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Anorectal malformations: Early outcome analysis in a tertiary care center in India

Priya Mathew,[1] Arun Kumar Gupta,[1] Rahul Gupta,[1*] Dharmesh Sharma,[2]

1 Department of Pediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India

2 Department of Preventive and Social Medicine, SMS Medical College, Jaipur, Rajasthan, India

Correspondence*: Dr. Rahul Gupta, MS, M.Ch. (Paediatric Surgery) FMAS, Associate Professor, Department of Paediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India. **E-mail**: meetsurgeon007@gmail.com

KEYWORDS

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Associated anomalies,
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ABSTRACT

Background: Delayed diagnosis, referral, and presence of associated anomalies may influence the outcome of Anorectal malformations (ARM). The aim of this study was to analyze the early outcomes (1 month postoperatively) of ARM presenting in the neonatal period.

Methods: A prospective study was carried out in our tertiary care teaching institute from December 2018 to March 2020. All neonates admitted in the NICU with ARM were studied.

Results: There were 315 neonates; out of these, 236 (74.92%) were male and 79 (25.07%) were female. High ARM (HARM) was present in 265 (84.13%) and low ARM (LARM) in 50 (15.87%). Associated congenital anomalies were noted in 121 (38.41%). Major associated anomalies consisted of gastrointestinal (GIT) (41.32%), oesophageal (31.4%), and genitourinary (GU) (19.83%). Out of 306 procedures for ARM, 196 (64.05%) neonates underwent left transverse colostomy (LTC). The most frequent postoperative complications were thrombocytopenia (115) followed by sepsis (98). Colostomy prolapse was uncommon (2). The overall mortality was 87/315 (27.61%) neonates - 82/265 (30.94%) in HARM and 5/50 (10%) in LARM. Neonatal mortality was significantly high with birth weight <2500gm 55/153 (35.94%; p value= 0.0001), associated malformations 82 (67.76%, p value= 0.003); and delayed presentation 40/87 (45%), and with primary perforation 5/6 (83.33%).

Conclusions: Higher mortality was associated with low birth weight, double/ triple atresia, neonatal GIT perforation, sepsis on admission, and those with oesophageal and cardiac anomalies. More than 1/3rd (38.41%) patients had associated anomalies; thus, a detailed systematic evaluation of all subtypes is paramount.

INTRODUCTION

Anorectal malformations (ARM's) are a diverse group of congenital anomalies encompassing the lower gastrointestinal tract, urinary, and /or genital system.[1,2] ARM has an incidence of 1:5000 live births.[1,2] According to Peña and Bechit, ARM has a varied presentation, ranging from low perineal fistula to high anomalies with complex surgical management.[1,3] It is frequently associated with anomalies affecting other organ systems referred to as the VACTERL association (5% to 31%).[4,5]

Complex associated anomalies, low birth weight, delay in access to pediatric surgeons, septicemia, gut perforation have an adverse impact on the prognosis. Risk factors such as hypothermia, hypoglycemia, poor immunity, and sepsis are modifiable and can lead to a reduction in neonatal mortality.[6] Timely diagnosis, management of associated anomalies, efficient surgical repair provide the best chance for a good functional outcome in patients.[7] We aimed to analyze the early outcomes (morbidity and mortality statistics) of a cohort of patients of ARM presenting in the neonatal period in a high-volume tertiary care teaching institute from the north-western region of India.

METHODS

This was a prospective study performed with IRB approval, to evaluate the early outcomes (1 month postoperatively) of neonatal ARM in our tertiary care teaching institute. The patients were from the state of Rajasthan and adjoining districts from neighboring

states in the region. The study period extended over a period of 16 months from December 2018 to March 2020.

Inclusion criteria:

All neonates with ARM admitted during this period were included in the study.

Exclusion criteria:

1. Patients with Cloacal Exstrophy, Syringomyelia, and Cloacal Regression Syndrome.
2. All the neonates underwent any surgical intervention at an outside hospital.

A thorough clinical evaluation of all the patients was performed. ARM subtypes were classified in accordance with the Krickenbeck classification.[8] Anomalies were categorized as cardiac, gastrointestinal, genitourinary, neurological, skeletal, Syndromic, and miscellaneous.

Radiological examination with Babygram and X-ray prone cross-table lateral view with raised pelvis was done after 18 to 24 hours of life in neonates without visible fistula. Abdominal ultrasound was done to detect any other abdominal pathology associated with ARM. Echocardiography was done in patients suspected to have cardiac anomalies. Routine use of ultrasound for urologic abnormalities and echocardiography was not considered due to resource limitations. A VACTERL association, first described in 1965, was recognized.[9] Esophageal atresia (EA) was classified according to the Gross anatomic classification.[10]

Preoperatively, all patients had intravenous fluids to correct fluid and electrolyte deficits, nasogastric suction, and broad-spectrum antibiotic coverage. Optimal control of hypothermia, hypoglycemia correction, and respiratory support was given. Nasogastric tube suctioning, intravenous fluids, and broad-spectrum antibiotics were continued in the postoperative period.

All neonates with LARM underwent primary anoplasty without a protective diverting colostomy. In HARM, the preferred colostomy was either the left transverse colostomy (LTC) or high sigmoid loop colostomy (SLC). Depending on the type of CPC, the neonates underwent fistula ligation with (a) pouchostomy (Type 1 and Type 2), or (b) excision of the pouch with end colostomy/end ileostomy. Patients were kept nil orally till the stoma started functioning or the child had passed meconium. Colostomy care was explained to the mother. Distal loop washes were performed in the postoperative period. In neonates with esophageal atresia, oral feeds were attempted only after a contrast study was done on the 7th postoperative day to rule out an anastomotic leak. The patients were followed up for 1 month postoperatively to analyze the early outcomes in our study.

The details were entered in the prescribed proforma and then into excel sheets. The clinical, operative records and other details of these patients were analyzed. Charts were reviewed. A “P” value of less than 0.05 was considered significant. All statistical data analysis was obtained with the statistical package for social sciences (SPSS) version 10.0 for Windows.

RESULTS

There were 315 neonates with ARM; 236 (74.92%) were males and 79 (25.08%) were females (M: F = 3:1). High-type ARM was seen in 265 (84.12%) patients, while low-type ARM in 50 (15.87%) neonates as shown in [Table 1]. High-type male ARM (with/without recto-urinary) was the most common variety with 183 (58.09%) neonates, followed by 47 (14.9%) males with low-type anomaly as shown in [Table 1]. The average birth weight was 2420 grams (range 930 to 3500 grams) [Table 2]. In our study, 171 (54.28%) neonates presented to the neonatal surgical ICU (NSICU) after 24 hours of birth (2 to 28 days), while 144 (45.71%) presented within 24 hours of birth.

Table 1: Type of ARM in the present study

Type of Anomaly	Sub-types	Frequency (Male + Female)	Percentages (%)
High Arm (HARM)		265 (189+76)	84.13
	HARM with/without Recto urinary (urethral/bladder) fistula (male)	183	58.09
	Persistent cloaca	21	6.67
	Vestibular fistula	27	8.57
	HARM with/without Rectovaginal (urinary) fistula (female)	24	7.61
	Rectal atresia	10 (6+4)	3.17
Low ARM (LARM)		50 (47 +3)	15.87
	Anteriorly placed anus/Anal stenosis/ Covered anus/ Bucket handle (male)	47	14.9
	Anteriorly placed anus/ Covered anus (female)	3	0.95
Total		315 (236 +79)	100

Associated malformations were documented in 121 (38.41%) neonates. The percentage of associated anomalies was more in males 94 (39.83%) than females 27 (34.17%). Associated malformations were present more in HARM 112/265 (42.26%) than LARM 9/50 (18%). Among 112 neonates with HARM, there

were 85 males and 27 females, while all 9 were males in the LARM subgroup [Table 3]. Three (0.95%) patients had triple atresia, i.e., esophageal atresia (EA), duodenal atresia (DA), and ARM. Two (0.63%) patients had ARM with duodenal atresia and 35 (11.11%) neonates had ARM with EA as shown in [Table 2]. CPC was found in 50 (15.87%) patients (Type 1 = 7, Type 2 = 13, Type 3 = 4, Type 4 = 24, Type 5 = 1, Type 6 =1).

Table 2: Predictors of neonatal surgical mortality in ARM patients [*Chi-square test].

Associated Malformations	**Frequency**	**Survival**	**Mortality**	**p value**
Gastrointestinal malformations	50 (41.32%)	10 (20%)	40 (80%)	p = 0.003*
Cardiac Malformations	22 (18.18%)	5 (22.72%)	17 (77.27%)	
Urogenital Malformations	24 (19.83%)	15 (62.5%)	9 (37.5%)	
Skeletal Malformations	14 (11.57%)	4 (28.57%)	10 (71.42%)	
DOWNS syndrome	3 (2.47%)	0 (0%)	3 (100%)	
Neurologic	2 (1.65%)	1 (50%)	1 (50%)	
Miscellaneous	6 (4.95%)	4 (66.66%)	2 (33.33%)	
Total	121	39 (32.23%)	82 (67.77%)	
Body Weight Range				
<1000gms (ELBW)	1 (0.31%)	0 (0%)	1 (100%)	p = 0.0001*
1000-1499gms (VLBW)	7 (2.22%)	1 (14.28%)	6 (85.71%)	
1500-2499gms (LBW)	145 (46.03%)	97 (66.89%)	48 (33.10%)	
>2500gms (Normal)	162 (51.42%)	130 (80.24%)	32 (19.75%)	
Total	315	228 (72.38%)	87 (27.61%)	
Type of atresia				
Triple atresia (EA+DA+ARM)	03	0 (0%)	03 (100%)	p = 0.933*
Double atresia *(a) EA+ARM* *(b) DA+ARM*	37 *35* *02*	06 (16.22%) *06* *0*	31 (83.78%) *29* *02*	
Total	40	06 (15%)	34 (85%)	
Rectal Atresia				
Rectal Atresia (Isolated)	9	3	6	P=0.645*
Rectal Atresia + Pure EA	1	0	1	
Total	10 (3.17%)	3 (30%)	7 (70%)	
Cloaca				
Cloaca (Isolated)	10	8	2	P=0.828*
Cloaca + Hydrometrocolpos / Vaginal Atresia	2	1	1	
Cloaca + CPC 1	3	2	1	
Cloaca + CPC 2	3	3	0	
Cloaca + CPC 3	2	2	0	
Cloaca + Umbilical Polyp	1	1	0	
Total	21 (6.66%)	17 (80.95%)	4 (19.05%)	
VF (Vestibular Fistula)				
V.F (Isolated)	20	19	1	P=.0001*
V.F + EA	5	1	4	
V.F + EA+D.A	1	0	1	
V.F + Dextrocardia	1	1	0	
Total	27 (8.57%)	21 (77.78%)	6 (22.22%)	
LARM				
LARM Isolated	43	40	3	P=0.009*
LARM + EA	3	3	0	
LARM +PS Hypospadias	1	1	0	
LARM + Skeletal abnormality	2	1	1	
LARM + Inguinal hernia	1	0	1	
Total	50 (15.87%)	45 (90%)	5 (10%)	
Primary/ Secondary perforation				
Primary	06	01 (16.66%)	05 (83.33%)	**P=0.543***
Secondary	06	03 (50%)	03 (50%)	
Total	12	04 (33.33%)	08 (66.66%)	

Ten (3.17%) patients had rectal atresia, out of which 1 neonate had rectal atresia with isolated esophageal atresia [Table 2]. Twenty-one (6.66%) neonates presented with a cloaca, out of which 2 neonates had associated Hydrometrocolpos, 8 had associated CPC as shown in [Table 2]. The vestibular fistula was the most common indication for admission with 27 (8.57%) neonates in the female group. Among these

27 neonates, 5 had associated esophageal atresia and 1 neonate had duodenal atresia with esophageal atresia [Table2].

Out of a total of 121 associated anomalies, gastrointestinal anomalies were identified as being the most common with 50/121 (41.32%) neonates. EA (38/121 (31.4%), Type C- 36 patients, Type A- 2 patients) was the most common individual anomaly associated with ARM in our patients; 5 patients also had duodenal atresia in this cohort. Thirty-five patients of HARM had EA while only 3 patients of LARM had it.

There were 23 (7.3%) patients with the VACTERL association. Some of the associated anomalies were, cardiac anomalies in 22/121(18.18%), urogenital anomalies in 24/121 (19.83%), skeletal in 14 (11.57%), and neurologic anomalies 2 (1.65%) as shown in [Table 3]. 2D Detailed summary of the survival percentages is shown in Table 2.

Table 3: Summary of associated malformations in the present study

Associated Malformations		**Type Frequency (n=121)**	**Subtype Frequency (n=121)**
Gastrointestinal malformations		50(41.32%)	
	EA Type C	*38 (31.4%)*	36 (72%)
	EA Type A		2 (4%)
	Duodenal Atresia		5 (10%)
	Malrotation		3 (6%)
	Meckel's Diverticulum		3 (6%)
	Duplication of Appendix + Caecum		1 (2%)
Cardiac Malformations		22(18.18%)	
	Septal Defects		16 (72.72%)
	Dextrocardia		1 (4.54%)
	Right Aortic Arch		3 (13.63%)
	Tetralogy Of Fallot		2 (9.09%)
Urogenital Malformations		24(19.83%)	
	Distal Hypospadias		4 (16.66%)
	Proximal Hypospadias		1 (4.17%)
	Undescended Testis		1 (4.17%)
	Scrotal Transposition		2 (8.33%)
	Hydronephrosis		5 (20.83%)
	Posterior urethral valve		2 (8.33%)
	Ureteral Duplication		1 (4.17%)
	Renal Agenesis		1 (4.17%)
	Vaginal Atresia		3 (12.5%)
	Bicornuate Uterus		2 (8.33%)
	Disorders of sex development		2 (8.33%)
Skeletal Malformations		14(11.57%)	
	Vertebral Defects		4 (28.57%)
	Multiple Limb Anomalies		9 (64.28%)
	Cleft Palate		1 (7.14%)
DOWNS syndrome		3 (2.47%)	
Neurologic	Lumbosacral Meningomyelocele	2 (1.65%)	
Miscellaneous		6 (4.95%)	
	Omphalocele Minor		1 (16.66%)
	Lumbar Hernia		1(16.66%)
	Inguinal Hernia		1(16.66%)
	Umbilical Polyp		1(16.66%)
	Cleft Lip		2 (33.33%)
Total		**38.41%**	

Among the 306 procedures done for ARM, left transverse loop colostomy was the most common (196) for the high-type anomaly. It was followed by posterior sagittal anoplasty in 45 neonates with low ARM. A total of 32 patients received end stoma in patients with CPC. Other procedures undertaken are summarised in [Table 4]. All procedures were performed between 2 to 48hrs following diagnosis. Nine patients died before any surgical intervention could be attempted.

Among the 44 procedures undertaken for associated anomalies, Fistula ligation and end-to-end esophageal anastomosis were the most common (25 neonates). A detailed summary of procedures for various associated malformations is shown in [Table 5].

Intraoperative surgical complications during repair of ARM included bleeding in 4 neonates. Six patients had pneumoperitoneum due to bowel perforation in the preoperative period and another 6 in the postoperative period. Two patients had distal jejunal perforation, and one each in the stomach, pouch, caecum, and stoma. The rest of the details are summarized in Table 6.

Table 4: Summary of procedures for ARM in the present study

Type	Procedure	Frequency (N=306)
High type anomaly	Left transverse loop colostomy	196
	Sigmoid loop colostomy	1
	Loop ileostomy	0
	Abdominoperineal pull through	5
High- type + Atypical Malrotation	Divided transverse colostomy	1
High type + Necrotising enterocolitis perforation	Divided ileostomy	1
Vestibular fistula	Evaluation- Conservative management	8
High Type +Congenital pouch colon (CPC)		
	Fistula ligation, excision of the pouch, end colostomy	32
	Fistula ligation, partial excision of pouch, pouchostomy	9
	Fistula ligation, excision of the pouch, end ileostomy	4
CPC + Gangrenous Caecum & Ascending Colon	Double exteriorisation of ileum, transverse colon	1
Persistent Cloaca	Right transverse loop colostomy	1
Low-type anomaly		
	Posterior sagittal anoplasty	45
	Cutback procedure	2
Not operated, Death		9
Total		**315**

Table 5: Summary of procedures for various associated malformations in the present study

Associated malformations	Procedures	Frequency
Esophageal atresia	Total	30
	Fistula ligation and end-to-end esophageal anastomosis (major leak – 2, minor leak -8)	25
	Fistula ligation, esophagostomy, and gastrostomy.	3
	Esophagostomy and gastrostomy	2
Duodenal atresia	Kimura's duodenoduodenostomy	2
Vaginal atresia with Hydrometrocolpos	Vaginostomy	3
	Suprapubic catheter insertion into the urinary bladder	2
Meningomyelocele	Repair of MMC	1
Malrotation	Ladd's procedure	2
Meckel's diverticulum	Wedge resection	2
Omphalocele minor	Repair and purse-string umbilicoplasty	1
Umbilical polyp	Polypectomy	1
Total		**44**

Postoperative complications were thrombocytopenia (115), sepsis (98), pneumonitis/pneumonia (30), esophageal anastomotic leak (10; major leak- 2, minor leak- 8), wound infection (5), colostomy prolapse (2), colostomy retraction (1), and colostomy stenosis (1).

Overall, there were 87 (27.61%) deaths and 228 (72.38%) survivors. Mortality in neonates with a birth weight below the normal (2500 gm) was 55/153 (35.94%); among this group, mortality rates with extremely low birth weight (ELBW) was 1/1 (100%), very low birth weight (VLBW) was 6/7 (85.71%), and low birth weight (LBW) was 48/145 (33.10%). The mortality outcomes were statistically significant (p value= 0.0001) with low birth weight as shown in Table 2.

Neonatal mortality in High-type ARM were 82/265 (30.94%) and in Low type ARM were 5/50 (10%). Neonatal mortality in CPC was 13/50 (26%), rectal atresia 7/10 (70%), vestibular fistula 6/27 (22.22%), and cloaca 4/21 (19.05%). Isolated vestibular fistula

(p value= 0.001) and LARM (p value= 0.009) had lower mortality rates. Neonatal mortality with associated malformations was 82/121 (67.76%) and was found to be statistically significant (p value= 0.003). A detailed summary is shown in Table 2. Out of all the deaths, 40 (45%) neonates had presented to our NSICU, beyond 24 hours of birth. Poor prognosis was present in patients presenting late with perforation 8/12 (66.67%), especially those with primary perforation 5/6 (83.33%).

Table 6: Summary of gastrointestinal perforation in neonates with ARM, surgical intervention, and outcome. (LTC – Left transverse colostomy)

Type of ARM	Site of perforation	Primary/Secondary	Type of repair	Outcome	Weight (grams)
High type	Sigmoid	Primary	Perforation repair & LTC	Death	1800
	Sigmoid	Primary	Perforation repair & LTC	Death	2500
	Ileal	Primary (NEC)	Divided ileostomy with the release of adhesions	Death	2600
	CPC 4, perforated pouch	Primary	Fistula ligation, excision of CPC, sigmoid colostomy.	Death	2500
	CPC4- gangrenous caecum, ascending colon	Primary	Resection of gangrenous segment, double exteriorisation of ileum, transverse colon.	Death	2600
	CPC 1 + perforated pouch	Primary	Fistula ligation, excision of CPC, and ileostomy.	Discharge	2000
	Caecal	Secondary	Perforation repair	Death	2600
	Gastric	Secondary to LTC	Gastric perforation repair + Anterior gastrostomy	Death	2600
	LTC- Stoma perforation	Secondary to LTC	Proximal end colostomy+ distal Hartmann pouch	Discharge	2300
	Distal jejunum	Secondary to LTC	Repair of perforation, Omental patch + drain placement	Death	2400
	Distal jejunum	Secondary to LTC	Divided colostomy + jejunal perforation repair	Discharge	1600
	CPC 2 with perforation of gangrenous pouch	Secondary to Pouchostomy	1. Pouchostomy 2. Excision of gangrenous pouch and end colostomy.	Discharge	3000
Total	n=12	Primary= 6 Secondary=6		Death=8 Discharge = 4	

DISCUSSION

ARMs form a diverse group of congenital malformation ranging from minor, easily treated defects like anal stenosis that have an excellent functional prognosis to imperforate anus with a recto-urethral fistula (most common defect in males) to complex defects e.g., persistent cloaca that is difficult to manage. [1,3,11,12] We managed an average of 236 neonates annually and approximately 20 neonates monthly in our resource-limited setup.

The higher no. of HARM (84.13%) in our study contrast with a recent large study from the Netherland in which more than half of the patients constituted low-type defects.[13]

In our study, the mortality rate was higher in neonates 47/144 (32.64%) presenting within 24 hours of birth than those presenting late 40/171 (23.39%). The higher mortality rate among the former group was due to the presence of associated esophageal atresia and cardiac malformations. Significant mortality figures in the latter group could be attributed to delayed presentation. Delay in the presentation of patients with ARM leads to the progression of neonatal intestinal obstruction, sepsis, aspiration pneumonia, intestinal perforation, and sometimes death. [14-18] Delayed presentation is common among female neonates because there is still some decompression through the vestibular fistula in most instances, unlike in males where abdominal distension occurs over a few days in most patients with a recto-urethral fistula. [19, 20] Urosepsis with septicemia is also more likely in males.[21]

The low birth weight (LBW) neonates remain at a much higher risk of mortality than the infants with normal weight at birth. It has been found that neonatal mortality has an inverse relationship with birth weight.[22] The same was observed in our study [Table 2].

In our study, the most common (8.57%) defect in females was the vestibular fistula (VF), which is according to most series.[1]. At our institute, for a neonate with VF, the criteria for admission to NSICU is subacute obstruction due to the non-passage of meconium or life-threatening associated anomalies which needed urgent surgical intervention.

A persistent cloaca is characterized by the fusion of the rectum, vagina, and urinary tract to form a single common channel. Hydrocolpos is characterized by an expanded fluid-filled vaginal cavity; present in half of the patients with persistent cloaca.[1] In our study, persistent cloaca was the second most common (6.67%) malformation. Hydrocolpos was noted in 3 cases, for which tube vaginostomy was done. In 2 patients, a suprapubic catheter was inserted into the urinary bladder. Urologic assessment is important in persistent cloaca due to the high percentage of associated defects.[1,23] CPC was present in 38.09% (8/21) neonates with persistent cloaca.

Rectal atresia is a rare entity. In our study, 10/315 (3.17%) of neonates had rectal atresia with 6 males and 4 females; LBW was present in 6/10 (60%) patients. The mortality was high 7/10 (70%) in this cohort. Only 1/10 (10%) neonate with associated EA presented <24 hours of birth, while the rest 9/10 (90%) presented late- >24 hours (2-8 days). The former neonate had an unfavorable outcome due to associated malformation, LBW, and postoperative sepsis. Among the LBW 6/10 (60%) neonates, only 1/6 (16.67%) had a favorable outcome, while in the group 4/10 (40%) with normal birth weight, 2/4 (50%) had a favorable outcome. The presentation of RA was delayed in our series owing to the presence of a normal anus, and delayed referral. Meticulous clinical evaluation, plain abdominal radiograph, and urine examination for meconuria may help in detecting this rare association early. The senior author (RG) recommends gentle insertion of a 6Fr infant feeding tube or a soft red rubber catheter per anus in all patients with delayed passage of meconium >24 hours to rule out RA.

CPC is a subgroup of congenital anomaly in ARM in which, whole or part of the colon is replaced by a pouch-like dilatation that communicates distally with the urogenital tract by means of a fistula. [24] It is included in “Rare and regional variants” as per Krickenbeck Classification. The incidence of CPC among ARM cases has been reported to range from 2% to 18%; it was 15.87% in our series. A high incidence of 30-40% has also been reported from the Indian sub-continent. [24] Traditionally; it has been divided into 4 subtypes, as per the length of colon involved.[24] Type 4 was the most common (24/50) subtype in our study. Type 5 CPC is a rare form of CPC, i.e. two colonic pouches with intervening normal colon.[25] It was seen in only one patient in the present series. Type 6 CPC which is an extremely rare and recently described subtype was seen in one patient.[26]

X-ray prone cross-table lateral view with the raised pelvis (Prone cross-lateral film) is performed when clinical signs do not reveal (define) the type of ARM in 18-24 hours. The babies are kept in the genu-pectoral position for 3 minutes by holding their face down with hips flexed. Prone lateral radiographs are obtained. It was performed to see the level of distal gas shadow in 81.90% (258/315) of our patients. In previous studies, it was performed in 10-20% of neonates. [12,27,28] In our study, all the neonates were advised “Babygram” to diagnose associated intestinal atresias, congenital pouch colon (CPC) and its type, other associated anomalies, and to confirm the diagnosis of bowel perforation (late presentation).[29]

In the present study, 38.41% of patients had associated anomalies, while the reported frequency ranged between 50-65%.[30,31] Neonates with HARM have a higher incidence of associated anomalies than LARM. Urogenital anomalies are the most common and among them, VUR is the most common (one-third) associated genitourinary anomaly. [11,12,32] Cardiac anomalies were detected in 18.18% of neonates in our case are less frequent than the reported frequency (1/3rd patients).[33] In the present series, the detection of cardiac anomalies was three times the previous study.[30] This was due to better utilization of Echocardiography in neonates with ARM.

GIT anomalies were detected in 15.87% (50/315) in the present study, while it was 20% and 27.31% (59/216) in recent studies. [30,34] They were the most common 41.32% (50/121) anomalies among associated malformations in our study. There is an increasing incidence of EA in our geographical area and our center is managing a high volume of neonates with EA.[35,36]

In LARM, posterior sagittal anoplasty is the preferred surgical intervention. In HARM, a colostomy is a preferred choice in the neonatal period. Most pediatric surgeons prefer a protective colostomy before definitive surgery.[37] In HARM, two types of colostomies can be performed: divided and loop colostomy. In our series, for patients with HARM, without radiological evidence of CPC, a transverse loop was exteriorized in the left upper abdomen in 196/306 (64%). In patients with suspicion of CPC or plain radiograph suggestive of CPC, a left hockey stick incision was given to deal with the pouch, fistula ligation, and perform end stoma as per the operative findings. A supra-umbilical right transverse incision was given, if there were signs of peritonitis or associated GI malformation or atypical anatomy or persistent cloaca with Hydrometrocolpos. The most used sites for stoma creation are high sigmoid/descending colon followed by left transverse colostomy.[34,38] We prefer (64.04%) left transverse loop colostomy at our center. The advantages of this

procedure are that it is easier to perform, smaller skin incision, less operative time, and fewer intraoperative complications. Stoma care is easier than in sigmoid colostomy. The disadvantage with left transverse colostomy is that it carries a high risk of prolapse, as the transverse colon is more mobile than the sigmoid colon. [30,34,37,38] In our study, only 2 neonates who underwent left transverse colostomy were noted to have a prolapsed stoma. In both cases, the prolapse was reducible and did not necessitate colostomy revision. Prolapse can be avoided with a small fascial defect, applying seromuscular sutures between the proximal and distal loops and fastidious technique of anchoring the loops to the incised fascia.[39,40]

The standard procedure for the management of the CPC is a three-staged procedure as single-stage management is associated with increased mortality.[41] In our institute, neonates with CPC are managed as per the Saxena-Mathur classification.[42] Depending on the type of CPC, 32 neonates underwent fistula ligation, excision of the pouch, and end colostomy; 9 had fistula ligation, partial excision of the pouch, and pouchostomy, and 4 neonates underwent fistula ligation, excision of the pouch, and end ileostomy.

In our study, mortality was found to be quite high 87/315 (27.61%) as compared to Western studies (6%).[34] The higher mortality rate was found in neonates with HARM 82/265 (30.94%), birth weight below 2500 gm 55/153 (35.94%), late presentation (to our NSICU) beyond 24 hours of birth 40/87 (45%), and associated malformations 82/121 (67.76%).

Among various variables, the worst prognosis was seen with low birth weight (p value= 0.0001) followed by associated malformations (p value= 0.003). Isolated vestibular fistula (p value= 0.0001) and LARM without associated malformations (p value= 0.009) had good prognosis.

The reasons for very high mortality in our setup were delayed detection (lack of trained workforce and limited resources in peripheral health centers). Mortality in our present series (27.61%) was better than a similar study conducted in our institute in 2016 (31.02%).[30] This was due to an increase in infrastructure facilities, relatively lesser overcrowding in NSICU, increase manpower, and improvement in neonatal care and resources.

The most common cause of mortality was septicemia (78), followed by associated cardiac malformations (21), pneumonia (19), and esophageal anastomotic leak (2). Morbidity and mortality in our series were not directly related to the type of stoma created. This has been reported in a recent series.[34] Ours being a high-volume center, due to space constraints, there is overcrowding in NSICU. Despite being diligent about infection control, there are high chances of cross-infection and sepsis.

CONCLUSION

Anorectal malformations are common in our setting with the high type being more frequent. The routinely done surgical intervention was left transverse colostomy for HARM. More than 1/3rd (38.41%) of patients with ARM had associated anomalies. With an increasing number of organ systems involved, the survival rate decreases. Higher mortality was associated with low birth weight, double/ triple atresia, neonatal GIT perforation, sepsis on admission, and those with esophageal and cardiac anomalies in our study. Hence, a detailed systematic evaluation of all neonates with ARM (both high and low type) should be done, irrespective of its type. Although rare, hollow viscus perforation in ARM should always be suspected in delayed ARM presenters.

A strategy to reduce neonatal mortality and morbidity by initial resuscitation, timely referral, strict infection control measures, adequate nutritional support and maintenance of optimal physiologic status, improvement in nursing care, up-gradation of infrastructure facilities, and availability of bedside echocardiography and ultrasonography is recommended. A multidisciplinary approach with neonatologists, pediatric surgeons, anesthesiologists, and radiologists would greatly enable the highest possible standards of care.

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