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POST-OPERATIVE OUTCOMES OF PEADIATRIC PATIENTS WITH ANORECTAL MALFORMATION: A PROSPECTIVE STUDY

General surgery

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ABSTRACT

Introduction: Anorectal Malformations (ARM) represents a complex group of congenital anomalies resulting from the abnormal development of the hindgut, allantois and mullerian duct leading to incomplete or partial uro rectal septal malformations. ARM is a relatively common congenital cause of intestinal dysmorphology in the new-born. **Aims:** To find out and evaluate the outcomes of paediatric patients with anorectal malformations **Materials and Methods:** The present study was an Observational Study. This Study was conducted from 18 months at Department of General Surgery, AIMSR, Bathinda. Total 50 patients were included in this study. **Result:** We found that more number of patients had Abnormal anal opening [11 (45.8%)] in Fair Outcome compared to Good Outcome [6 (25.0%)] and Poor Outcome [1 (50.0%)] which was statistically significant (p<0.0001). More patients had [2 (100.0%)] L.ARM Anomaly in Poor Outcome compared to Good Outcome [15 (62.5%)] and Fair Outcome [10 (41.7%)] but this was not statistically significant (p=0.2286). **Conclusion:** We found that more number of patients had abnormal anal opening in Fair Outcome compared to Good Outcome and Poor Outcome which was statistically significant. More patients had L.ARM Anomaly in Poor Outcome compared to Good Outcome and Fair Outcome but this was not statistically significant.

KEYWORDS

Anorectal malformation, Follow up, Growth Intelligence, Motor function and development.

INTRODUCTION

Anorectal Malformations (ARM) represents a complex group of congenital anomalies resulting from the abnormal development of the hindgut, allantois and mullerian duct leading to incomplete or partial uro rectal septal malformations. ARM is a relatively common congenital cause of intestinal dysmorphology in the newborn. There are epidemiological differences in the level and extent of the abnormality. The spectrum of the lesions vary from fairly minor lesions like Covered anus, to some of the most complicated ones like Exstrophy and Rectal atresia. It is one of the most complicated defects to correct and forms the backbone of paediatric surgery.

ARM forms a significant load on the surgical services, particularly in the developing countries, not only in the emergency situations but also in terms of long term corrective procedures. Although there have been major advances in the management of these children in the last 15 years, these patients still represent a continuing challenge as a result of the significant reconstructive problems involved as well as the fact that a significant number suffer from faecal and urinary incontinence along with the possibility of inadequacies – sexual, non-correctable defects. With the development in the surgical specialities the management has improved and what was a certain disaster has now been converted to a normal livelihood and they now see patients who have married and borne children with a normal life span.

Anorectal malformation (ARM) is a common anomaly that results from defective development of the distal gastrointestinal tract affecting both sexes, the management of which is challenging [1]. There is a geographical variation in the incidence of ARM but it is reported as 1 in 5,000 live births in developed countries and more common in boys than girls. ARM is the commonest cause of intestinal obstruction in neonates [2] and the commonest congenital anomaly causing intestinal obstruction in the older children. Of the 151 cases of neonatal intestinal obstruction studied by Ameh and Chirdan, ARM accounted for 68.9% and 39.4% of 71 cases studied by Osifo and Okolo [3] in different parts of Nigeria. In the older children, it also accounted for 22.3% of 213 cases of intestinal obstruction studied by Ogundoyin et al. [4].

It is a multisystemic birth defect which affects boys and girls involving the distal part of the gastrointestinal and genitourinary systems. As a result of this association, patients suffering from anorectal malformation tend to have faecal and urinary incontinence. This problem may persist even after being attended to by a paediatric surgeon with consequent psychosocial effects on the children and their parents [5].

MATERIALS AND METHODS

The present study was carried out in the Department of General Surgery, AIMSR, Bathinda.

Patients were selected consecutively as and when they present to hospital after applying inclusion and exclusion criteria.

Duration: Study was carried out for a period of 18 months after getting approval from the Institution Research Committee(IRC), AIMSR and Institutional Ethics Committee(IEC), Adesh University .

Type of study: Observational.

Subjects: All cases in the Department of General Surgery, AIMSR, Bathinda in the period of 18 months as diagnosed cases of congenital anorectal malformation.

Place: Department of General Surgery, AIMSR, Bathinda.

Inclusion Criteria

- All cases of ARM admitted in AIMSR, Bathinda during the period from first presentation to completed all the stages of surgical procedures with in this period and within this period and with followup of 3 months

Exclusion Criteria

- Those cases who have not given consent for the surgery
- Those cases found to be unfit for surgeries due to severe associated anomalies

RESULT AND DISCUSSION

The present study was an Observational Study. This Study was conducted from 18 months at Department of General Surgery, AIMSR, Bathinda. Total 50 patients were included in this study.

In our study, out of 50 patients, most of the patients were [20 (40.0%)] ≤30 days of age. Age was not statistically significant with Outcome (p=0.7895). The mean age (in days) was higher [150.0000± 169.7056] in Poor Outcome compared to [137.0833± 122.3246] Fair Outcome and Good Outcome [103.6250± 118.3020] but this was not statistically significant (p=0.6030).

Pakarinen MP, et al [6] (2010) observed that low anorectal malformation comprises about half of all anorectal anomalies. Most of the literature concerning management of anorectal anomalies is centred around the treatment and outcome of high anomalies. The management of low anomalies has been considered significantly less challenging than high anomalies. Also, the outcome of low anomalies

has traditionally been considered good. However, recent more critical long-term follow-up reports show a different picture. Many patients with low anomalies suffer from long-term anorectal functional problems, especially constipation but also soiling that occurs in a significant percentage of patients. In this review, they compile the recent views on the diagnosis, surgical treatment and outcome of low anorectal anomalies.

We found that, male population [34 (68.0%)] was higher than the female population [16(32.0%)] but this was not statistically significant (p=0.8163).

Levitt MA, et al [7] (2010) found that Constipation in anorectal malformations (ARM) is extremely common, particularly in the lower types. Failure to adequately treat it can lead to significant morbidity. From their series of over 2000 patients with ARM, we reviewed 398 with good prognosis for bowel control and a tendency toward constipation; rectoperineal fistula (63), recto vestibular fistula (114), recto bulbar urethral fistula (104), imperforate anus with no fistula (46), rectal atresia or stenosis (9), and cloaca with a common channel below 3 cm (62). Those lost to follow-up, not yet toilet-trained (<3 years old), or with poor prognostic features were excluded. We compared morbidities in patients we operated on and managed primarily (group A, n = 268) to those managed at other institutions who suffered from constipation or incontinence and were referred to us for treatment (group B, n = 130). Those we managed primarily were subjected to an aggressive senna-based laxative program, started after their primary repair or after colostomy closure.

Rintala RJ, et al [8] (2010) suggested that anorectal malformations (ARMs) and Hirschsprung disease (HD) are the most common congenital colorectal defects in the new-born. The functional problems are more pronounced in patients with ARMs. Compared with healthy people, both patients with ARMs and those with HD have limitations in their quality of life. Inferior quality of life is more common in patients with ARMs. There is very little published data on long-term outcomes of adults with ARMs and HD. The effect of ageing on the functional outcome and quality of life remains unclear, although some preliminary data suggest that the bowel function and quality of life may deteriorate with ageing.

Our study showed that, more number of patients had Abnormal anal opening [11 (45.8%)] in Fair Outcome compared to Good Outcome [6 (25.0%)] and Poor Outcome [1 (50.0%)] which was statistically significant (p<0.0001). More patients had [2 (100.0%)] L.ARM Anomaly in Poor Outcome compared to Good Outcome [15 (62.5%)] and Fair Outcome [10 (41.7%)] but this was not statistically significant (p=0.2286).

Rangel SJ, et al [9] (2011) showed that the antegrade continence enema (ACE) has been shown to be a safe and effective method for managing faecal incontinence in the paediatric population. The purpose of this study was to examine their experience with the ACE procedure using the appendix as a catheterizable conduit in children with anorectal malformations (ARMs). They reviewed the charts of all patients who underwent an ACE procedure using the appendix as a catheterizable conduit between January 1992 and January 2010. Preoperative diagnosis (ARM type), operative details, functional outcomes, and postoperative complications were assessed. Technical modifications over time included selective cecoplication, implementation of the umbilical V-V appendicoplasty technique, and laparoscopy for cecal mobilisation. Mean age was 9.9 ± 0.6 years, and 67% were male. The most common preoperative diagnosis was rectourethral fistula in boys (39%) and persistent cloaca in girls (61%).

De Vos C, et al [10] (2011) Examined that laparoscopic-assisted anorectoplasty (LAARP) has gained popularity since its introduction in 2000. Further evidence is needed to compare its outcome with the gold standard of posterior sagittal anorectoplasty (PSARP).A retrospective review of patients presenting with anorectal malformation (ARM) in the period 2000 - 2009. Demographics, associated abnormalities, and operative and post-operative complications were assessed. The functional outcome in children older than 3 years was assessed, applying the Krickenbeck scoring system and, where possible, by interviewing parents. Patients with cloacal abnormalities were excluded. Patients with a LAARP were compared with those managed by PSARP. Results. Seventy-three patients with ARM were identified during the study period. Male to female ratio was 1.6:1. All 32 low ARMs (perineal and vestibular fistulae) were excluded. Thirty-nine had levator or supra-levator lesions. Twenty males presented with recto-bulbar, 3 with recto-prostatic, and 1 with a recto-vesical fistula; 2 had no fistula; and in 2 the data were insufficient to determine the level. Among the females, 6 had recto-vaginal fistulae, 4 had cloacas and 1 had an ARM without fistula. There were 3 syndromic ARMs (2 trisomy 21 and 1 Baller-Gerold syndrome).

It was found that, majority number of patients had [5 (100.0%)] RVF in Good Outcome compared to Poor Outcome [1 (100.0%)] and Fair Outcome [6 (42.9%)] it was statistically significant (p=0.0418) and we also showed that, most of the patients had [3 (100.0%)] no Associated Anomaly in Good Outcome compared to Poor Outcome [1 (1000.0%)] and Fair Outcome [3 (37.5%)] though it was not statistically significant (p=0.1070). Higher number of patients had [17 (77.3%)] Left Transverse Colostomy in Good Outcome compared to Fair Outcome [14 (58.3%)] and Poor Outcome [1 (50.0%)] which was statistically significant (p<0.0001).

Arnoldi R, et al [11] (2014) showed that the purpose of this study was to investigate the outcome of patients operated for anorectal malformations (ARMs) with good prognosis.Thirty patients underwent clinical evaluation by Rintala score and anorectal manometry recording anal resting pressure (ARP), rectoanal inhibitory reflex (RAIR), and rectal volume (RV).

We showed that Association of Complications with Outcome was not statistically significant (p=0.0632). Majority number of patients had [20 (100.0%)] PSARP in Definitive procedure in Good Outcome compared to Fair Outcome [14 (100.0%)]. Only 2 patients had Anal Stenosis in Fair Outcome which was statistically significant (p<0.0001).

De Blaauw I, et al [12] (2013) found that the European consortium on anorectal malformations (ARM-NET) was established to improve the health care of patients and to identify genetic and environmental risk factors. The aim of the present study was to present the first results on clinical data of a large European cohort of ARM patients based on theirregistry.In 2010, the registry was established including patient characteristics and data on diagnosis, surgical therapy, and outcome regarding complications. Patients born between 2007 and 2012 were retrospectively added. A descriptive analysis of this cohort was performed. Two hundred and three ARM patients were included. Syndromes or chromosomal abnormalities were present in 9%.

We observed that, higher number of patients had [24 (100.0%)] Dilation in Good Outcome compared to Fair Outcome [23 (95.8%)] though it was statistically significant (p<0.0001).

Borg HC et al [13] (2013) examined longitudinal follow-up of changes in bowel function in children with anorectal malformations (ARMs) with or without spinal cord pathology and neurogenic bladder dysfunction (NBD) as they grow. A successive improvement in functional outcome with age in children with ARM and normal spinal cord was seen with respect to continence, soiling and constipation. Continence was achieved earlier in girls than in boys (at 10 years: girls 80%, boys 36%). Soiling and constipation decreased with age both in grade and frequency (at 10 years low grade soiling: girls 53%, boys 64%). Boys with spinal cord malformation with NBD in combination with prostatic/bladder neck fistula (PRF/BNF) and sacral agenesis had the worst functional outcome with minimal possibility of improvement over time. Functional outcome in girls with NBD and tethered cord did not differ significantly from those without NBD and with a normal spinal cord.

In our study, the mean Weight was lower [2.4000± .8485] in Poor Outcome compared to [5.3750± 3.1645] Good Outcome and Fair Outcome [7.2833± 3.6617] but this was not statistically significant (p=0.0500) and we also found that, the mean Maternal Age was higher [29.1250± 3.4302] in Fair Outcome compared to [28.0000± 4.2426] Poor Outcome and [27.6667± 4.2495] Good Outcome but this was not statistically significant (p=0.4289).

CONCLUSION

1) Keeping in consideration with the research derivation, it can be concluded that the surgical treatment of Anorectal malformation has improved and diagnosis has become easier when recognizing the etiological factors and investigations. However it is important

to note that ARM outcomes can be affected due to its distribution of subtypes, associated anomalies, and mostly complications.

2) Based on our study of 50 patients we found that Males are commonly affected by this condition M:F 2.12 : 1 most patients presented within 4 days of birth.
3) It was found in the study High Anomalies are more frequent in males than females, clinical evidence differentiating low and high/intermediate are present in approximately 90% cases.
4) 40% of our patients had associated anomalies, Mortality is high in patients with high lesions as most of these patients are associated with serious associated anomalies. 78%of patients had good results. Associated Anomalies like cardiac , genitourinary , GI , Renal Anomalies associated with high type had poor outcome
5) Low Anomalies have a better outcome following surgery than the other variants; the only problem they faced was constipation in these patients.
6) Patients underwent surgical procedure according to the type of anomaly and were planned for staged procedure, High Anomalies required a colostomy at birth followed by Posterior sagittal anorectoplasty as definitive procedure followed by colostomy closure within 9-12 months of age.
7) The overall sensitivity of invertogram in detecting type of anomaly is low. Invertogram is done as a matter of routine but should not be taken as a fool proof investigation. Clinical determinants are the deciding norms.
8) Both the Abdomino perineal PSARP and PSARP procedures can successfully treat ARM with comparable outcomes.The functional results of posterior sagittal approach is better than traditional procedures for High and Intermediate anomalies.
9) The functional outcome of patients treated with low ARM is excellent.
10) The most common cause of death in ARM patients is Associated Anomalies.

Table: Distribution of Clinical Presentation

Number	Clinical Presentation	Frequency	Percent
1.	Absent anal opening, not passed meconium	26	52.0%
2.	Abnormal anal opening, Meconium passed	17	34.0%
3.	Passed meconium through urethral opening	4	8.0%
4.	Membranous anus, Not Passed Meconium	3	6.0%
	Total	50	100.0%

Table: Association between Associated Anomaly: Outcome

OUTCOME				
Associated Anomaly	Fair	Good	Poor	TOTAL
Hydronephrosis + VUR	3	1	2	6
Row %	50.0	50.0	0.0	100.0
Col %	37.5	100.0	0.0	50.0
B/L Undescended testis	1	0	0	1
Row %	100.0	0.0	0.0	100.0
Col %	12.5	0.0	0.0	8.3
Cardiac Anomalies	2	0	2	4
Row %	100.0	0.0	0.0	100.0
Col %	25.0	0.0	0.0	16.7
Down Syndrome	1	0	0	1
Row %	100.0	0.0	0.0	100.0
Col %	12.5	0.0	0.0	8.3
Esophageal atresia, TOF	0	0	1	1
Row %	0.0	0.0	100.0	100.0
Col %	0.0	0.0	100.0	8.3
TOTAL	8	1	5	13
Row %	66.7	25.0	8.3	100.0
Col %	100.0	100.0	100.0	100.0

Chi-square value: 15.7500**; p-value:** 0.1070

REFERENCES

1. Levitt MA,. Anorectal malformations. Orphanet J Rare Dis. 2007 Jul 26;2:33. [PMC free article] [PubMed] [Google Scholar] [Ref list]
2. Ameh EA, Chirdan B. Neonatal Intestinal Obstruction in Zaria, Nigeria. East Afr Med J. 2000;77(9):510–513. [PubMed] [Google Scholar] [Ref list]
3. Osifo DO, Okolo JC. Neonatal Intestinal Obstruction in Benin, Nigeria. Afr J Paediatr Surg. 2009 Jul-Dec;6(2):98–101. [PubMed] [Google Scholar] [Ref list]
4. Ogundoyin OO, Afolabi AO, Ogunlana DI, Lawal TA, Yifieyeh AC. Pattern and outcome of childhood intestinal obstruction at a Tertiary Hospital in Nigeria. Afr Health Sci. 2009 Sep;9(3):170–3. [PMC free article] [PubMed] [Google Scholar] [Ref list]
5. Taskinen S, Valanne L, Rintala R. The effect of spinal cord abnormalities on the function of the lower urinary tract in patients with anorectal abnormalities. J Urol. 2002 Sep;168(3):1147–9. [PubMed] [Google Scholar] [Ref list]
6. Pakarinen MP, Rintala RJ. Management and outcome of low anorectal malformations. Pediatric surgery international. 2010 Nov;26(11):1057-63.
7. Levitt MA, Kant A, Peña A. The morbidity of constipation in patients with anorectal malformations. Journal of pediatric surgery. 2010 Jun 1;45(6):1228-33.
8. Rintala RJ, Pakarinen MP. Outcome of anorectal malformations and Hirschsprung's disease beyond childhood. InSeminars in pediatric surgery 2010 May 1 (Vol. 19, No. 2, pp. 160-167). WB Saunders.
9. Rangel SJ, Lawal TA, Bischoff A, Chatoorgoon K, Louden E, Peña A, Levitt MA. The appendix as a conduit for antegrade continence enemas in patients with anorectal malformations: lessons learned from 163 cases treated over 18 years. Journal of Pediatric Surgery. 2011 Jun 1;46(6):1236-42..
10. De Vos C, Arnold M, Sidler D, Moore SW. A comparison of laparoscopic-assisted (LAARP) and posterior sagittal (PSARP) anorectoplasty in the outcome of intermediate and high anorectal malformations: gastro-intestinal. South African Journal of Surgery. 2011 Feb 1;49(1):39-43.
11. Arnoldi R, Macchini F, Gentilino V, Farris G, Morandi A, Brisighelli G, Leva E. Anorectal malformations with good prognosis: variables affecting the functional outcome. Journal of pediatric surgery. 2014 Aug 1;49(8):1232-6.
12. de Blaauw I, Wijers CH, Schmiedeke E, Holland-Cunz S, Gamba P, Marcelis CL, Reutter H, Aminoff D, Schipper M, Schwarzer N, Grasshoff-Derr S. First results of a European multi-center registry of patients with anorectal malformations. Journal of Pediatric Surgery. 2013 Dec 1;48(12):2530-5.
13. Borg HC, Holmdahl G, Gustavsson K, Doroszkiewicz M, Sillén U. Longitudinal study of bowel function in children with anorectal malformations. Journal of Pediatric Surgery. 2013 Mar 1;48(3):597-606.