

Functional continence in anorectal malformation after posterior sagittal anorectoplasty

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Abstract. *Background and aim:* Incontinence remains a surgical challenge for patients with anorectal malformations. This study aimed to determine the outcome function of continence in anorectal malformation patients after posterior sagittal anorectoplasty (PSARP) at Dr. Wahidin Sudirohusodo Hospital, considering gender; type of anorectal malformation; sacral ratio; vertebral, anal, cardiac, tracheal, esophageal, renal, and limb (VACTERL) anomalies; and age when undergoing PSARP. *Methods:* This descriptive, analytical, cross-sectional study used primary data (Rintala questionnaire) and secondary data (medical records) at single-center hospitals in Makassar, Indonesia. It investigated 39 patients who had received PSARP over 5 years. *Results:* Of the 39 patients, 24 (61.5%) were male, and 15 (38.5%) were female. The age at which PSARP was performed was ≤ 6 months for 22 (56.4%) patients and after 6 months for 17 (43.6%) patients. A relationship existed between the age when PSARP was performed and continence outcomes ($p=0.034$). Patients who were treated at younger than 6 months had better continence function than those who underwent PSARP after 6 months. The type of anorectal malformation has no influenced the functional outcome of PSARP: most patients with no fistula type 19 (48.7%) ($p=0.198$). Patients without VACTERL anomalies had better functional outcomes than patients with VACTERL anomalies. *Conclusions:* Functional outcomes in anorectal malformation patients were related to the age at which PSARP was performed, type of anorectal malformation, and presence of VACTERL anomalies. The sacral ratio had no relationship with functional outcomes. (www.actabiomedica.it)

Key words: anorectal malformation, posterior sagittal anorectoplasty, outcome, functional

Introduction

Anorectal malformation (ARM) is a congenital abnormality characterized by failure of the anus to develop normally at the perineum. ARM is the most common congenital anomaly, with a prevalence of 1 in 5,000 live births. Diagnosis is confirmed after birth by physical examination (1–4).

The management of ARM has continued to develop from simple cutback and translocation anoplasty and pull-through abdominoperineal and sacroperineal procedures until De Vries and Pena in 1982 introduced

the posterior sagittal anorectoplasty (PSARP) procedure for treating anal atresia in children and infants, as well as several new techniques that have been developed using laparoscopic assistance (5–9). The reconstruction goal for ARM is to obtain adequate defecation function and fecal continence. Several scoring systems have been developed to evaluate the results of postoperative reconstruction in terms of bowel function and continence, such as the Holschneider, Krickenbeck, Kelly's, Pena's, Templeton, and Rintala scoring systems (9–12). Little study has evaluated continence outcomes in Indonesia. This study aimed to

determine continence outcomes after PSARP using the Rintala score in Makassar.

Materials and Methods

This was a categorical, analytical, descriptive, cross-sectional study, derived from patient medical records and questionnaires. The research was conducted in December 2022 at Dr. Wahidin Sudirohusodo Hospital, Makassar, Indonesia. The sample was 39 ARM patients who had undergone PSARP. The inclusion criteria for the study were patients diagnosed with ARM who underwent the first PSARP procedure at our institution, with sufficient data in the medical record, who could still be contacted, were willing to be included in the research, and were aged 3 years or older when the research was conducted, at least 3 months after undergoing PSARP. The exclusion criteria were patients who refused to participate or died before the research was conducted. After each patient met the inclusion criteria, a demographic evaluation was carried out using their medical record, and then a direct interview with parents or guardians was conducted using the Rintala questionnaire to assess the patient's continence function. Sex is a biological characteristic that is divided into male or female. Age at the time of PSARP is determined at ≤ 6 months or >6 months. ARM type is based on the Krickenbeck classification (12)(Table 1). VACTERL-related anomalies are associated if there are 3 or more vertebral, anal, cardiac, tracheoesophageal, renal and limb abnormalities. Sacral ratio assessed based on x-ray pelvic AP view (BC/AB) value <0.74 or ≥ 0.74 (Figure 1).

Table 1. Krickenbeck classification of anorectal malformation.

Major clinical groups	Rare/regional variants
Perineal (cutaneous) fistula	Pouch colon
Rectourethral fistula	Rectal atresia/stenosis
Prostatic	Rectovaginal fistula
Bulbar	H fistula
Rectovesical fistula	Others
Vestibular fistula	
Cloaca	
No fistula	
Anal stenosis	

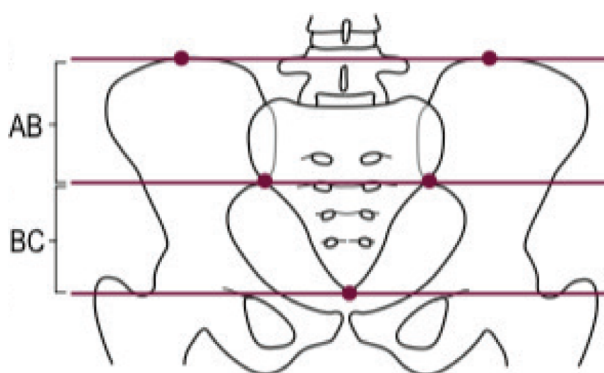


Figure 1. Landmark to calculate sacral ratio.

Rintala scoring system

The reason for choosing the Rintala scoring system for assessment is that the Rintala questionnaire is the only one that has been validated and verified on healthy children (12,13). The Rintala questionnaire has seven questions, all of which have a score between 0 and 3, except the frequency of defecation, which has a value between 1 and 2. The overall score can range from 1 to 20. Based on the scoring system (Table 2), continence outcomes can be categorized as excellent (18–20), good (11–16), fair (9–11), or poor (6–9) (12).

Data analysis

The data were analyzed with univariate correlation analysis to determine the distribution of proportions of demographic characteristics and continence scoring components. The data processing used IBM SPSS Version 21 (Armonk, NY, USA; IBM Corp.). The Chi-square test was used to determine the relationship between two variables in two groups, and multivariate tests were used to assess the relationship between more than two variables (age; anorectal malformation type; VACTERL association; and sacral ratio).

Results

The study used univariate analysis to analyze the relationship between continence function and characteristics in patients with ARM after PSARP

Table 2. Rintala questionnaire.

Ability to hold back	
Always	3
Problem less than once a week	2
Weekly problem	1
No voluntary control	0
Feels/reports urge to defecate	
Always	3
Most of the time	2
Uncertain	1
Absent	0
Frequency of defecation	
Every other day to twice a day	2
More often	1
Less often	1
Soiling	
Never	3
Soiling less than once a week	2
Frequent soiling	1
Daily soiling	0
Accidents	
Never	3
Less than once a week	2
Weekly accidents, often requiring protective aids	1
Daily, require protective aids during day and night	0
Constipation	
No constipation	3
Manageable with diet	2
Manageable with laxatives	1
Manageable with enemas	0
Social problems	
No social problems	3
Sometimes (foul odors)	2
Problem causing restriction to social life	1
Severe social and/or psychic problems	0
Interpretation	
Excellent	18–20
Good	9–16
Fair	7–11
Poor	6–9

at Wahidin Sudirohusodo Hospital in January 2017–December 2022 (Table 3).

Of the 39 patients during this research period (Table 4), 24 (61.5%) were male, and 15 (38.5%) were female. The age at PSARP was ≤ 6 months for 22 (64.1%) patients, showing that this age was more frequent. The perineal fistula was present in 15 (38.5%) patients, rectourethral fistula was present in 1 (2.6%) patient, rectovaginal fistula was present in

Table 3. Characteristics of ARM patients.

Variable	n	%
Sex		
Male	24	61.5
Female	15	38.5
Age at PSARP (months)		
≤ 6	22	56.4
> 6	17	43.6
Anorectal malformation type		
Perineal fistula	15	38.5
Rectourethral fistula	1	2.6
Rectovestibular fistula	3	7.7
Rectovaginal fistula	1	2.6
No Fistula	19	48.7
VACTERL association		
Yes	4	10.3
No	35	89.7
Sacral ratio		
< 0.74	22	56.4
≥ 0.74	17	43.6

3 (7.7%) patients, rectovestibular fistula was present in 1 (2.6%) patient, whereas no fistula was present in 19 (48.7%) patients. Four (10.3%) patients had VACTERL anomalies, whereas 35 (89.7%) did not. A sacral ratio of < 0.74 was present in 22 (56.4%) patients, whereas 17 (43.6%) had a sacral ratio of ≥ 0.74 .

As shown in Table 3, three variables had a p-value of < 0.05 , showing a relationship between the dependent variable and the independent variable, followed by the VACTERL association anomaly variable and continence function ($p=0.022$). There was no relationship between malformation type and continence function ($p=0.198$). The operating age variable was a function of continence ($p=0.34$). The sacral ratio variable had a p-value of 0.105, indicating no relationship with continence function.

Discussion

The sex distribution of patients with ARM was predominantly male. This aligns with research by Makrufardi et al., in which 43 neonate patients were 30.2% girls and 69.8% boys (14). The age at PSARP was classified into < 6 months and ≥ 6 months, with

Table 4. Functional outcomes in ARM patients after PSARP.

Variable	Interpretation				p-value
	Poor n (%)	Fair n (%)	Good n (%)	Excellent n (%)	
Age at PSARP (months)					
≤6	0 (0)	0 (0)	8 (20.5)	14 (35.9)	0.034
>6	0 (0)	4 (10.3)	7 (17.9)	6 (15.4)	
Anorectal malformation type					
Perineal fistula	0 (0)	0 (0)	4 (10.3)	11 (28.2)	0.198
Rectourethral fistula	0 (0)	0 (0)	1 (2.6)	0 (0)	
Rectovestibular fistula	0 (0)	0 (0)	2 (5.1)	0 (0)	
Rectovaginal fistula	0 (0)	0 (0)	1 (2.6)	0 (0)	
No Fistula	0 (0)	3 (7.7)	7 (17.9)	9 (23.1)	
VACTERL association					
Yes	0 (0)	2 (5.1)	1 (2.6)	1 (2.6)	0.022
No	0 (0)	2 (5.1)	14 (35.9)	19 (48.7)	
Sacral ratio					
<0.74	0 (0)	3 (7.7)	11 (28.2)	8 (20.5)	0.105
≥0.74	0 (0)	1 (2.6)	4 (10.3)	12 (30.8)	

most being aged ≥ 6 months. Similar research conducted by Sukarnjanaprai et al. showed that 40 patients (68.9%) were aged >6 months when PSARP was performed (15). Patients who underwent PSARP aged ≤ 6 months had better continence outcomes compared to those who underwent PSARP aged >6 months ($p=0.034$), indicating a relationship between the age when PSARP was performed and continence function. This aligns with research conducted by Sukarnjanaprai et al. in 2017, which showed a relationship between the age at which PSARP was performed and continence function in the form of total continence, voluntary bowel movements, constipation, and soiling ($p=0.047$). This relationship is related to the sphincter complex atrophying because it has not been used for a long time and the development of neurological function between the sphincter muscle complex, anal canal and brain (15). The most common type of malformation found was anorectal malformation without fistula, where no relationship was found between the type of malformation and the patient's continence function; this is in line with research conducted by Sukarnjanaprai et al., which showed that there were 19 patients with anorectal malformation without fistula (32.8%) where there was no relationship between the type of anorectal malformation and functional outcome ($p=0.13$) (15).

No scores were interpreted as poor in this study. This was because continence disorders were detected early and corrective surgery was carried out quickly. Twenty-five patients (64.1%) had no associated abnormalities other than ARM. This is similar to the research of Hondel et al., which found that 58 ARM patients (54%) had no associated anomalies (16). A sacral ratio of <0.75 was found in 24 patients (61.5%). This agrees with research conducted by Sukarnjanaprai et al., which showed a sacral ratio of <0.75 in 23 patients (58.9%) (15). In the group of patients who had VACTERL association anomalies, 2 patients (5.1%) had moderate continence function, 1 (2.6%) had good function, and 1 (2.6%) had normal function. In the group of patients without VACTERL association anomalies, 2 patients (5.1%) had moderate continence function, 14 (35.9%) had good function, and 19 (48.7%) had normal function. The p-value of 0.022 shows a relationship between the VACTERL association anorectal malformation anomaly and continence function after PSARP. This aligns with research by Totonelli et al., which compared two groups (VACTERL- vs VACTERL+) by assessing continence function, finding a significant difference in patients with VACTERL requiring colon management ($p=0.025$). Patients with VACTERL anomalies have worse clinical outcomes in terms

of the need for diet or laxative treatment and bowel management (17). Among patients with a sacral ratio of <0.74 , continence function was moderate for 3 patients (7.7%), good for 11 (28.2%), and normal for 8 (20.5%). Among those with a sacral ratio of ≥ 0.74 , 1 patient had moderate continence function, 4 had good function (10.3%), and 12 had normal function (30.8%). The p-value of 0.105 shows no relationship between the sacral ratio and continence function. This study used imaging techniques in the anteroposterior position because data were not obtained from lateral views in all patients. This result aligns with research conducted by Macedo et al., which showed that of the 42 patients assessed, 17 were considered continent, 7 were partially incontinent, and 18 were incontinent. No significant difference existed in the sacral ratio between the three groups. The number of incontinent patients had no significant difference by sacral ratio ($p=0.77$) (18). Our result contrasts with research conducted by Zadeh et al., which concluded that a low sacral ratio was associated with poor continence function in children. This is related to innervation originating from the lumbosacral area, which influences sphincter activity (19).

Conclusion

Functional outcomes in patients with ARM after PSARP were related to the age at the time of the procedure, the type of location of the malformation, and the presence or absence of accompanying VACTERL anomalies. The sacral ratio had no relationship with the functional outcome of ARM patients after PSARP.

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Data Availability Statement: The data presented in this study are available on request from the corresponding author.

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