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Original Research Article

Assessment of post-surgical functional outcome in children with anorectal malformation

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ABSTRACT

Background: Anorectal malformations (ARMs) comprise a spectrum of congenital anomalies that continue to present a challenge for paediatric surgeons. Advances in modern surgical techniques and neonatal care have greatly improved survival among ARM patients over the last decades, and early mortality is now unusual in the absence of fatal associated cardiac or chromosomal defects. The aim of this study is to measure the functional outcome of ARM by most recent krickenburg classification.
Methods: The present longitudinal study was conducted in Department of paediatric surgery, Dr. BRAM hospital, Raipur, Chhattisgarh during study period February 2016 to September 2017. Those patients who had completed their all stages of surgery for anorectal malformation at-least 6 months back and arriving at outpatient department of paediatric surgery were included.
Results: Maximum number of patients were in age group of 3 to 5 years (77.8%) and minimum were 9 to 11 years (3.7%). A 50.6% male and 49.4% female child were included in the study. Maximum number of patients had vestibular fistula (38.27% followed by perineal fistula (24.69%), rectobulbar (18.51%) then rectovaginal (7.4%) and rectoprostatic (7.4%). Minimum patients had pouch colon (2.4%) and cloaca (1.09%). Voluntary bowel movement was present in 50% of rectoprostatic and 66.6% of rectovaginal fistula. Eighty percentage rectobulbar and 83.87% vestibular fistula had voluntary bowel movement. Cloaca and Pouch colon had no voluntary bowel movement. In recto-vaginal fistula 66.6% had soiling, of which 50% had grade 1 and 16.6% had grade 2 soiling. In recto-bulbar fistula 40% cases had soiling, out of which 20% had grade1 and 20% had grade 2. In recto-vaginal fistula cases 33 % had constipation, of which 16.5% had grade 1 and 16.6 % had grade 2. In recto-prostatic fistula 16% had constipation which was grade1.
Conclusions: In this study, author showed that functional outcomes comparable to matched peers are achieved in the majority of low ARMs after minimally invasive, individualized perineal procedures and regular surgical follow-up. In high type of ARM soiling is the prominent feature while in intermediate ARM constipation is more common.

Keywords: ARM, Cloaca, Functional outcome, Vestibular fistula

INTRODUCTION

Anorectal malformations (ARMs) comprise a spectrum of congenital anomalies that continue to present a challenge for paediatric surgeons.[1] ARMs affect around 1:2000-2500 births, ranging in severity from mild anterior displacement of the anus to very complex malformations of the hindgut and urogenital tract.[2-4] Advances in modern surgical techniques and neonatal care have greatly improved survival among ARM patients over the

last decades, and early mortality is now unusual in the absence of fatal associated cardiac or chromosomal defects.[5]

Accordingly, the focus of surgical care has shifted beyond initial survival of the patient towards ensuring that children treated for ARMs to grow up having bowel function that is compatible with a good quality of life.[6] For most, this means being able to actively participate in their social environment without significant limitations from bowel function, for which fecal continence is a major determinant.[7-9] Posterior sagittal anorectoplasty (PSARP), first introduced by De Vries and Peña and followed later by its limited modification anterior sagittal anorectoplasty (ASARP), represents the basis of the modern surgical approach to ARMs with termination of the anal canal outside the voluntary sphincter complex.[10,11]

The outcome for patients with ARM is related to the severity of the anomaly. There is a lot of confusion regarding the nomenclature and uniformity in clinical diagnosis, investigations and surgical approaches for these malformation and therefore postoperative result are also difficult to correlate and compare. The aim of this study is to measure the functional outcome of ARM by most recent krickenburg classification, this include three parameters: voluntary bowel movements (VBM) (yes/no), soiling (yes/no, if yes grade 1-3), and constipation (yes/no, if yes grade 1-3).

METHODS

The present longitudinal study was conducted in Department of paediatric surgery, Dr. BRAM hospital, Raipur Chhattisgarh during study period February 2016 to September 2017.

Those patients who had completed their all stages of surgery for anorectal malformation at least 6 months back and arriving at outpatient department of paediatric surgery were included. Anatomical anomalies were classified according to Krickenbeck classification and standard procedures were done.

Data was collected using a functional outcome questionnaire for a minimum of six months after surgical reconstruction. Outcome measurements were related to the krickenbeck scoring system which is most recent scoring system and not many studies have been done to validate it. Data was compiled in MS excel and checked for its completeness and correctness. Then the data was analyzed by using suitable statistical software.

Method for assessment of outcome established in Krickenbeck[9]

Voluntary bowel movements (yes/no)

- Feeling of urge
- Capacity to verbalize
- Hold the bowel movement

Soiling (yes/no)

- Grade 1 Occasionally (once or twice per week)
- Grade 2 Every day, no social problem
- Grade 3 Constant, social problem

Constipation (yes/no)

- Grade 1 Manageable by changes in diet
- Grade 2 Requires laxatives
- Grade 3 Resistant to diet and laxatives

Inclusion criteria

- Children age more then 3 years and less then 14 years.
- Follow up postoperative cases of ARM who have completed all stages of surgery at least 6 months back
- Not on any therapy

Exclusion criteria

- Children age more than 14 years and <3 year.
- Cases who have not completed all stages of surgery.
- Cases with other associated anomaly which will affect the continence.
- Cases with anal stenosis or rectal ectasia at the time of assessment.
- On therapy

RESULTS

Maximum number of patients were in age group of 3 to 5 years (77.8%) and minimum were 9 to 11 years (3.7%). 50.6 % male and 49.4 % female child were included in the study.

A 79% of total patient have institutional delivery while 21% have home delivery. A 53.1% patient were 1st born child subsequently 35.8% were 2nd born, 8.3 were 3rd born and 2.5% were 4th born child.

Maximum number of patients had vestibular fistula (38.27% followed by perineal fistula (24.69%), rectobulbar (18.51%) then rectovaginal (7.4%) and rectoprostatic (7.4%).

Minimum patients had pouch colon (2.4%) and cloaca (1.09%). ASARP was performed in 48.14, PSARP in 27.16%, anoplasty in 20.98% and abdomino-perineal pull through was done in 3.7% patients in this study of 81 patients (Table 1).

Table 1: Background characteristics of study subjects.

		Frequency	%
Age	3-5 years	63	77.8
	6-8 years	15	18.5
	9-11 years	3	3.7
Sex	Male	40	49.4
	Female	41	50.6
Krickenbech major variant	Perineal fistula	20	24.69
	Bulbar fistula	15	18.51
	Prostaic fistula	6	7.4
	Vestibular fistul	31	38.27
	Cloaca	2	2.4
Krickenbach rare variant	Pouch colon	1	1.09
	Rectovaginal fistula	6	7.4
Standard procedure for ARM	Abdomino-perineal Pullthrough	3	3.70%
	Anoplasty	17	20.98%
	ASARP	39	48.14%
	PSARP	22	27.16%

In this study voluntary bowel movement was present in 50% of rectoprostatic and 66.6% of rectovaginal fistula. 80% rectobulbar and 83.87% vestibular had voluntary bowel movement. Cloaca and Pouch colon had no voluntary bowel movement (Table 2).

Table 2: Distribution of patients according to voluntary bowel movement.

Krickenbeck subtypes	Voluntary bowel movement	
	Yes	No
Cloacal anamaly	0 (0%)	2(100%)
Pouch colon	0(0%)	1(100%)
Rectoprastatic fistula	3(50%)	3(50%)
Rectovaginal fistula	4(67%)	2(33%)
Rectobulbar fistula	12(80%)	3(20%)
Vesitbular fistula	26(83.8%)	5(16.12%)
Perineal fistula	20(100%)	0(0%)

Pouch colon (1 patient) had grade 2 soiling. In recto-vaginal fistula 66.6% had soiling, of which 50% had grade 1 and 16.6% had grade 2 soiling. In recto-bulbar fistula 40% cases had soiling, out of which 20% had grade1 and 20% had grade 2. In vestibular Fistula 22.58% had soiling (grade 1).

None of the patients with cloaca and pouch colon had constipation. In recto-vaginal fistula cases 33 % had constipation, of which 16.5% had grade 1 and 16.6 % had grade 2. In recto-prostatic fistula 16% had constipation which was grade1. A 46% of recto-bulbar fistula patient shows constipation, from which 20% had grade 1 and 26% grade 2, while in vestibular fistula cases 59% had constipation of which 35.5% had grade 1 and 22.6 had grade 2 constipation (Table 3).

Table 3: Distribution of patients according to soiling grades and constipation grades.

Krickenbeck subtypes	Soiling grades				Constipation grades			
	Soiling absent	**1**	**2**	**3**	**Constipation absent**	**1**	**2**	**3**
Cloaca	0 (0%)	0(0%)	1(50%)	1(50%)	2(100%)	0(0%)	0(0%)	0(0%)
Pouch colon	0(0%)	0(0%)	1(100%)	0(0%)	1(100%)	0(0%)	0(0%)	0(0%)
Rectoprostatic fistula	2(33%)	2(33%)	2(33%)	0	5(83%)	1(17%)	0(0%)	0(0%)
Rectovaginal fistula	2(33%)	3(50%)	1(17%)	0(0%)	4(67%)	1(17%)	1(17%)	0(0%)
Rectobulbar fistula	9(60%)	3(20%)	3(20%)	0(0%)	8(53%)	3(20%)	4(27%)	0(0%)
Vesitbular	24(77%)	7(23%)	0(0%)	0(0%)	13(42%)	11(35.5%)	7(22.5%)	0(0%)
Perineal fistula	19(95%)	1(5%)		0(0%)	6(30%)	9(45%)	4(20%)	1(5%)

DISCUSSION

Over the years, many different scoring systems have been employed for the evaluation of outcomes following the surgical treatment of ARMs, which has presented challenges for the later comparison of outcomes between series. Fortunately, the major scoring systems have placed fecal continence as the most important endpoint in patients with ARMs and have focused on the evaluation of this from different perspectives.[12] The Kelly score introduced a quantitative scoring system based on functional and objective criteria.[13] Later, Holschneider and Metzer built on the concept of quantitative clinical scoring and added manometric parameters to the evaluation.[14] The Wingspread Score approaches the problem from a slightly different angle by gauging the functional outcome from the degree of therapy required for symptom control.[15] These systems have all contributed to the development of further models of evaluation. The system of Peña importantly brought in the concept of voluntary bowel movements (VBMs) as one of its major criteria of assessment.[16] VBMs, defined as the ability to recognise the urge to defecate, the

capacity to verbalise this and the ability to hold the movement, have since established a key role as in the reporting of outcomes for ARM patients.[17] Author measure the functional outcome of ARM by most recent krickenburg classification, this include three parameters: voluntary bowel movements(VBM) (yes/no), soiling (yes/no, if yes grade 1-3), and constipation (yes/no, if yes grade 1-3).

Voluntary bowel movement was present in 50% of rectoprostatic and 66.6% of rectovaginal fistula. Eighty percentage rectobulbar and 83.87% vestibular fistula had voluntary bowel movement. Cloaca and Pouch colon had no voluntary bowel movement. The results were comparable to global function results. Perhaps the most important factor in fecal continence is bowel motility. In a normal individual the rectosigmoid remains quiet for variable periods of time (one to several days), depending on specific defecation habits. The peristaltic contraction of the rectosigmoid that occurs prior to defecation is normally felt by the patient. The normal individual can voluntarily relax the striated muscles, which allows the rectal contents to migrate down into the highly sensitive area of the anal canal. There, accurate information is provided concerning the consistency and quality of the stool. The voluntary muscles are used to push the rectal contents back up into the rectosigmoid and to hold them, if desired, until the appropriate time for evacuation. At the time of defecation, the voluntary muscle structures relax. The main factor that provokes the emptying of the rectosigmoid is a massive involuntary peristaltic contraction that is sometimes helped by a Valsalva maneuver. Most patients with ARM suffer from disturbance of this sophisticated bowel motility mechanism. Patients who have undergone a PSARP or any other type of sacroperineal approach, in which the most distal part of the bowel was preserved, show evidence of an over efficient bowel reservoir (mega-rectum). The main clinical manifestation of this is constipation, which seems to be more severe in patients with lower defects. Those patients treated with techniques in which the most distal part of the bowel was resected behave clinically as individuals without a rectal reservoir. This is a situation equivalent to a perineal colostomy. Depending on the amount of colon resected, the patient may have loose stools.

In the current study, Pouch colon (1patient) had grade 2 soiling. In rectovaginal fistula 66.6% had soiling, of which 50% had grade 1 and 16.6% had grade 2 soiling.

In intermediate type ARM with rectobulbar fistula 40% cases had soiling, out of which 20% had grade1 and 20% had grade 2. In vestibular Fistula 22.58% had soiling (grade 1).

In low type ARM only 2.5% patient had soiling. The results were comparable to the study done by Pena et al.[18] Nixon and Puri used a questionnaire to the parents, hospital records, personal interview, and clinical examination to classify bowel control into three groups: normal control, occasional soiling, and frequent soiling or colostomy. [19] A patient with a good result was defined as being continent most of the time, suffering only occasional soiling during diarrhea and physical stress. A fair result was one in which there was occasional soiling with normal stool consistency, but acceptable social continence. Frank incontinence or permanent colostomy was considered a poor result.

The etiology of defecation problems is multifactorial and includes: (1) sacral malformations (2) altered rectosigmoid motility (3) sphincteric insufficiency and (4) secondary psychological problems. In some cases, fecal incontinence is a complication of surgery (e.g., a mislocated rectum); however, in most children fecal incontinence is secondary to the defect. Children with sacral agenesis, and males with a rectal fistula to the bladder neck had the highest rates of fecal incontinence, followed by females with a high confluence cloaca.

In this study, none of the patients with cloaca and pouch colon had constipation. In rectovaginal fistula cases 33% had constipation, of which 16.5% had grade 1 and 16.6% had grade 2. In rectoprostatic fistula 16% had constipation which was grade1. In this study 46% of rectobulbar fistula patient shows constipation, from which 20% had grade 1 and 26% grade 2, while in vestibular fistula cases 59% had constipation of which 35.5% had grade 1 and 22.6 had grade 2 constipation. Study shows 70% perineal fistula had constipation, 45% grade 1, 20% grade 2 and 5% had grade 3. Abnormal colonic motility, usually presenting with constipation, has been reported to be a problem in patients with low ARM and in females with a vestibular fistula. Since the advent of PSARP for higher anomalies cumulative evidence has shown that chronic constipation is one of the main functional complications encountered following repair. The incidence of constipation following the PSARP procedure varies in the literature between less than 10% and 73%. Constipation seems to be more common when internal-sphincter-preserving techniques have been used. The cause of constipation is unclear; extensive mobilization of the anorectum may cause partial sensory denervation of the rectum and impair rectal sensation. Rectosigmoid hypomotility has also been suggested. Many patients suffering from postoperative constipation have a dilated rectosigmoid. In some, the dilatation of the rectum is present at birth, others appear to develop dilatation later in life. The dilatation is only rarely related to stenosis of the bowel outlet. Segmental colonic transit time studies in patients with ARM has shown that those with low anomalies have rectosigmoid hypomotility, whereas those with high anomalies have a generalized colonic motility disturbance.

The data shows that soiling and constipation can be present in same child due to pseudo-incontinence. The effective and timely management of constipation, which affects all groups of patients with ARMs, is central to

achieving optimal outcomes. With the implementations of newer techniques like PSARP significantly improves the functional outcome due to splitting of the voluntary sphincter muscles in the midline.

CONCLUSION

In this study of measurement of functional outcome of ARM according to krickenbeck scoring system author found that it is easy to perform, convenient, no need of physical examination or any procedure like manometry and even no need to attend the child because questionnaire can be asked to his parents also while the results are comparable with other scoring systems.

In this study, author showed that functional outcomes comparable to matched peers are achieved in the majority of low ARMs after minimally invasive, individualized perineal procedures and regular surgical follow-up. In high type of ARM soiling is the prominent feature while in intermediate ARM constipation is more common.

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