

Internal Anal Sphincter and Late Clinical Outcome in Patients with High Type Anorectal Malformation

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Received: 11 Jun. 2007; Received in revised form: 25 Oct. 2007; Accepted: 12 Nov. 2007

Abstract- The purpose of this study is to investigate the age-related improvement of defecation function in high type anorectal malformation in relation to the assessment of the internal anal sphincter. The function of defecation was studied every 5 years up to 15 years postoperatively according to the Japanese scoring system in our hospital in 50 patients operated for high type (recto-urethral fistula) imperforate anus with posterior sagittal anorectoplasty (PSARP) procedure in 25 patients, and 25 patients with endorectal pull-through (ERPT). The internal anal sphincter was assessed by anorectal manometry and histology, and the results were analyzed with the clinical outcomes. The defecation scores of the PSARP cases exceeded those of ERPT cases at all age groups, the averaged total score were 7.0 in the PSARP cases vs. 4.6 for ERPT cases at 5 years old, 7.5 vs. 5.2 at 10 years old, and 8.0 vs. 6.7 at 15 years old. The anorectal reflex was seen in 17 of 25 (68%) PSARP cases examined, whereas seen in 5 (20%) of ERPT cases examined. Histologically, the well-developed and thickened internal circular muscle at the rectal end was found only in 40% of the cases, whereas discontinuation and hypoplasia of the muscle were seen in most of the cases examined. The present results indicate that the internal sphincter muscle at the rectal end may be histologically maldeveloped in high type anorectal malformations; however, they can potentially develop after transplanted and contribute to the improvement of passive continence in the late post-operative period.

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Acta Medica Iranica 2009; 47(2): 139-142.

Key words: Internal anal sphincter, congenital abnormalities, anus, imperforate

Introduction

The clinical impact of the utilization of the intrinsic circular muscle located at the rectal end as internal anal sphincter is widely accepted in the surgical repair of the high type anorectal malformations (1-3) however, the practical significant of this.

Muscle has remained yet controversial in high type anomaly (4,5). The present study aimed to describe the histology of the internal anal sphincter muscle in high type anomaly, and to review the late clinical outcomes in relation to the histological and manometrical findings and the surgical utilization of internal anal sphincter.

Patients and Methods

50 male patients with recto-urethral fistula, the most common high type anorectal malformation in male, were

involved in the current study. The patients had the definitive surgery in our department from 1988 to 2001, and the age at the latest assessment ranged from 6 years through 20 years. Of 50 cases, 25 patients were operated with posterior sagittal anorectoplasty (PSARP) procedure, and 25 with ERPT procedure (6-8).

The post-operative function of defecation was scored in 50 patients at the age of 5,10,15 years according to the clinical scoring system proposed by the Japanese study group of anorectal anomalies (JSGA) (Table 1) (9,10). Defecation problems were studied in EACH patient by retrospective review or interview with full-trained persons. The anorectal manometry was studied postoperatively at the averaged age of 4,8,14 years in all 2 group patients. The pressure profile of the reconstructed anal canal was measured by the open-dip method, and anorectal reflex was examined using k-y microballoon system in each patient.

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Table 1. Clinical scoring for defecation function (Japanese study group of anorectal anomalies)

A - Sensation score	
2	always sensible
1	sometimes lost
0	always insensible
B - constipation score	
4	no constipation
3	infrequent use of laxatives and enemas
2	daily use of laxatives and enemas
1	constipation requiring finger evacuation of stools
C - soiling score	
4	no soiling
3	soiling only at diarrhea
2	infrequent (less than once a week) soiling
1	frequent (twice a week or more) soiling
0	daily soiling
D - staining score	
2	no staining
1	infrequent (less than daily) staining
0	daily staining

In all patients, a 5 mm× 50 mm strip-tissue of the rectal end containing whole thickness was harvested from the posterior wall at the time of the definitive surgery. The tissues were sectioned in the sagittal direction, and the hematoxylin and eosin histology was examined.

Results

Table 2 summarizes the defecation scores of each clinical item in different operative procedures at each age. The scores were generally improved with aging. The patients operated with PSARP procedure revealed definitely higher scores compared to those operated with

ERPT procedures at all age groups; averaged total Score was 7.0 in the PSARP cases VS. 4.6 in ERPT cases at 5 years old, 7.5 VS. 5.2 at 10 years old, and 8.0 VS. 6.7 at 15 years old. In PSARP cases, the sensation was scored full in all patients even at the early age of 5 years, while the soiling score was not fully improved in the older ages. In PSARP group the constipation Score was fully improved by the age of 10 years, whereas the soiling score remained slightly lower. In contrast, in the patients operated with ERPT procedure, the constipation score was not fully improved until older ages, and the soiling score was further lower than the constipation scores at all ages. Defecation habit was established at the averaged age of 4.0 years (ranging 2 to 7 years) in PSARP cases, whereas established at 8.7 years (ranging 6 to 10 years) in the ERPT cases according in the ERPT cases according to the retrospective review, and sometime toilet habit was not fully established in older ages. The toilet troubles during school hours were complained frequently regardless of the operative procedures, such as shortage of time for toileting and discrimination among friends. Among the recently interviewed patients, however 6 out of 25 patients (24%) operated with PSARP procedure complained mild staining during exercises, with no episode of soiling. In contrast in the patients operated with ERPT procedure, 15 out of 25 patients (60%) were suffering from soiling (10 patients) and severe staining (5 patients) during exercises.

17 (68%) out of 25 patients operated with PSARP procedure showed positive anorectal reflex postoperatively, whereas only in 5 (20%) cases operated with ERPT procedure showed the significant reflex. The averaged resting anal pressure was 33.0 cmH₂O in the PSARP cases, and 24.0 cmH₂O in the ERPT cases (Table 3).

Table 2. Scoring of the post-operative function of defecation

Total score Age	5 years	10years	15 years
PSARP	7.0 ± 0.95	7.5 ± 0.53	8.0 ± 0.70
ERPT	4.6 ± 1.14	5.2 ± 1.30	6.7 ± 0.58
Soiling score			
PSARP	4.4 ± 0.67	4.4 ± 0.52	4.6 ± 0.55
ERPT	3.0 ± 1.41	3.0 ± 0.89	4.3 ± 0.58
Constipation score			
PSARP	4.4 ± 0.79	5.0	5.0
ERPT	4.2 ± 1.10	3.6 ± 0.89	4.3 ± 1.15
Sensation score			
PSARP	3.0	3.0	3.0
ERPT	2.8 ± 0.45	3.0	2.7 ± 0.58
Staining score			
PSARP	2.0 ± 0.43	2.1 ± 0.41	2.4 ± 0.58
ERPT	1.2 ± 0.45	1.6 ± 0.55	2.3 ± 0.58

Table 3. Anorectal manometry

Procedure	Resting anal pressure	Anorectal reflex
PSARP	33.0 ± 11.7 cmh ₂ o	68% (17/25)
ERPT	24.0 ± 15.0 cmh ₂ o	20% (5/25)

Histology evaluation revealed, well-defined thickening of the intrinsic circular muscle at the rectal end was in 20(40%) cases, while the muscle bundles were loose, hypoplastic, and surrounded by fibrous tissues in other 10 (20%) patients, among these 10 patients, vacuolar degeneration of muscle fibers was seen in 4 patients (8).

(16%) patients showed the discontinuation or partial thinning of the internal circular muscle at the rectal end. The outer longitudinal and internal circular muscle layers were widely separated at the rectal end with interstitial fibrous tissue in all of 20 patients. Relationship was not evident between the histology of the internal circular smooth muscle at the rectal end and the JSGA total clinical score at 5 years old. In contrast to the internal circular bundles, outer longitudinal muscle was well developed in all patients.

Discussion

Postoperative function of defecation in anorectal malformations may be affected by many factors such as type of anomaly, aging, function of the internal anal sphincter muscle, and the surgical procedures. Among these factors, the functional potential and the appropriate surgical utilization of the internal anal sphincter identified at the rectal end seemed most important for the late clinical outcome.

To study the clinical significant NCE of the internal anal sphincter in the high type anomaly, age-related alteration of the defecation score was analyzed together with the manometric and histological assessment of the internal anal sphincter muscle in the current study. The study was designed to review the cases with a single high type anomaly in male who were operated on in a single institution to eliminate the bias factors such as surgical skill and the type of anomaly. The patients operated with posterior sagittal anoectoplasty (PSARP) (11).

Clinical assessment according to the JSGA scoring system showed age-related improvement of the scores, especially in continence, in each surgical procedure. The scores were generally improved until 15 post-operative years, and no longer altered thereafter. The higher scores, however, may be provided not only by the im-

proved physical function of defecation but also by the innovations of their toilette habit. The patients seem to acquire the appropriate life style to adopt the incontinence for their social lives as they grow older by using enema or laxatives to empty their bowel. Therefore, the clinical assessment by the scoring at higher ages may not fully reflect the postoperative function of the internal anal sphincter.

The current assessment yet showed definitely higher scores in PSARP patients compared to ERPT patients at all age groups, though not statistically significant. In the PSARP procedure, the rectal end including the most caudal internal circular smooth muscle is transplanted downward to the anus, whereas in ERPT procedure, the muscle remained is higher at the fistula site.^{12,13} These clinical outcomes may indicate that the transplanted internal circular muscle works post-operatively as internal anal sphincter to preserve passive continence after the PSARP procedure.

This hypothesis is supported by another observation that anorectal reflex was seen post operatively in 77.8% of the patients operated with sacro-perineal procedure, whereas seen in none of the patients operated with ERPT procedures. The clinical importance of the thickened internal circular smooth muscle has been emphasized in the surgical repair, since this muscle was first identified at the rectal end in high type anorectal malformations (1-3,11,14) and some articles insisted that this muscle functioned postoperatively like internal anal sphincter to preserve passive continence (1,15).

Nevertheless, the internal circular smooth muscle located at the rectal end showed a wide spectrum of histology, and the well-developed and thickened muscle similar to the internal anal sphincter in normal children is rarely seen in the present series. Meier-ruge et al also reported that the intrinsic muscle layers and/or internal anal sphincter were characteristically abnormal and hypoplastic in high type anomalies (16).

The present results may suggest that the most caudal internal circular muscles have the potential to develop as the internal anal sphincter, even though they are histologically loose and undeveloped at the definitive surgery. Husberg et al reported irregularity and varied thickness of internal anal sphincter in the postoperative magnetic resonance imaging (MRI), which did not correlate with the function (17).

The present histological findings and clinical outcomes seemed compatible with those reported by husberg et al in contrast, the lower clinical scores and negative anorectal reflex in ERPT cases indicate that the internal circular muscles preserved at the higher site rarely

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acquire the function as internal anal sphincter. The present results encourage the transplant of rectal end with minimal trimming in the surgical repair of high type anomalies. However, the disturbance of rectal innervations may occur during the surgical mobilization of the rectal end in the PSARP procedure, which may cause severe constipation (18,19).

Holsehneider et al recently pointed out that the innervations of the rectal end might be disturbed primarily in anorectal malformations from his histological investigation, suggesting that this may be the cause of the high incidence of constipation after the transplant procedure 5. Further studies should be required to improve the function of defecation in the high type anorectal malformations. In conclusion, the present results indicate that the internal sphincter muscle at the rectal end may be histologically maldeveloped in high type anorectal malformation; however, they can potentially develop after transplanted and contribute to the improvement of passive continence in the late post-operative period.

Acknowledgements

The authors would like to thank MRS. M. saeedi, who cooperated a lot in typing and editing the text and accompanied us during the analysis.

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