

Faecal Continence in Anorectal Malformation, a Correlation with Preoperative and Postoperative Parameters

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ABSTRACT

BACKGROUND

Anorectal malformations are one of the most common congenital defects encountered in paediatric surgery. Operative procedures aim in attaining bowel control which implies the ability to detect and retain flatus and stool until the appropriate time for evacuation. The study aims to determine the fecal continence in anorectal malformation and its correlation with few preoperative and postoperative parameters.

METHODS

60 patients who had completed surgery for anorectal malformation and on follow up in department of Paediatric Surgery, Medical College Kottayam were studied in terms of vertebral anomalies, type of fistula, surgical procedure, position of anus and functional results. Functional results were assessed by Kelly score.

RESULTS

Kelly's score showed that cases treated with PSARP (26.7%) had fair scores. SPM had good score in 28.6%, fair in 50% and poor in 21.4%. Skeletal anomalies was seen in 16% cases of which 90% had only fair scores. Score was good for anocutaneous fistula and vestibular fistula. All bladder neck fistulas and bulbar fistulas had fair scores (100%) while for prostatic fistula, the scores was good in 6.7%, fair in 73.3% and poor in 20%

CONCLUSIONS

Kelly's scoring showed inadequate stooling outcome scores in cases with vertebral anomalies, prostatic fistulas. Scores were fair following PSARP, while in SPM the cases were seen in all groups- good, fair and poor. Many of the patients had improvement in their scores when specific intervention was implemented and thus will improve their quality of life.

KEYWORDS

Anorectal malformation, Kelly score, Faecal incontinence

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BACKGROUND

Bowel control is the result of a normal sphincter function, anorectal sensation, and colonic motility. All these factors are affected in children with anorectal malformations¹. The act of defecation and fecal continence are complex physiological functions depending on a variety of factors including the rectum, internal and external sphincters, pelvic floor muscles, sensory and motor innervation, and integrity of neural (both autonomic and somatic) pathways. Anorectal anomalies present a spectrum of defects. The incidence worldwide is 1 in 2000 to 1 in 5000 live births.^{1,2} The estimated risk for a couple having a second child with an anorectal malformation is approximately 1%.³ Most babies (40% to 60%) with anorectal malformations have one or more abnormalities that affect other systems.^{1,4,5} Sacral deformities are the most frequently associated defect.³ Lumbosacral anomalies such as hemivertebrae, scoliosis, butterfly vertebrae, and hemi sacrum are common. The most frequent spinal problem is tethered cord, which is seen in 25% cases.⁶ of the more complex the anorectal anomaly the more likely is the presence of an associated spinal and vertebral anomaly. Absence of more than two sacral vertebrae represents a poor prognostic sign in terms of bowel continence and urinary control. Hemivertebra also has a negative implication for bowel control. The muscle groups of the sphincter mechanism form a funnel like structure in the pelvis. These muscles are innervated by the pudendal nerve, both motor to the voluntary muscles and sensory to the skin around the anus and anal canal. They are derived from the sacral plexus roots S2 to S4, as well as the autonomic nervous system via the nervi erigentes, from the same segments of the spinal cord. The junction of the levator musculature with the fibers about the anal dimple is defined by a vertical group of striated muscle fibers called the muscle complex. In children with ARMs, there are varying degrees of striated muscle development from normal-looking striated muscle to no muscle seen at operation. Good prognostic indicators for bowel control include a normal sacrum, prominent midline groove (good muscles) and types like rectal atresia, vestibular fistula, imperforate anus without fistula and low anomalies. Bad prognostic indicators include abnormal sacrum, flat perineum (poor muscles), and types like rectovesical fistula and complex anomalies.⁷ Spinal and vertebral anomalies have significant impact on the functional outcome in terms of urinary function and fecal incontinence. Surgical correction of ARM has evolved from simple cut back anoplasty to abdominoperineal pull through, sacroperineal pull through, PSARP, primary neonatal PSARP⁸ and laparoscopy assisted bowel pull through.^{9,10} Constipation is one of the most common sequelae after surgical repair of anorectal malformations. The lower the malformation, the more likely the development of constipation. Constipation that is not properly managed will lead to mega-rectosigmoid, resulting in overflow pseudo incontinence. Many require regular enemas, washouts, or procedures, such as antegrade colonic enemas for management of constipation

or fecal incontinence.¹¹ A patient with a good muscle mechanism and a normal sacrum, is likely to have good bowel control. The long-term studies suggest that good bowel control can be achieved after correction of low anomalies in about 90% of patients.¹² Quality of life is affected in those with fecal incontinence and urological complications.¹³

METHODS

This is an observational study conducted over a period of one year among OPD- and IPD-patients of Department of Paediatric Surgery of Medical College, Kottayam.

Sample Size

60 patients.

Patients who had undergone all stages of surgery for anorectal malformation were included. Children presenting with ARM in the neonatal period, who had undergone initial colostomy/cutback procedure followed by definitive surgery and final stage of colostomy closure during the study period were also included. Cases presenting as cloaca were excluded from the study.

The study was conducted in Department of Paediatric Surgery, Medical College Kottayam during the study period August 2017 to August 2018. Those patients who had completed all stages of surgery for anorectal malformation and arriving at the outpatient department of paediatric surgery for review were included. Pre-operative parameters assessed were the type and severity of vertebral anomalies and type of fistula. Intraoperative findings and type of surgery formed the operative parameters of assessment. Post-operative assessments included calculation of anal index and continence scoring. Post-operative assessment of these patients was done by interviewing the parents for bowel movement pattern, episodes of soiling, constipation, clinical examination scoring and retrospectively studying the medical records available with the patient. After the corrective surgery the patients were evaluated at 6 months post bowel pull through procedure. Those patients who had completed all stages of surgery earlier were evaluated by clinical examination and scoring at time of examination and data collected from their medical records. Data was compiled in MS excel and analysed by using statistical software The Kelly score of continence This system of scoring awards points for three basic parameters continence, staining and sphincter. A score of 5-6 is good, 3-4 is fair, and 0-2 is poor score.¹⁴

Continence

- Normal, no soiling 2
- Occasional accidents, faeces/flatus escape 1
- No control, frequent accidents 0

Staining

- Always clean 2
- Occasional staining 1
- Always stained 0

Sphincter

- Strong and effective squeeze 2
- Weak and partial squeeze 1
- No contraction 0

RESULTS

Of the 60 patients included in the study, majority were below 3 years of age (45%) while 15% were above 6 years of age. 58.3% male children and 41.7% female children were included in the study. Majority had high ARM 46.7% followed by low ARM in 45% and intermediate ARM in 8.3%. 50% of cases had associated anomalies, commonest was genitourinary anomaly (31.7%). Isolated genitourinary was seen in 21.7%. Second commonest were skeletal 16.7% and CVS 16.7% anomalies. Genitourinary anomalies were seen in 3 cases of intermediate ARM (15.7%) and 16 cases of high ARM (84.2%).

Skeletal anomalies were seen in 16% cases and were agenesis of more than one vertebra in 6.7%, hemi vertebrae in 6.7% and agenesis of one vertebra in 5%. Out of the 11 patients with skeletal anomalies, 90% had fair score on Kelley scoring. All patients with an abnormal sacral ratio had higher degrees of soiling (grade 2) and poor Kelley scoring. These points towards a poorer stooling outcome in the presence of vertebral anomalies. Less number of skeletal anomalies in our study may be due to the fact that routine MRI was not done for all patients. Sacral ratio was calculated from AP and lateral imaging films. Line A passing through the superior most point of iliac bone. Line B at the level of posterior inferior iliac spines and inferior most point of both sacroiliac joints. Line C through most distal point of visible coccyx. Normal sacral ratio is >0.7 (BC/AB) correlates with good functional prognosis.¹⁵

Arm	Normal	Skeletal Abnormality			Total	Chi-square
		Agenesis of 2 or more vertebrae	Agenesis of 1 vertebra	Hemivertebrae		
Low (n=27)	27	0	0	0	27	13.970 p=0.030
Intermediate (n=5)	4	1	0	0	5	
High (n=28)	18	3	3	4	28	
	64.3%	10.7%	10.7%	14.3%	100.0%	
Total	49	4	3	4	60	
	81.7%	6.7%	5.0%	6.7%	100.0%	

Table 1. Skeletal Anomalies

Out of the 60 cases 23.3% had no fistula. Among those with fistula, majority of the patients had vestibular fistula (26.7%) followed by recto prostatic (25%), recto bulbar (10%) perineal fistula (10%) and bladder neck fistula (3%). Kelly's score was good for anocutaneous fistula and vestibular fistula. All cases with bladder neck fistula (100%) had only fair score, none had good score while Prostatic fistula had scores good in 6.7%, fair in 73.3% and poor in 20%. Bulbar fistula also had fair scores in all cases (100%).

Kelley's	N	Mean Anal Index	S.D.	ANOVA (f value)
Good	30	.3767	.07097	1.123 p=0.332
Fair	27	.3650	.06384	
Poor	3	.3167	.07638	
Total	60	.3684	.06816	

Table 2. Kelly Score and Mean Anal Index

Fistula	Kelley's			Chi-square
	Good	Fair	Poor	
No fistula	8	6	0	14
	57.1%	42.9%	.0%	100.0%
Bladder neck	0	3	0	3
	.0%	100.0%	.0%	100.0%
Prostatic urethra	1	11	3	15
	6.7%	73.3%	20.0%	100.0%
Bulbar urethra	0	6	0	6
	.0%	100.0%	.0%	100.0%
Vestibular	15	1	0	16
	93.8%	6.2%	.0%	100.0%
Anocutaneous	6	0	0	6
	100.0%	.0%	.0%	100.0%
Total	30	27	3	60
	50.0%	45.0%	5.0%	100.0%

Table 3. Type of Fistula and Kelly Score

The surgical procedures were PSARP in 26.7%, ASARP in 25%, SPM in 23.3%, anoplasty in 20% and abdominoperineal pull through in 5% patients. Functional assessment with Kelly's score showed good outcome in 50%, fair in 45% and poor in 5% of the 60 cases. Kelly's score was good in all of the low ARM cases treated with anoplasty (100%). Out of the 19 cases who were treated with ASARP 93.3% had good score and rest of the 6.7% had fair score. All cases treated with PSARP had fair scores irrespective of type of fistula. All cases of abdominoperineal pull through (3 cases) also had a fair score. SPM had good score in 28.6%, fair in 50% and poor score in 21.4%.

Surgery	Kelley's			Chi-square
	Good	Fair	Poor	
Anoplasty	12	0	0	12
	100.0%	.0%	.0%	100.0%
ASARP	14	1	0	15
	93.3%	6.7%	.0%	100.0%
PSARP	0	16	0	16
	.0%	100.0%	.0%	100.0%
Abdominal PSARP	0	3	0	3
	.0%	100.0%	.0%	100.0%
SPM	4	7	3	14
	28.6%	50.0%	21.4%	100.0%
Total	30	27	3	60
	50.0%	45.0%	5.0%	100.0%

Table 4. Procedure and Kelly Score

Anal position index (API) which is the ratio of anal-fourchette distance to coccyx-fourchette distance for females and the ratio of anal-scrotum distance to coccyx-scrotum distance for males, defined the normal position of the anus in the new-born. Out of the 60 patients, 43 patients had an abnormal anal index. These patients were managed with diet modification (12 patients), laxatives (26 patients), and daily enemas (5 patients). Those patients who were managed with enemas (5 cases) had abnormal anal indices in all 5 cases. Anal index and continence was studied but was found that the relation is not statistically significant. Inaccuracy in measurements or non-availability of a standard method for measuring may have contributed to such a result in our study.

DISCUSSION

Most patients who undergo repair of an ARM suffer from variable degrees of faecal incontinence, constipation and urinary symptoms depending upon the type of anomaly, associated anomalies and the effectiveness of the definitive procedure. In our study skeletal anomalies were seen in 16% cases and were agenesis of more than one vertebra in 6.7%, hemi vertebrae in 6.7% and agenesis of one vertebra in 5%. Venkat Shankar Raman et al reported that poorer stooling outcomes were twice as common in children with local pelvic MRI abnormalities as compared to children with normal MRI.¹¹ In our study, out of the 11 patients with skeletal anomalies, 90% had fair score on Kelly scoring. All patients with an abnormal sacral ratio had higher degrees of soiling (grade 2) and poor Kelly scoring. These points towards a poorer stooling outcome in the presence of vertebral anomalies. Less number of skeletal anomalies in our study may be due to the fact that routine MRI was not done for all patients. Pernilla Stenströma et al, found that males with rectourethral fistulas and concomitant sacral malformations, had significantly less voluntary bowel movements (100%) and a higher frequency of faecal incontinence compared to those without sacral malformations 100% and 55%, respectively.¹⁶

Functional assessment with Kelly's score showed good outcome in 50%, fair in 45% and poor in 5% of the 60 cases. Comparing the Kelly's score for low, intermediate and high ARM, it was seen that score was good in all of the low ARM cases treated with anoplasty (100%). Intermediate ARM cases had good score in 40% and fair score in 60%. High ARM cases had good score in 7.1%, fair in 82.1% and poor score in 10.7%. V Bhatnagar found that the clinical outcome depends on the type of anomalies (continence rates: low-90%; high- ~50%). The study also found that anorectal manometry assesses functional compliance, correlates well with Kelly's score and can predict long-term results, and should be used for decision regarding re-operation.¹⁷ We had one child who underwent SPM, and the child had persistent urinary leak from the neorectum following SPM. The child was treated with a redo surgery (PSARP). We have not used anorectal manometry for functional assessment, but Kelly score itself is a good predictor of bowel function.

All cases treated with PSARP (26.7%) had fair scores. All cases of abdominoperineal pull through (3 cases) also had a fair score. SPM had good score in 28.6%, fair in 50% and poor score in 21.4%. In the study Sacroperineal mobilization versus posterior sagittal anorectoplasty: a study on outcome, K Sivakumar compared the outcomes of PSARP and SPM and found that functional assessment by Kelly score, voluntary bowel movement and sensation doesn't reveal any difference between two procedures for high ARM, whereas for intermediate anomalies, Stephens' procedure seems to give better functional result.¹⁸ Rintala et al compared long term outcomes in PSARP and SPM, and found that PSARP patients had significantly higher anorectal resting and squeeze pressures and voluntary sphincter force¹⁹. 23.3% of our cases had no fistula. Among those with

fistula, majority of the patients had vestibular fistula (26.7%) followed by recto prostatic (25%), recto bulbar (10%) perineal fistula (10%) and bladder neck fistula (3%). Kelly's score and type of fistula was studied and was good for anocutaneous fistula and vestibular fistula. All cases with bladder neck fistula (100%) had only fair score, none had good score while Prostatic fistula had scores good in 6.7%, fair in 73.3% and poor in 20%. Bulbar fistula had fair scores in all cases (100%).

Reisner et al described anal position index (API). API was defined as the ratio of anal-fourchette distance to coccyx-fourchette distance for females and the ratio of anal-scrotum distance to coccyx-scrotum distance for males, to define the normal position of the anus in the newborn^{20,21} and suggested that API of less than 0.46 in boys and less than 0.34 in girls was indicative of anterior displacement of the anus²². Out of the 60 patients in our study, 43 patients had an abnormal anal index. These patients were managed with diet modification (12 patients), laxatives (26 patients), and daily enemas (5 patients). Anal index values showed that those patients who were managed with enemas (5 cases) had abnormal anal indices in all 5 cases. Mohamed shahin et al in a study to investigate the association of anterior displacement of the anus with constipation during infancy reported an incidence of anteriorly displaced anus in 24.25%, with the incidence significantly higher in females than males (32.0% and 16.5%, respectively).²² The incidences of anteriorly displaced anus in infants with constipated events during first 4 months were higher than normal infants. Anal index and continence were evaluated in our study but was found that the relation is not statistically significant.

CONCLUSIONS

Most patients who undergo repair of an ARM suffer from variable degrees of faecal incontinence, constipation and urinary symptoms depending upon the type of anomaly, associated anomalies and the effectiveness of the definitive procedure. The commonest associated anomaly was genitourinary anomaly. All the patients with an abnormal sacral ratio had higher degrees of soiling (grade 2) and poor Kelly scoring. These points towards a poorer stooling outcome in the presence of vertebral anomalies. Recto prostatic fistulas had poor Kelly scores compared to other type of fistulas probably due to decreased muscle more associated with this high fistula. Kelly's scoring showed a good functional outcome following PSARP with all the patients having fair scores, while in SPM the cases were seen in all groups of Kelly scoring- good fair and poor. ASARP had good scores in majority of the cases. Anal index and continence were evaluated in our study but was found that the relation is not statistically significant. Anal position index needs further studies and accurate measurements are needed which may prevent incorrect anal placement. To conclude, it is the intrinsic factors of the malformation like type of anomaly low or high and association with vertebral

defects form the true determinants of continence rather than the procedure which is undertaken. Many of the patients had improvement in their scores when specific intervention was implemented and continued with regular follow up. Post-operative assessments, specific interventions and regular long term follow up can improve the continence of ARM patients and thus will improve their quality of life.

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