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Functional Outcome of Anorectal Malformations and Associated Anomalies in Era of Krickenbeck Classification

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ABSTRACT

Objective: To describe the management and functional outcome of anorectal malformations and associated anomalies according to Krickenbeck classification.

Study Design: Case series.

Place and Duration of Study: The Aga Khan University Hospital, Karachi, from January 2002 to December 2012.

Methodology: Anorectal anomalies were classified according to Krickenbeck classification. Data was collected and proforma used regarding the primary disease associated anomalies, its management and functional outcome, according to Krickenbeck classification. Cases included were: all those children with imperforate anus managed during the study period. Qualitative variables like gender and functional outcome were reported as frequencies and percentages. Quantitative variables like age were reported as medians with interquartile ranges.

Results: There were 84 children in study group. Most common associated anomaly was cardiac (38%), followed by urological anomaly (33%). All children were treated by Posterior Sagittal Anorectoplasty (PSARP). Fistula was present in 64 out of 84 (76%) cases. The most common fistula was rectourethral (33%), followed by recto vestibular (31%). According to Krickenbeck classification, continence was achieved in 62% children; however 27% children were constipated, followed by 12% children having fecal soiling.

Conclusion: Functional outcome of anorectal malformation depends upon severity of disease. A thorough evaluation of all infants with ARM should be done with particular focus on cardiovascular (38%) and genitourinary abnormalities (33%).

Key Words: Anorectal malformation. Functional outcome. Krickenbeck classification.

INTRODUCTION

Anorectal malformations (ARMs) are common anomalies in neonates.¹ The catastrophe happens as a result of antenatal dysmorphogenesis of cloaca and urorectum.² Currently survival rate of ARMs is much improved because of advancement in surgical techniques and availability of improved neonatal perioperative intensive care facilities. Mostly, children are diagnosed postnatally, and have associated other anomalies; mostly, urological, cardiac, and musculo-skeletal systems.³ Majority of these babies have associated recto urinary fistula. Recto urethral and recto vestibular fistulae are the most common presentations respectively in males and females.³

Ammussat, the father of proctoplasty, in 1835 attempted the first classification of ARMs. His work was followed by Ladd and Gross in 1934, producing the first standard classification of this anomaly. Further development led to an international classification on the basis of puborectalis muscle, followed by International classification

in 1970 and Wingspread classification in 1984. Pena's classified ARMs on the bases of presence and position of fistula. He also shared his experience with a new surgical technique, i.e. Posterior Sagittal Anorectoplasty (PSARP).

All these classifications categorized the ARMs but there were variations in terms of follow-up of these children as it was difficult to compare the functional outcome of these children. Krickenbeck group in 2005 published their findings and incorporated criteria from Wingspread and Pena's works. They gave the concept of categorization of ARMs in to three categories, i.e. diagnostic category, surgical procedure category, and functional outcome category. Since then it is this Krickenbeck classification that has been used to measure functional outcome of patients of anorectal malformation.

To the best of authors' knowledge, there is no national study available regarding the classification of this anomaly according to this classification. This study was conducted with the objective of describing the management and functional outcome of anorectal malformations and associated anomalies, according to Krickenbeck classification.

METHODOLOGY

It was retrospective case series. Prior approval was taken from the Medical Record Audit Committee at The

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Aga Khan University Hospital, Karachi. Medical records of all the patients were reviewed who had needed surgical intervention and presented over a period of 10 years from January 1, 2002 to December 31, 2012 at the Aga Khan University Hospital, Karachi, Pakistan. The inclusion criterion was all children with anorectal malformations who were born and presented to Emergency / Pediatric Surgery clinics at the study centre and needed surgical intervention during the study period. Children who were shifted out of the Hospital and those with inadequate follow-up information were excluded from the study.

There was a standard protocol of managing all children presented with anorectal malformation with fistula as multistage surgery; first divided sigmoid colostomy followed by standard Posterior Sagittal Anorectoplasty (PSARP), and colostomy closure usually 6 to 8 months after the PSARP. Regular fortnightly follow-up initially and then monthly clinic visits were required depending on the outcome of all these children in the surgical outpatient clinic for monitoring of regular anal dilatation and postoperative wound care. Throughout the postoperative course, there was a close communication with parents all the time regarding the need of regular anal dilatation and toilet training.

Cases were identified via Hospital Information Management System (HIMS) by using International classification of Disease (ICD-9-CM) codes. ARMs were classified and functional outcomes were assessed according to Krickbeck classification using detailed questionnaire completed at each child visit whenever possible or by contacting the parents via telephone. Collected data were double entered in Epi-Data (version 3.2) by two different data entry persons. SPSS (Statistical Software for Social Sciences) version 17 was used for statistical analysis. Qualitative variables like gender and functional outcome were reported as frequencies and percentages. Quantitative variables like age were reported as medians with interquartile ranges.

All the possible efforts were made to maintain the confidentiality of patients. No identifiable information was collected. Data was stored under lock and key in the custody of the principal investigator.

RESULTS

There were 84 children including 57 (68%) males and 27 (32%) females. The median age at presentation was the day of birth, as 56% of children presented immediately after birth. Associated anomalies were present in 45 (53%) children. Cardiac anomalies (17/45, 38%) were the most common co-existed anomalies with anorectal malformations, followed by urological anomalies in 15/45 (33%) children. Fistula was present in 64/84 (76%) children. Out of those who had associated fistula,

recto-urethral fistula was present in 21/64 (33%), followed by recto-vestibular fistula in 20/64 (31%, Table I).

Three-stage procedures were performed in 64/84 (76%). All children with fistula were managed initially by making divided descending colostomy followed by PSARP, while 5 children (6%) were offered single-stage limited PSARP. All of them were newborn and were offered single-stage procedure (Table II). There was one child who had a perineal repair and 8 children were managed by cutback anoplasty (Table III). All children with PSARP had colostomy closure later on. There were 18 children who developed complications later (Table IV). The most common complication was anal stenosis (8/84, 10%). Four children later presented with recurrent fistulae, which were managed by Redo PSARP.

The functional outcome of the patients, who were more than 3 years, was assessed according to the Krickbeck classification and found that 32/52 (62%) were continent. Most of them had low anorectal anomalies (17/32, 53%). However, there were 14/52 (27%) children who had constipation followed by different grading of soiling (6/52, 12%), reported by the parents (Figure 1).

Table I: Frequency of different types of fistulae associated with anorectal anomalies.

Presence of fistula - 64	
Rectourethral	21/64 (33%)
Rectovesicular	11/64 (17%)
Rectovestibular	20/64 (31%)
Rectoperineal	06/64 (9%)
Rectovaginal	06/64 (9%)

Table II: Types of surgical interventions.

Surgical intervention - 84	
Divided descending colostomy	64 (76%)
Total PSARP	69 (82%)
Limited PSARP (single stage)	05 (6%)
Anoplasty	08 (10%)
Perineal repair	01 (1%)

Table III: Frequency of other associated anomalies with anorectal malformations.

Other anomalies - 13	
Ectopic anus	05/84 (6%)
Anal atresia	03/84 (4%)
Pouch colon	02/84 (2%)
Cloacal malformation	03/84 (4%)

Table IV: Surgical complications.

Surgical complication - 18	
Anal stenosis	08 (10%)
Stoma prolapsed	05 (6%)
Recurrence of fistula	04 (5%)
Stoma stenosis	01 (1%)

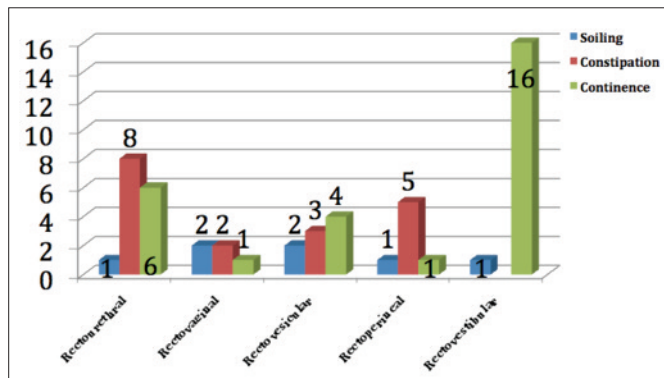


Figure 1: Follow-up according to Krickenbeck classification (age > 3 years, n = 52).

DISCUSSION

Anorectal malformations comprise a wide spectrum of defects ranging from slight malpositioning of the anus with excellent functional outcomes to complex anomalies of the hindgut and urogenital organs that are difficult to manage.⁴ Commonly known as imperforate anus, it affects 1 in 4000 to 5000 live births worldwide with a slight male predominance.⁵ The anomaly may occur in isolation but is commonly associated with other anomalies with incidence ranging from 40 - 60% in different series. The commonest of these are in the urinary tract (35%), the vertebral system (18%), and in the developing heart (10%). A known association of anomalies is known as the VACTERL group (vertebral, anorectal, cardiac, tracheoesophageal, renal and limb).⁶

Many classification systems for ARM have been devised over the years; the first of which was in 1970s which described low, high, intermediate and miscellaneous lesions for both genders based on the position of the terminal rectum to the levator ani. The same anatomic relationship formed the basis of the widely used 1984 Wingspread classification where the categories aforementioned were subdivided for males and females separately.^{7,8} A more surgically oriented classification was then proposed in 1995 by Pena as a result of his experience with posterior sagittal anorectoplasty (PSARP).⁴ This was based on the presence and position of fistula and on the relationship of the terminal colon to the levator sling muscles of the pelvic floor. The advantage of the classification of Pena was that the type of the fistula provided information not only about localization of the blind pouch but also on the anticipated extent of mobilization of the atretic rectal segment necessary to perform sacro perineal or abdomino-sacroperineal pull-through.^{9,10} This classification system was also the first one which attempted to determine prognosis for each group in terms of functional bowel outcomes.

With recognition of rarer anomalies not previously included in any classification and development of advanced surgical procedures other than PSARP, the

Krickenbeck International Classification emerged in 2005, which is based on consensus recommendations of world authorities.¹¹ This classification system is composed of 3 distinct elements: a diagnostic category, a surgical procedure category, and a category documenting functional outcome criteria. With the inclusion of all defects including rarer ones and surgical options, Krickenbeck classification aims to rationalize functional outcome among different clinical and surgical groups to allow more meaningful comparisons.¹²

Since the development of this system is fairly recent, there is paucity of literature on large long-term outcome studies using this classifications especially in our region. Long term functional outcome in children with ARM, primarily entails bowel function which is of vital importance as fecal incontinence and/or constipation remain major postoperative complications that impede social and psychological development of these patients.^{13,14} Continences, defined as the ability to initiate voluntary bowel movement with no soiling, regular bowel habits with no constipation, in turn defined as the passage of infrequent or hard stools, and overall quality of life, are the parameters looked at when assessing functional prognosis in such patients.¹⁵ In this study, there were 32/52 (62%) children who were continent. Mother's education is part and parcel of the better functional outcome as most of these children were toilet trained mainly because of mother's compliance to toilet training drills.

The Krickenbeck classification allows for international criteria for their treatment and development of a uniform international scoring system for comparable follow-ups. One of the first such studies was conducted in 2008 by Hassett *et al.* which evaluated the 10-year outcome of children born with ARM and treated by posterior sagittal anorectoplasty.³ This group of authors advocated with attempts to rationalize and demonstrate application of the Krickenbeck classification for both diagnosis and functional outcome in terms of constipation, urinary control and soiling.

Here, the researchers evaluated the follow-up of these children according to Krickenbeck classification and we found that continence, followed by constipation and fecal soiling, was the most common functional outcome in this study. Commonly, constipation occurs as a consequence of chronic dilatation of the rectal pouch due to failure to evacuate stool adequately. Mostly, it is seen in low fistulae.¹⁶⁻¹⁸ Soiling occurs because of defects in the sphincter mechanism or as a consequence of overflow from chronic constipation.¹⁹ Similar results were seen in this study as 9/18 (50%) children, who were constipated, had low anorectal anomalies.

Cardiovascular anomalies (38%) followed by urological anomalies (33%) were the most commonly associated anomalies in this study. Similar results were found in a

study from Singapore, in which they found similar associated anomalies (28% and 19%, respectively).²⁰

To the best of authors' understanding, there is no national study published which stratifies the functional outcome of this anomaly. This study is one of its own kind from Pakistan, addressing the need of uniform application of this classification for measuring functional outcome. The authors strongly suggest to make an anorectal malformation registry at national level so that we will come across the multicenter functional outcome of this anomaly for better understanding and management. Like all retrospective studies, there are the same limitations, as some time it was very difficult to recall although all possible measures were taken to double-confirm the findings by other observers. In addition to the parents' understanding about the functional outcome, there were variations of responses by mothers in terms of different grading of constipation.

CONCLUSION

Functional outcome of anorectal malformation is related to severity of disease. Children with low anorectal malformations usually have a good functional outcome; however, soiling is likely to be a long-term complication. There should be uniform approach to know the functional outcome which should be part and parcel for preoperative counselling to parents. A thorough evaluation of all infants with ARM should be done with particular focus on cardiovascular and genitourinary abnormalities.

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