refractory course of constipation, a rectal biopsy becomes necessary to rule out neuronal dysplasia and Hirschsprung's disease and other associated conditions. We propose performing an anorectal myomectomy at this same sitting which can be therapeutic along with the diagnostic procedure of rectal biopsy.

Aim

The aim of this study is to assess the diagnostic role and therapeutic value of anorectal myomectomy in children with CRC.

Materials and Methods

Inclusion and exclusion criteria

This study included 107 patients who fit the criteria of CRC. This included:

Address for correspondence: Dr. Rajeev Gurunath Redkar, Lilavati Hospital and Research Centre, A-791, Bandra Reclamation, Bandra West, Mumbai - 400 050, Maharashtra, India.
E-mail: rajeev.redkar@mail.com

Access this article online

Quick Response Code:

Website: www.afrjpaedsurg.org

DOI: 10.4103/ajps.AJPS_99_17

How to cite this article: Redkar RG, Raj V, Bangar A, Hathiramani V, Chigicherla S, Tewari S. Role of ano rectal myomectomy in children with chronic refractory constipation. Afr J Paediatr Surg 2018;15:31-5.

1. Inability to pass stools daily in spite of the maximum dose of laxative therapy
2. Need of daily rectal stimulation in the form of enemas or suppositories to pass stools
3. The presence of the above two conditions for more than 6 months
4. A contrast enema showing megarectum with a zone of coning or an abrupt narrowing.

Children with the signs of intestinal obstruction, diagnosed cases of Hirschsprung's disease and those with known metabolic or endocrine conditions or who were on medications which are known to cause constipation were excluded from the study. All children had a contrast enema before the procedure showing dilated rectum and sigmoid colon.

Surgical procedure

The patients were admitted 3 days before the surgery. During this preoperative period, all patients underwent complete bowel preparation with polyethylene glycol solution (25 ml/kg over 4 h repeated twice a day), simple enemas with normal saline and antibiotics (oral metronidazole). The anorectal myomectomy was carried out under general anaesthesia with the patient in high lithotomy position. The Lone Star Retractor System® [Cooper Surgical, Inc. Nory, GE, Figure 1] was used and a standard posterior anorectal myomectomy was done starting 2 cm above the dentate line via smily incision. An adequate strip of muscle (minimum length of 2 cm) was removed in all patients [Figure 2]. The distal and proximal ends of the muscle strip were marked with catgut and polyglactin stitches respectively to aid in histopathological co-relation. The mucosal defect was closed with interrupted polyglactin stitches of appropriate size, and a lubricated, antibiotic-impregnated rectal pack was placed for 24 h. The patient was discharged once the child passed stools after pack removal.

Follow-up

The patients were followed up from 6 months to 13 years postoperatively. The follow-up evaluations were based on clinical evaluation including postoperative bowel habits, the need for medication, relief of symptoms and dietary modifications.

Results [Table 1]

The mean age of the studied population was 4 years 1 month (7 months to 9 years). There were 71 boys and 36 girls and out of the 107 patients 8 were lost to follow-up.

Of the 99 followed up patients, 37 (37.37%) patients had normal histology, 11 (11.11%) had ultrashort segment Hirschsprung's disease, 10 (10.10%) had hypoganglionosis, 22 (22.22%) had hypertrophic nerves and immature ganglia and 19 (19.19%) had Hirschsprung's disease.

The success of myomectomy was based on the daily and complete passage of stools without the need for medication or enemas. Therefore, out of the 99 patients, 86 (86.86%) showed a good response to anorectal myomectomy. Of these, 32 had normal histology, 14 had histology suggestive of Hirschsprung's disease, 8 had hypoganglionosis, 10 had ultrashort segment Hirschsprung's disease and 22 had hypertrophic nerves with immature ganglia.

Poor response was seen in 13 (13.13%) patients of whom 5 had normal histology, 5 had Hirschsprung's disease, 2 had hypoganglionosis, and 1 had ultrashort segment Hirschsprung's disease. The five patients in whom a definitive diagnosis of Hirschsprung's disease was made were found to have aganglionosis up to the rectosigmoid junction and were treated appropriately with pull-through surgery and are doing well postoperatively with normal bowel movements.

In the 37 patients with normal histology, 32 (86.48%) did not require any medication or enemas postoperatively, whereas five patients (13.51%) still require oral laxatives to pass stools daily.

The group with 19 patients which showed Hirschsprung's histology, 14 patients (73.68%) are doing well without enemas or medications. The remaining five patients (26.31%) required further pull-through surgery and were found to have longer segments of aganglionosis.

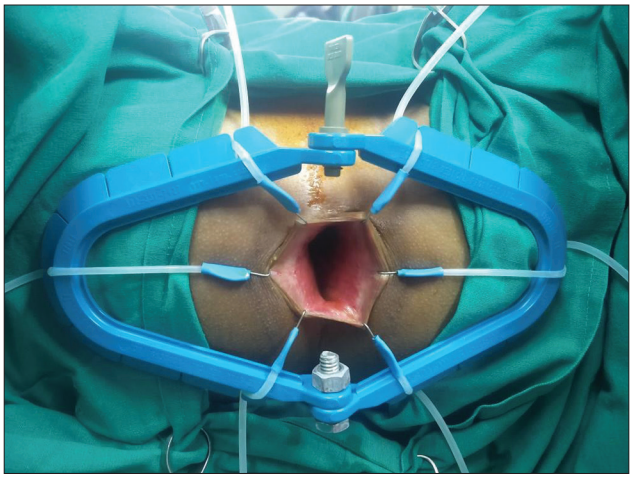

Figure 1: The Lone Star Retractor System

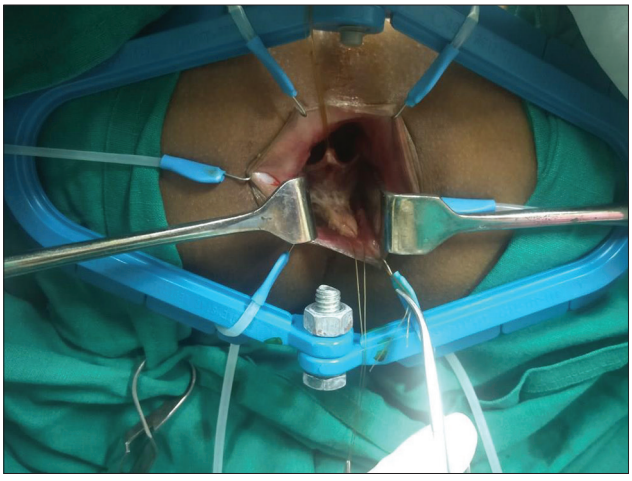

Figure 2: The posterior muscle strip being excised

Table 1: Histology versus outcomes

Histology	*n* (%)	Improved postmyomectomy	Still requiring medications or enemas postmyomectomy	Further intervention
Normal	37 (37.37)	32 (86.48)	5 (13.51)	On medication
Hirschsprung's disease	19 (19.19)	14 (73.68)	5 (26.31)	Improved after pull through surgery
Ultra short segment Hirschsprung's disease	11 (11.11)	10 (90.90)	1 (9.09)	On medication
Hypoganglionosis	10 (10.10)	8 (80)	2 (20)	On medication
Hypertrophic nerves and immature ganglia	22 (22.22)	22 (100)	0	-

Table 2: Histology report table

Histology	*n*	Distal end	Proximal end
Normal	17	Normal (17)	Normal (17)
Hirschsprung's disease	8	No ganglion cells (8)	No ganglion cells (8)
Ultra short segment Hirschsprung's disease	7	No ganglion cells (7)	Ganglion cells present (7)
Hypoganglionosis	4	Hypoganglionosis (4)	Normal ganglions (3) Hypoganglionosis (1)
Hypertrophic nerves and immature ganglia	10	Immature ganglia and hypertrophic nerves (10)	Immature ganglia and hypertrophic nerves (4) Normal ganglia and hypertrophic nerves (3) Normal ganglia and nerves (3)

Among the patients who showed hypertrophic nerves ($n = 22$), hypoganglionosis ($n = 10$) and ultrashort segments of Hirschsprung's disease ($n = 11$) on their histopathology, all but three (one with ultrashort segment Hirschsprung's disease and two with hypoganglionosis) improved in their symptomatology not requiring enemas or medication for passing stools daily.

We were able to record the histopathological analysis of the distal and proximal ends of the myomectomy specimen in 46 patients and the results are tabulated in Table 2.

Discussion

Chronic constipation is the most common presentation of childhood constipation. It is characterised by persistent difficulty in defecation and most commonly is caused by painful bowel movements, resulting in withholding of faeces by the child, without objective evidence of an organic disorder. Stool-withholding behaviour can lead to prolonged faecal stasis in the colon, with reabsorption of fluids and decrease in the size and hard stools,[2] resulting in increased rectal compliance and decreased rectal sensitivity. Such patients get into a vicious cycle of hard stool, painful defection and retention of stool which further aggravates their problems. These patients may also have abdominal distension, pain or discomfort and this condition is an important factor for psychosocial disorders among children and their families.

The management of chronic constipation depends on two critical factors for achieving good results: the child's adherence to conventional treatment (adequate dietary fibre intake and dietary modifications, behavioural modifications, drug therapy and faecal disimpaction), and parents' understanding of the importance of incorporating these modifications. Nevertheless, long-term follow-up studies have shown that approximately 50% of children require treatment for long periods of time,[3] showing no significant improvement with the use of conventional therapeutic measures.

In most children with chronic constipation, the aetiology is unknown. In a reported series of children with idiopathic constipation, Keshtgar *et al*.[4] which included 144 children a variety of underlying abnormalities were documented. Therefore in such children, it is necessary to establish a differential diagnosis between functional and organic constipation, such as Hirschsprung's disease, intestinal dysganglionosis, other systemic and neurological disorders or malformations.[5]

In spite of normal histology in 37 of our patients, four of them still required oral laxatives in the postoperative period. We attribute this to an undiagnosed form parasympathetic over activity and subtle variants of intestinal dysmotility.

Anorectal myomectomy can be a useful technique to reach a histological diagnosis and can also be used as a therapeutic intervention. The usefulness of diagnostic myomectomy to provide muscle biopsy specimen is well understood.

The concept of correcting an area of anatomical or functional bowel obstruction by myomectomy is not new. Myomectomy involves excising a strip of muscle along one wall which breaks the circumferential action of the muscle group. It is known that the internal sphincter does not relax in response to rectal distension in patients with Hirschsprung's disease. This hypertonicity and hypercontractibility extend into the aganglionic segment. This abnormal response of the aganglionic segment and the internal sphincter contribute to

an element of obstruction in Hirschsprung's disease which is similar to the spasm in the cardio-oesophageal region in achalasia and pylorus in pyloric stenosis.

In the early part of the last century, the similar principle of myomectomy was applied to the oesophagus (Heller's cardiomyotomy for achalasia cardia), pylorus (Ramstedt's pyloromyotomy for hypertrophic pyloric stenosis) and small and large intestine (Martin's total colonic myotomy for long-segment Hirschsprung's disease) by various researchers. Its role in the anorectal region was proposed and described with successful results by Hurst,[6] Bentley[7] and Thomas.[8] Bentley *et al*. described posterior excisional anorectal myomectomy in the management of chronic faecal accumulation.[9]

Several techniques have been described for anorectal myomectomy[10-13] but all depend on excision of a longitudinal strip of muscle for a variable distance with variable approaches. This study was designed to evaluate the role of posterior myomectomy in children with refractory constipation for the diagnosis and as a modality of treatment.

In Holschneider's[14] review of the literature, 144 (76%) of 189 patients with refractory constipation had excellent results after myomectomy. De Caluwé *et al*.[15] in their study have reported 2–6 years follow-up of 15 patients after internal sphincter myomectomy with clinical evaluation and manometric studies. Seven of these 15 patients had regular bowel motions without any medication and six needed small doses of laxatives.

In a similar study, Doodnath and Puri[16] have reported 87.5% success rate with follow-up based on clinical evaluation. Redkar *et al*.[17] in 2012 published their preliminary results wherein 92.86% of the patients with CRC were benefited following anorectal myomectomy. Ahmadi *et al*.[18] classified patients according to presence of ganglionic cells into 3 groups; Group A (normal ganglion cells in proximal and distal ends of muscle stripe), Group B (no ganglion cells in both proximal and distal ends) and Group C (normal ganglion cells only in the proximal end). They reported no meaningful difference in the results of surgery between Groups A, B and C, and they got to benefit from dilatation and anorectal myomectomy.

Anal stricture and incontinence have been reported as complications of myomectomy; however, no such complications were seen in any of our patients. Further intervention with pull-through procedures was not affected by this procedure, and no technical difficulty was observed in the five patients with aganglionosis up to the rectosigmoid level, who underwent the pull-through surgery for their definitive management.

Conclusion

In our study, we found that 37.37% of children with normal histology suffered from CRC.

The biopsy and myomectomy procedure proved diagnostic in 100% and therapeutic in 86.48% of the patients with normal histology. The rest 13.52% of patients with normal histology required some form of constipation management in the form of enemas and medication in spite of anorectal myomectomy.

Abnormalities of bowel innervation were diagnosed in 62.62% of patients, and 87% of these patients improved with the myomectomy procedure. Fourteen of the 19 patients (73.68%) with Hirschsprung's disease on histology were treated by anorectal myomectomy alone while the rest 26.32% required a formal pull-through operation.

Anorectal myomectomy is an effective and technically simple procedure in selected patients with CRC, for both diagnostic and therapeutic purpose. This gives a histopathological confirmation in all patients and helps therapeutically in 86.86% of patients with CRC. This is a gold standard for the diagnosis and could benefit the patients with CRC therapeutically as well.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Levitt MA, Martin CA, Falcone RA Jr., Peña A. Transanal rectosigmoid resection for severe intractable idiopathic constipation. J Pediatr Surg 2009;44:1285-90.
2. Constipation Guideline Committee of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Evaluation and treatment of constipation in infants and children: Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. J Pediatr Gastroenterol Nutr 2006;43:e1-13.
3. Staiano A, Andreotti MR, Greco L, Basile P, Auricchio S. Long-term follow-up of children with chronic idiopathic constipation. Dig Dis Sci 1994;39:561-4.
4. Keshtgar AS, Ward HC, Clayden GS, Sanei A. Thickening of the internal anal sphincter in idiopathic constipation in children. Pediatr Surg Int 2004;20:817-23.
5. Bustorff-Silva JM, Costa-Pinto EA, Fukushima E. Role of anorectal manometry in the differential diagnosis of chronic constipation in children. J Pediatr (Rio J) 2000;76:227-32.
6. Hurst AF. An address on the sphincters of the alimentary canal and their clinical significance: Delivered before the Manchester Medical Society (October 8th, 1924). Br Med J 1925;1:145-51.
7. Bentley JF. Some new observations on magacolon in infancy and childhood with special reference to the management of megasigmoid and megarectum. Dis Colon Rectum 1964;7:462-70.
8. Thomas CG Jr. Posterior sphincterotomy in Hirschsprung's disease. Surg Gynecol Obstet 1967;124:365-6.
9. Bentley JF, Nixon HH, Ehrenpreis TH, Spencer B, Lister J, Dhuamel B, *et al*. Seminar on Pseudo-Hirschsprung's disease and related disorders. Arch Dis Child 1966;41:143-54.
10. Ibay RS. Anorectal myectomy for short aganglionic megaeolom. Indian J Pediat 1983;50:61-4.
11. Alexander JL, Aston SJ. A technique for posterior myectomy and internal sphincterotomy in short-segment Hirschsprung's disease. J Pediatr Surg 1974;9:169-70.
12. Scobie WG, Mackinlay GA. Anorectal myectomy in treatment of ultrashort segment Hirschsprung's disease. Report of 26 cases. Arch Dis Child 1977;52:713-5.
13. Shehata SM, El-Banna IA, Gaber AA, El-Samongy AM, Attia MA.

Long-term evaluation of modified lateral anorectal myomectomy for low-segment hirschsprung disease. Arch Surg 1998;133:269-71.
14. Holschneider AM. Anal sphincter achalasia. In: Hirschsprung's Disease. New York: Hippocrates; 1982. p. 203-18.
15. De Caluwé D, Yoneda A, Akl U, Puri P. Internal anal sphincter achalasia: Outcome after internal sphincter myectomy. J Pediatr Surg 2001;36:736-8.
16. Doodnath R, Puri P. Long-term outcome of internal sphincter myectomy in patients with internal anal sphincter achalasia. Pediatr Surg Int 2009;25:869-71.
17. Redkar RG, Mishra PK, Thampi C, Mishra S. Role of rectal myomectomy in refractory chronic constipation. Afr J Paediatr Surg 2012;9:202-5.
18. Ahmadi J, Ashjaei B, Kalantari M, Nahvi H, Ebrahimsoltani A, Nejat F, *et al*. Study of outcome and complications of anorectal myectomy in children with ultrashort segment Hirschsprung's disease. Acta Med Iran 2006;44:259-62.