

# Internal Sphincter Myectomy for Adult Hirschsprung's Disease: A Single Institute Experience

Jirawat Pattana-arun MD\*, Tawornrath Ruanroadroun MD\*,  
Kasaya Tantiphalachiva MD\*, Chucheeep Sahakitrungruang MD\*,  
Puttarat Attithansakul MD\*, Arun Rojanasakul MD\*

\* Division of Colon and Rectal, Department of Surgery, Chulalongkorn University,  
King Chulalongkorn Memorial Hospital, Bangkok, Thailand

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**Objective:** Adult Hirschsprung's disease is a rare disease and frequently misdiagnosed as the long-standing refractory constipation. Almost all cases have short or ultra-short aganglionic segment of distal rectum. The clinical features are different from those in childhood when the diseased segment is long. Amongst the few successful operations that have been used to treat this condition, internal sphincter myectomy has been proposed as a simple and low morbidity procedure, but only a few literatures reported the results. The present study aimed to evaluate the outcomes of anorectal myectomy in adult Hirschsprung's disease.

**Material and Method:** All medical records of adult Hirschsprung's disease between January 1, 1997 and April 30, 2008 were retrospectively reviewed. The histological criteria for diagnosis were increase in the number of cholinergic nerve fibers in the lamina propria, muscularis mucosae, and submucosa, and the absence of ganglia in the submucosa. All cases underwent internal sphincter myectomy as the first operation. Post-operative complications, number of defecation per week, and the need for a second operation were studied.

**Results:** Seven patients met the criteria. All patients had the long history of constipation. Anorectal myectomy was performed as the first operation in all cases. Four patients (57%) had good results, without complication and no further operation was needed up to the last follow-up (26-86 months). Two cases underwent subtotal and total colectomy after myectomy to achieve good results eventually. Only one patient had a poor result after Left colectomy and Total proctocolectomy with ileal pouch anal anastomosis.

**Conclusion:** Internal sphincter myectomy, the simple and complication-free procedure, provides the satisfactory outcomes for adult Hirschsprung's disease. This technique should be the first operation for this condition.

**Keywords:** Hirschsprung's disease, Myectomy

*J Med Assoc Thai* 2010; 93 (8): 911-5

Full text. e-Journal: <http://www.mat.or.th/journal>

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Hirschsprung's disease is a congenital abnormality of ganglion cells in submucosal and myenteric plexuses of the distal intestine. This condition results in the inability of the involved colon and internal anal sphincter to relax. It is commonly seen in newborns and infants but rarely reported in adults<sup>(1-3)</sup>. The term "adult Hirschsprung's disease" has been arbitrarily applied to a case in which the patient is more than ten years of age when the diagnosis is established<sup>(4-6)</sup>.

The diagnosis in adult patients is more difficult than in newborn or infancy, because the short or ultra-short segment of distal rectum is involved, leading to latent and milder symptoms. Adolescents and adults who suffer from intractable chronic constipation may be caused by this disease. Other symptoms include various degrees of abdominal distension and pain, palpable fecal masses, fecal impaction, using of cathartics and/or enemas to achieve bowel movements<sup>(5-8)</sup>. The diagnosis of Hirschsprung's disease is supported by finding the transitional zone in the barium enema study and by absence of the rectoanal inhibitory reflex during anorectal manometry, and is confirmed by histopathological findings of aganglionosis.

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**Correspondence to:**

Pattana-arun J, Division of Colon and Rectal, Department of Surgery, Chulalongkorn University, King Chulalongkorn Memorial Hospital, Bangkok 10330, Thailand.  
Phone: 0-2256-4117, Fax: 0-2256-4194  
E-mail: [jpattanaarun@gmail.com](mailto:jpattanaarun@gmail.com)

The surgical procedures developed to treat the disease in children have been applied to adults. Each procedure has protagonists and antagonists. The Duhamel procedure appeared to be associated with a lower rate of major postoperative complications than other procedures, and the rate of good long-term results was higher<sup>(4,6,7,9)</sup>. Lynn reported that the results of rectal myectomy, which is a minor procedure for the treatment of short-segment Hirschsprung's disease, were good in 45.7% of patients<sup>(10)</sup>. However, long-term follow-up from several studies revealed unsatisfactory results<sup>(3,5,6,11-13)</sup>. In King Chulalongkorn Memorial Hospital, anorectal myectomy is the most commonly performed procedure for adult Hirschsprung's disease either alone or accompanied with other operations. In the present series of adult Hirschsprung's disease patients, the treatment and results of all cases were described herein.

#### **Material and Method**

The authors reviewed all the medical records of patients who were histologically diagnosed of Hirschsprung's disease and were operated at age more than fifteen years old between January 1, 1997 and April 30, 2008. The histologic criteria for the diagnosis of Hirschsprung's disease were an increase in the number of cholinergic nerve fibers in the lamina propria, muscularis mucosae, and submucosa, and the absence of ganglia in the submucosa<sup>(14)</sup>. The prevalent signs and symptoms, the results of investigations such as barium enema, and anorectal manometry, and the operative procedures including their results and complications were noted.

The technique of anorectal myectomy was modified from posterior midline incision reported by Lynn<sup>(10)</sup>. The mucosal incision started from the dentate line to the upper border of sphincter complex, at 3 or 9 o'clock, and then the internal sphincter was separated from the submucosa and the external sphincter. A strip of internal sphincter muscle 0.5 to 1.0 cm in width was cranially removed for pathological confirmation in the same distances. Then the incision was re-approximated with 4-0 absorbable suture, in continuous fashion.

The authors classified long-term results as: good, consisting of complete fecal continence with rare use of laxatives or enemas; fair consisting of occasional fecal incontinence or routine use of laxatives or enemas; and poor consisting of continued reliance on mechanical disimpaction with no improvement over the preoperative period. Surgical complications were classified as major, consisting of pelvic

sepsis, pelvic abscess, rectal perforation, and stricture requiring reoperation, and minor consisting of wound infection, stricture requiring dilatation, temporary urinary retention, and rectal hematoma<sup>(1)</sup>.

#### **Results**

There were seven patients (6 females) diagnosed as adult Hirschsprung's disease with pathological confirmation. The median age at the time of diagnosis was 24 (18-66) years old. All patients had chronic constipation; six patients (86%) had suffered from constipation since childhood. Laxatives were continuously used in 57% of patients. Abdominal pain and bloating were presented in 57% of cases. Barium enema was performed in four cases; the transitional zone could be demonstrated in only one case. Only one patient showed absence of rectoanal inhibitory reflex.

Of the four patients in whom anorectal myectomy was the definitive procedure, all had excellent results with the range of follow up between 26 to 86 months (mean = 55 months). There was no complication from the procedure in this group. The other three patients did not clinically improve after anorectal myectomy and required the second operation. Two patients underwent total colectomy and still had a good result without complication as long as the follow-up period of 18 and 23 months. One of them needed re-admission from partial small bowel obstruction, at 6 months after colectomy. The other underwent left hemicolectomy at 20 months after anorectal myectomy, because of no clinical improvement. At 42 months follow-up, her symptoms did not differ from the pre-operative period, and the transit time demonstrated rectal inertia. Finally, total proctocolectomy with ileal pouch anal anastomosis was performed. She did well and experienced partial small bowel obstruction, which responded well to conservative treatment, at 9 months after the last operation. At 96 months, she still had constipation and required self-enema two times a week (Table 1).

#### **Discussion**

The diagnosis of Hirschsprung's disease is more difficult in adults than in infants, partly because of the rarity of the disease and the higher incidence of short or ultra-short segment aganglionosis. Ultimately, rectal biopsy is required to make a definitive diagnosis<sup>(1,14,15)</sup>. Unlike the classic disease in early life, the typical transitional zone from barium enema may be absent, the short aganglionic segment may be dilated

**Table 1.** Detail of the patients with adults Hirschsprung's disease

No. of patient	Sex	Age (yrs)	Further procedures	Complications	Length of follow-up (mo)	Results
1	F	19	None	None	26	Good
2	M	25	None	None	37	Good
3	F	19	None	None	86	Good
4	F	30	None	None	50	Good
5	F	67	Subtotal colectomy	None	42	Good
6	F	19	Total colectomy	Partial small bowel obstruction	47	Good
7	F	29	Lt.hemicolectomy Total colectomy	Partial small bowel obstruction	96	Poor

sufficiently either by the enema tip or cleansing enemas to conceal its presence<sup>(16)</sup>. In such patients, barium enema without bowel preparation is indicated so that the transitional zone is not altered by a mechanical cleansing<sup>(17)</sup>. In four patients that barium enemas were performed in the present series, all were prepped with laxatives and the typical transitional zone could be demonstrated in only one patient. In a recent study, in children with Hirschsprung's disease, the sensitivity and specificity of this test is 76% and 97% respectively<sup>(18)</sup>. However, the figures in adults are unknown.

Anorectal manometry is a less-invasive diagnostic test; the absence of rectoanal inhibitory reflex is the pathognomonic finding. Accuracy of reflex manometry in establishing the diagnosis is high in most reported series. The sensitivity and specificity in children is 83% and 93% respectively<sup>(18)</sup>. From the presented data, only one from three patients had absence of the rectoanal inhibitory reflex. Since this is a retrospective study, many uncontrolled factors could affect the result. Some of the false-negative results might be a consequence of relaxation of the external anal sphincter or displacement of the transducer probe with side-hole sensors.

The surgical procedures developed to treat this disease in children have been applied to adults, but no significant differences between the results of these procedures have been demonstrated because of the small number of cases and surgeon's preference. Swenson abdominoperineal pull-through operation was reported in 1948 as the first definitive surgery for Hirschsprung's disease<sup>(19)</sup>. The largest experience with this procedure has been reported from the Mayo Clinic<sup>(5)</sup>. Fourteen from seventeen patients had excellent results. The failure rate was 18% (3 patients).

Moreover, the incidence of major postoperative complications was high. Three patients had abscesses (2 presacral and 1 perineum). One of them developed rectal stricture. Impotence occurred in two patients. Anastomotic leakage presented in one case. Despite the successful results, it is generally thought that extensive pelvic dissection may result in a high incidence of major postoperative complications.

Duhamel retrorectal pull-through operation was reported in 1960<sup>(20)</sup>. This operation obviates any extensive pelvic dissection and avoids damage to the sensory fibers of the rectum that may be encountered during the Swenson procedure. Modifications to the original procedure have focused on the elimination of the rectal spur that may be the cause of difficult defecation by obstruction and stasis<sup>(21, 22)</sup>. The largest series of this procedure was from St. Mark's Hospital<sup>(7)</sup>. The results were good in 36 of 39 patients without any operative mortality. However, the major postoperative complications developed in five patients (3 abscesses, 2 fecal fistulas). Two patients initially suffered from fecal impaction in the rectal pouch due to the long colon-rectal septum, which was divided by diathermy with good result. In general, Duhamel procedure appeared to be associated with a lower rate of major postoperative complications than other procedures, and the rate of good long-term results was higher<sup>(1,2,5,6,9,13,23)</sup>.

Soave proposed the endorectal pull-through procedure in 1964<sup>(24)</sup> and some reports reported good results<sup>(25-27)</sup>, but the early literatures reported the high incidence of postoperative complication such as anastomotic leakage, perirectal cuff abscess, and rectal stricture<sup>(5,13)</sup>.

Lynn procedure for posterior anorectal myectomy advocated its usefulness as an initial

approach or complementary procedure approach to short-segment Hirschsprung's disease<sup>(10)</sup>. Although the long-term follow-up from several reports could not demonstrate the good results, 17 of the 35 patients (48.6%) required further surgery, but the result of this low-to-no risk and relatively simple procedure were good in the remaining 16 patients (45.7%)<sup>(3,5,6,10-13)</sup>.

In the presented series, the anorectal myectomy was performed as the first procedure for all seven adult Hirschsprung's disease patients. The results were good in four patients with no complications, and no second procedure was required during the range of follow up between 26 to 86 months. The other three patients did not improve and underwent further procedures. Two patients who underwent subtotal and total colectomy had good results eventually. The only complication occurred to the one with total colectomy. She developed an episode of partial small bowel obstruction, six months after the colectomy. It could be successfully treated conservatively. The last patient, with the longest duration of follow-up, of about six years, still had a poor result, even after left hemicolectomy and total proctocolectomy respectively. The pathologic report of the specimen from the last operation revealed ganglionic segment of ascending colon. From the present study, anorectal myectomy showed acceptable outcome, with good result in four of seven patients (57%) and no complication. In non-improved cases, two of three patients could achieve good results from subtotal or total colectomy, which are familiar to most surgeons.

### Conclusion

Anorectal myectomy, the simple and low morbidity procedure, showed an acceptable outcome and should be considered as the first operation for treatment of adult Hirschsprung's disease.

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## ผลการรักษาโรค Hirschsprung's disease ในผู้ใหญ่ด้วยวิธี anorectal myectomy

จิรวัดณ์ พัฒนอรุณ, ภาวรัฐ เรือนโรจน์รุ่ง, กษยา ตันติพลลาชีวะ, ชูชีพ สหกิจรุ่งเรือง, พุทธรัตน์ อธิษฐานสกุล, อรุณ โรจนสกุล

**วัตถุประสงค์:** โรค Hirschsprung's disease เป็นสาเหตุของภาวะท้องผูกเรื้อรังที่พบได้น้อยในผู้ใหญ่ และมักวินิจฉัยไม่ได้เนื่องจากไม่ค่อยนึกถึง ส่วนใหญ่พยาธิสภาพมักจำกัดอยู่ในช่วงสั้น ๆ ของลำไส้ใหญ่ จึงอาจทำให้อาการ และอาการแสดง ตลอดจนลักษณะทางคลินิกบางอย่างแตกต่างไปจากผู้ป่วยเด็ก ที่ผ่านมามีการรายงานวิธีการผ่าตัดรักษาต่าง ๆ กันไป แต่ละวิธีล้วนมีข้อดีข้อเสียแตกต่างกัน anorectal myectomy เป็นอีกวิธีหนึ่งซึ่งทำได้ง่าย และไม่มีผลข้างเคียงที่รุนแรง การศึกษานี้ต้องการประเมินผลของการรักษา Hirschsprung's disease ในผู้ใหญ่ด้วยวิธี anorectal myectomy ในโรงพยาบาลจุฬาลงกรณ์

**วัสดุและวิธีการ:** เวชระเบียนทั้งหมดของผู้ป่วยที่มีอายุมากกว่า 15 ปี ขึ้นไปซึ่งได้รับการวินิจฉัยว่าเป็น Hirschsprung's disease ตั้งแต่ 1 มกราคม พ.ศ. 2540 ถึง 30 เมษายน พ.ศ. 2551 ถูกนำมาศึกษาวิเคราะห์อย่างละเอียด ข้อมูลทางคลินิก ตลอดจน การตรวจค้นต่าง ๆ ได้รับการตรวจสอบ และบันทึกไว้เพื่อนำมาวิเคราะห์เกณฑ์ทางพยาธิวิทยาที่ใช้ในการวินิจฉัยโรค Hirschsprung's disease คือ การเพิ่มจำนวนของเส้นประสาท cholinergic ในชั้น lamina propria, muscularis mucosae และ submucosa ร่วมกับการที่ไม่มี ganglion cell ในชั้น submucosa ของลำไส้

**ผลการศึกษา:** มีผู้ป่วยที่ได้รับการวินิจฉัยตรงตามเกณฑ์ดังกล่าวทั้งสิ้น 7 ราย ทุกรายมีอาการท้องผูกเรื้อรังการตรวจด้วยภาพถ่ายเอกซเรย์หลังการสวนด้วยสารทึบรังสี และการตรวจวัดความดันในลำไส้ใหญ่ และทวารหนัก มีการทำในผู้ป่วย 4 ราย และ 3 รายตามลำดับ พบลักษณะเฉพาะที่เข้าได้กับภาวะนี้ในผู้ป่วยเพียงหนึ่งราย สำหรับแต่ละวิธีเท่านั้น ผู้ป่วยทุกรายได้รับการรักษาด้วย anorectal myectomy เป็นวิธีแรก และได้ผลดีไม่มีภาวะแทรกซ้อนในผู้ป่วย 4 ราย ตลอดช่วงที่เฝ้าติดตาม (26-86 เดือน) ผู้ป่วยสองรายมีอาการไม่ดีขึ้น จึงได้รับการผ่าตัด subtotal colectomy และ total colectomy ในภายหลัง สุดท้ายก็มีอาการดีขึ้น ส่วนผู้ป่วยรายสุดท้ายอาการยังไม่ดีขึ้น ถึงแม้จะได้รับการผ่าตัดอีกหลายครั้งตามหลัง anorectal myectomy

**สรุป:** anorectal myectomy จัดเป็นวิธีการผ่าตัดรักษา Hirschsprung's disease ที่ทำได้ง่าย และไม่มีผลข้างเคียงที่รุนแรง ตลอดจนผลการรักษาก็เป็นที่น่าพอใจจึงสมควรพิจารณาเลือกใช้เป็นวิธีแรกในการรักษา