

Original Article



Surgical Outcome of Anorectal Malformations of Neonates in Dhaka Medical College Hospital

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Abstract

Background: 'Imperforate anus' has been a well-known condition since antiquity. Nowadays, it is broadly termed as anorectal malformations (ARMs).

Objective: To determine the incidence, initial management and surgical outcome in children with ARMs.

Materials and Methods: This prospective descriptive study included neonates admitted with ARMs in Dhaka Medical College Hospital during 24-months period (January 2015 – December 2016). A total of 118 neonates were admitted (23.6 % of the total neonatal surgical admission) during this period.

Results: The majority of the ARMs neonates were presented at the first week of life 109 (92.37%). Gender distribution of neonates: males were 80, females were 38, ratio (m: f) was 2.11: 1, age at presentation of neonates ranged from 0.04 day (1 hour) to 28 days (mean 3.27 days), presenting weight of neonates ranging from 1.5 - 5 in kilograms (mean 2.67 kg). The majority of patients were a high variety of ARMs 80 (67.79%), low variety of ARMs 27 (22.88%) and cloaca 11 (9.32%). Most of the surgical procedures were pelvic colostomy 80 (21.1%) followed by anoplasty 27 (7.0%), and transverse colostomy 11(9.32%). 16 neonates died, so mortality was 13.56%. Mortality was higher in low-birth-weight babies: 68.75%. Premature/preterm (short gestational age) delivery 56.25%. 14 (17.5%) after pelvic colostomy and 2 (18.18%) died after transverse colostomy.

Conclusion: An effective healthcare system, proper training for medical professionals and meticulous perineal examination of all neonates at birth are key requirements of early detection of ARMs. Early presentation of surgical neonates ensures better surgical outcomes.

Keywords: Anorectal Malformations, Neonatal Intestinal Obstruction, Preterm, Surgical Outcome.

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Introduction

Anorectal malformations (ARMs) are one of the significant spectra of structural congenital defects involving the anal orifice, anal canal and rectum with an incidence of 1 per 2000 - 5000 live births.^{1,2} The catastrophe happens as a result of any arrest or insults during early development of cloaca and urorectal division.³ Mostly, children are diagnosed postnatally by careful examination of perineum, that includes DRE (Digital Rectal Examination) and radiological studies within 24 hours of birth.⁴ ARMs may present as an isolated anomaly, in association with other congenital anomalies (VACTERL meaning vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities) or as part of Down syndrome (a genetic condition caused when an unusual cell

division results in an extra full or partial copy of chromosome 21).^{5,6} Previously the used classified terms 'low', 'intermediate' and 'high' are not now found much useful in current therapeutic or prognostic terminology. Since 2005, Krickbeck's classification is widely used not only in establishing the diagnosis of diverse types of ARMs but also for evaluation of surgical outcome.⁷

ARMs are the leading congenital causes of intestinal obstruction in neonates.^{1,8} The treatment options, often, are influenced by factors related to the clinical presentation and available of perioperative hospital facilities, especially for complex congenital malformations and expertise of surgeons. Early management is recommended in the treatment of children with ARMs in order to prevent sepsis and other morbidities related to intestinal

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obstruction.⁸ The aim of present study was to evaluate the presentation, procedures performed in relation to the type of anomaly and early outcome analysis in the neonatal period at our high-volume tertiary care institute.

‘Delayed presentation’ where anal opening has not been noticed/identified before discharge and where the diagnosis is delayed until abdominal distension, poor feeding or vomiting alerts the mother or healthcare staff.⁹ Many of these neonates are brought to the hospital several days or sometimes weeks after the onset of illness with severe fluid and electrolytes deficits, malnutrition, anemia, and sepsis. These are late signs of anorectal malformations. Besides, surgical intervention in hemodynamically unstable and hypothermic babies often leads to high per- and postoperative morbidity and mortality.^{2, 8} This potentially life-threatening scenario is avoidable. All cases of anorectal malformations should be diagnosed within 24 hours of birth. In the present study, ‘early diagnosis’ is defined as the presentation of neonates within 24 hours of birth that are submitted to immediate hospital care.⁹

Birth weight is one of the most important determinants of neonatal survival.¹⁰ LBW (Low birth weight) babies constitute only about 14% of children born, and they account for 60%–80% of neonatal deaths. LBW arises through short gestation (preterm birth) or in-utero growth restriction, or both. Here, early neonatal deaths account for 75% of all neonatal deaths, and preventing these depends on attention to the causes of death that are unique to the first week of life, particularly birth asphyxia and prematurity. Many neonatal deaths take place within the first 24 hours after birth, at least more than 1 million per year globally. Late neonatal deaths are mainly due to infections.¹¹ It has been estimated that almost half of infant deaths in Bangladesh from diarrhea or pneumonia could be prevented if LBW was eliminated.¹²

Materials and Methods

All neonates with ARMs admitted to the Department of Pediatric Surgery, Dhaka Medical College & Hospital, over a period of 2 years from January’ 2015 to December’ 2016 were studied prospectively. A detailed history and careful physical examination were completed. An invert gram (cross-table lateral x-ray was not available in our setting) was performed in the examination did not reveal the type of ARM. In female patients with cloacal malformation/urethral fistula in whom the diagnosis was not established by perineal examination, a babygram was performed. Ultrasonography of the abdomen was performed to evaluate for cloacal malformation. If needed echocardiography also was performed for evaluation of cardiac abnormality. Intravenous fluids, nasogastric tube placement, and antibiotics were continued postoperatively. Gradual feeds were initiated after the postoperative passage of meconium or when the colostomy started functioning.

Patient’s age at presentation, sex, clinical presentation, preoperative investigations undertaken for establishment of diagnosis, associated malformations, intraoperative findings (operative records), postoperative complications, and treatment outcomes were analyzed and charts were prepared.

Exclusion criteria included

- i. Male or female ARM patients presenting beyond the neonatal period.
- ii. Patients with cloacal exstrophy, sirenomelia syndrome (also known as mermaid syndrome, a congenital anomaly with complete or partial fusion of lower limbs), and caudal regression syndrome (featured by the classical triad of presacral mass, sacral bone defect and anorectal malformation).
- iii. Patients presenting with colostomy or other operative interventions performed outside the institution.
- iv. Vestibular fistula is important variety of ARMs of female, treated by per rectal normal saline irrigation in the outpatient department and definitive surgery (ASARP) done at the age of 6 months.

Results

The majority of the neonates with ARMs presented during the first week of life [109 (92.37%)]. Gender distribution of neonates: male were 80, females were 38, ratio (m: f) was 2.11: 1, and age at presentation ranged from 0.04 days (1 hour) to 28 days (mean 3.27 days). (Table I)

Table I: Demographic distribution

Presenting time in weeks	Male (%)	Female (%)	Total (%)
1 st week	75 (63.56)	34 (28.81)	109 (92.37)
2 nd week	2 (1.69)	4 (3.39)	6 (5.08)
3 rd week	0	0	0
4 th week	3 (2.54)	0	3 (2.54)
Total	80 (67.79)	38 (32.20)	118 (100)

Table II: Birth history and weight pattern of neonates

Neonatal weight (kg)	Preterm (%)	Term (%)	Total (%)
< 2.5	24 (20.34)	7 (5.93)	31 (26.27)
>2.5	0	87 (73.73)	87 (73.73)
Total	27 (20.34)	94 (79.66)	118 (100)

87 (73.73%) of neonates presented at term delivery, all were having more than 2.5kg body weight. 31 (26.27%) of neonates presented with less than 2.5kg body weight. Among them, 24 (20.34%) were having preterm delivery and 7 (5.93%) having term delivery. (Table II)

Table III: Type of surgical procedure of ARMs

Type of ARM	Surgical procedure	Number (%)	Mortality (%)
Low variety	Anoplasty	27 (22.88)	0
High Variety	Pelvic colostomy	80 (67.79)	14 (17.50)
Cloaca	Transverse colostomy	11 (9.32)	2 (18.18)
Total		118 (100)	16 (13.56)

27 (22.88%) of neonates with low variety ARM for which anoplasty was done. There was no mortality. 80 (67.79%) high variety ARM for that pelvic colostomy was done. Among them, 14 newborn (17.50%) died after surgical procedure. Persistent cloaca is a type of high variety ARM. In this study, for 11 (9.32%) cloaca anomaly neonates transverse colostomy was done, where 2 (18.18%) neonates died following the surgical procedure. Thus, total mortality was 16 (13.56%). (Table III)

Table IV: Mortality related to birth history and weight.

Surgical outcome	Presenting time	Birth history		Weight (kg)	
		Preterm	Term	< 2.5 kg	>2.5 kg
Death	1 st week (14)	8 (50%)	6 (37.50%)	9 (56.25%)	5 (31.25%)
	2 nd week (2)	1 (6.25%)	1 (6.25%)	2 (12.50%)	0
Total		9 (56.25%)	7 (43.75%)	11 (68.75%)	5 (31.25%)

Mortality related to birth history and weight was assessed. It was shown that 9 (56.25%) mortality in preterm delivery. Among them, 8 (50%) were during the 1st week, and 1 (6.25%) were during the 2nd week. 11 (68.75%) mortality were documented in LBW baby (weighing less than 2.5 kg). Among them, 9 (56.25%) were during the 1st week and 2 (12.50%) were during the 2nd week. (Table IV)

Table V: Outcome of ARM with age at presentation (during the 1st week)

Presentation	Good	Dead	Total
Early (0-1 day)	33 (94.29%)	2 (5.71%)	35 (32.11%)
Late (2-7 days)	62 (83.78%)	12 (16.22%)	74 (67.89%)
Total	95 (87.16%)	14 (12.84%)	109 (100%)

To test the hypothesis, outcome was assessed with early and late presentation in cases of ARMs. It was observed that 16.22% of late presentation cases died, while only 5.71% died in cases of early presentation cases. It revealed that early presentation of neonates had more satisfactory outcome than for late presentation cases. (Table V)

Table VI: Odd ratio between early and late presentation of ARM during the 1st week

Presentation	Dead	Good	Total
Late (2 - 7 days)	12	62	74
Early (0 - 1day)	2	33	35
Total	14	95	109

Odd ratio = Probability of an event to occur ÷ probability of an event not to occur

$$= a/b \div c/d$$

$$= 12/62 \div 2/33$$

$$= 0.193 \div 0.06$$

$$= 3.22$$

To test the hypothesis, the outcome was assessed with the presentation by odd ratio (OR). We observed that the OR of late presentation was 3.22. It meant that the late presentation of neonates was a higher risk factor indicating worse surgical outcomes. (Table VI)

Discussion

The most common causes of neonatal intestinal obstructions are ARMs. Prenatal diagnosis of ARMs is not always possible. So, careful examination of the perineum at birth within 24 hours is crucial to know the type of ARMs.^{13, 14} Male and female ratio was 2.11: 1. One study reveals male and female ratio was 3: 12 while other study shows male and female ratio was 4: 1.¹⁵ Boys were the predominant group in our study, similar finding was noted in other studies.^{7, 16-17} Majority of neonates had normal birth weight (mean 2.65 kg ± standard deviation 0.5), similar

finding is seen in other studies.^{1,2}

The majority of neonates presented during the first week of life 109 (92.37%). 35 (29.66%) cases were presented before 24 hours (early presenter), and total 83 (70.33%) cases between 24 hours (day 1) to 28 days (late presenter). So, the majority of neonates were found as late presenters in our study. 'Delayed presentation' is defined variably in different articles. Some articles are defined as more than 24 hours, some other articles as more than 2-3 days,^{9, 8, 14, 17, 18} and some other articles as more than 7 days, or even months later.^{6, 19}

In this study, 31 (26.27%) cases were LBW babies, among them preterm babies (<37 weeks gestational age) were 20.34%. One other study shows preterm babies 10%.^{6, 16} Anoplasty was done for low variety ARM in 27 (22.88%) babies. The majority of the neonates accounting 91(77.12%) were of high variety ARMs. This finding is similar to another study.¹⁵ But, in some other study, it had been shown that anal stenosis is the major diagnosis accounting to about 88.89%.⁷ Among them, 80 (67.80%) cases were treated by pelvic colostomy, and transverse colostomy was performed for persistent cloaca in 11 (9.32%) cases. In our study, persistent cloaca was present in 9.32% of patients. In another European large study over 20 years period, incidence is shown as 6.07%.¹⁵

Early surgical outcomes during the neonatal period were good about 102 (86.44%). Mortality was about 16 (13.56%). Higher mortality was observed in other studies.^{2, 15} Significantly, more deaths occurred in preterm babies and low birth weight babies. Mortality was higher in preterm babies because of the immaturity of all physiologic functions. Mortality was high among children aged more than 24 hours (late presenter) in this study. In another study was shown that age at presentation did not affect the outcome of surgery but, small sample size and maximum age at presentation was 3 days.²⁰ The other studies were shown that delayed presentation was associated with poor surgical outcome and also increased mortality and morbidity.^{1, 2, 6} Higher mortality was observed in early presenters in this survey probably because most of them were underweight, preterm with multiple associated anomalies, this was similar to other studies.^{2, 15}

The disease process of neonates with ARMs varies day to day and the disease process starts ante-natally, but is most often diagnosed after birth. In the present study, 16.22 % death occurred in late presenters while only 5.71% death occurred in early presenters. Odd Ratio (OR) of late presentation was 3.22. It was found that delayed presentation of neonates was a higher risk factor for poor surgical outcomes.

Conclusion

The outcome following surgical correction of ARMs depends on the respective/specific type and the age at presentation. Early diagnosis and referral to pediatric centers, improvement of Neonatal Intensive Care Centers (NICUs), treatment of associated anomalies in other systems and prevention of sepsis all will help reduce morbidity and mortality. Delayed presentation is definitely a risk factor for poor surgical outcomes.

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