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Original Article

Factors affecting the outcome of neonates with anorectal malformation in a developing country

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ABSTRACT

Background: The survival and outcome of neonates with anorectal malformations (ARM) have much improved in the developed countries due to optimal perioperative and postoperative care but in developing countries, sepsis, low birth weight, delayed presentation, and lack of intensive care for neonates are still important in affecting the outcome. This study was carried out to evaluate factors of poor outcome (mortality) in neonates with ARM.

Methods: This is a prospective analytical study. A total of 44 consecutive neonates with Anorectal malformations (ARM) presenting to the Department of Pediatric Surgery, The Children's Hospital, Pakistan Institute of Medical Sciences, Islamabad, were included. Variables studied included age at presentation, gender, birth weight, type of malformation, sepsis at presentation, type of surgery performed, postoperative complications, and their relationship to the outcome. The statistical analysis was performed using SPSS version 21.

Results: A total of 44 neonates with ARM were included in the study. In the study population, 56.8 % (25) were males and 43.2% (19) were females. The mean age at presentation was 2.1 ± 0.5 days. The mean birth weight was 2.5 ± 0.6 kg. Overall mortality was 29.5% (13) with 13.63% (6) patients died pre-operatively. The most common cause of death in postoperative patients was sepsis (40%). There was a statistically significant relationship between low birth weight (P= <0.01) and sepsis at presentation (P=0.001) with mortality. No statistically significant association was found when the outcome was compared with age at presentation (P=0.21) and postoperative complications (P=0.16).

Conclusion: In developing countries, the lack of resources, lack of trained midwives/LHVs, intensive care are contributing factors to sepsis and delayed presentation, and ultimately mortality. Good antenatal care, awareness of the midwives/Lady Health Visitors to refer such patients in time, and provision of adequate intensive care can improve the outcome of surgery in ARMs.

Keywords: Imperforate anus, Sepsis, Low birth weight, Primary PSARP, Pelvic divided colostomy, Cloaca

INTRODUCTION

Anorectal Malformations (ARM) is considered as one of the most common congenital intestinal anomalies with an incidence of 1 in 4000 to 5000 births, affecting males slightly more than females.[1] ARM has a wide spectrum of disease presentation, from minor defects to complex malformation such as persistent cloaca and from isolated anomaly to associations with urogenital, cardiac, esophageal atresia, gastrointestinal, vertebral, and limb anom-alies.[2,3]

The mortality rate for ARM was high in the past. Alt-hough it was unsuccessful, the first surgical attempt was made in 1783 by performing an inguinal colostomy.

The outcome gradually started getting better with improvements in understanding the nature of the defect and with the emergence of new management modalities. The major determinant of poor prognosis in ARM is associated congenital anomalies (20-80%). Other factors include delayed presentation, sepsis, and gut perforation.[4]

This study was carried out to evaluate age at presentation, low birth weight, septicemia, and

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postoperative complications as factors of poor outcome (mortality) in neonates with ARM.

METHODS

This is a prospective descriptive study conducted in the Department of Pediatric Surgery, The Children's Hospital, Pakistan Institute of Medical Sciences, Islamabad, Pakistan. The study was carried out between October 2019 and March 2020. A total of 44 consecutive neonates with ARM were included in the study. Data were collected on a structured proforma after taking approval from the Institutional Review Board.

Variables studied include age at presentation, gender, birth weight, type of malformation, sepsis at presentation, type of surgery performed, postoperative complications, and outcome as survival or mortality. After stabilization of patients, a detailed history was taken and careful physical examination was done to classify the type of ARM and other visible associated abnormalities. Those with recto-vestibular fistula (RVF), persistent cloaca, and perineal fistula were confirmed on careful perineal examination. Those in whom meconium was present at meatus without micturition were classified as recto-urethral fistula and those in whom urine was mixed with meconium while micturition was classified as recto-bladder neck fistula. Lateral cross table X-ray was performed in ARM without fistula to find out the level of distal gas shadow. Ultrasound was performed in order to find out associated genitourinary anomalies. Echocardiography was not performed due to non-availability. All the neonates were managed in the pediatric surgical ward.

After initial resuscitation, a pelvic divided colostomy at descending colon was performed in cases of ARM with recto-urethral fistula, recto-bladder neck fistula, ARM without fistula where distal gas shadow >1 cm from the anal pit. Primary Posterior Sagittal Anorecto-plasty (PSARP) was performed in cases of Recto-vestibular fistula and ARM and Primary limited Posterior Sagittal Anorectoplasty with perineal fistula. Patients were followed up for postoperative complications.

Patients were categorized into two groups based on their birth weight of less than 2.5 kg and more than 2.5 kg.

Sepsis was defined as Leukocytosis/ leukopenia, thrombocytopenia, increased Prothrombin time and International Normalized Ratio (INR), Positive C-Reactive Proteins and Fibrin degradation products on laboratory investigations.

The statistical analysis was performed using statistical SPSS version 21. Descriptive statistics like mean and standard deviation for quantitative data and frequency and percentages for qualitative data were calculated. A Chi-square test was applied to associate different parameters with the outcome. P-value < 0.05 was considered significant.

Written informed consent was taken from parents/guardians and approval to carry out the study was taken from the institutional ethical review board.

RESULTS

A total of 44 neonates with ARM were included in the study. Out of 44 neonates, 56.8% (n=25) were males and 43.2% (n=19) were females. The mean age at presentation was 2.1 ± 0.5 days with a minimum age of one day and maximum age of 3 days. The mean birth weight was 2.5 ± 0.6 kg with a minimum weight of 1.5 kg and a maximum weight of 4.3 kg. The type of anorectal malformations in neonates presented are shown in Table 1. At presentation, 22.7% (n=10) neonates had sepsis and out of the 6 died before surgery. All the patients with sepsis presented to us after 24 hours of life. Types of surgeries performed in the remaining 38 neonates are shown in Table 2. Postoperative complications were Anal stenosis in 2 patients with primary PSARP which were managed by anal dilatations, 2 patients had wound dehiscence which was managed conservatively, and 4 patients had postoperative sepsis. In our study, 70.5% (n=31) of patients had successful outcomes and were discharged. Overall mortality was 29.5% (n=13) out of which 13.63% (n=6) died preoperatively and 15.9% (n=7) died postoperatively. The most common cause of death in postoperative patients was sepsis (40%).

Table 1: Types of Anorectal Malformations (ARM)

Type	Frequency	Percentage
ARM with perineal fistula	8	18.1
ARM with recto-urethral fistula	2	4.5
ARM with recto-bladder neck fistula	2	4.5
ARM with recto-vestibular fistula	10	22.7
ARM without fistula	18	40.9
Common cloaca	4	9

Various variables such as age at presentation, birth weight, sepsis at presentation, and postoperative complications were compared for their association with outcome. There was a statistically significant relationship between low birth weight and mortality (P= <0.01). Mortality in neonates with birth weight less than 2.5 kg was 62.5% (10/16). Whereas, only 10.7% (3/28) of patients with a birth weight of more than 2.5kg died. There was also a significant relationship between sepsis at presentation and mortality (P=0.001). Seven out of ten patients with sepsis at presentation died.

There was no statistically significant association was found when the outcome was compared with age at presentation (P=0.21) and postoperative complications (P=0.16) Table 3.

Table 2: Surgeries performed in different types of ARM

Type of malformation	Pelvic Divided Colostomy	Primary PSARP*
ARM with perineal fistula	0	4
ARM with recto-urethral fistula	2	0
ARM with recto-bladder neck fistula	2	0
ARM with recto-vestibular fistula	4	6
ARM without fistula	16	0
Common cloaca	4	0

*PSARP= Posterior Sagittal Anorectoplasty

Table 3. Association of different variables with poor outcome

Variable	p-value
Low birth weight	<0.01
Sepsis at presentation	0.001
Delayed presentation (after 24 hrs)	0.21
Postoperative complications	0.16

DISCUSSION

Neonates with ARM are traditionally managed with staged procedures involving a diverting colostomy at the first stage and definitive repair in the second stage followed by the closure of colostomy at the third stage. Recently, patients with low malformations such as recto-vestibular fistula in girls are being increasingly managed as a single-stage procedure, Primary PSARP to avoid the morbidity of colostomy.[5] Our practice is to do primary PSARP in low malformations and staged procedures in high ARMs.

Various factors affect the outcome of ARM in neonatal age. In our study, one of the most important factors affecting the outcome of ARM was the low birth weight (LBW). Mortality in neonates with birth weight less than 2.5 kg was 62.5% (10/16). In a large retrospective study conducted by Cassina et al., low birth weight (<2.5 kg) was found to be a significant risk factor of mortality in ARM (P= 0.001).[6] In another study, mortality in ARM was studied based on their birth weight by dividing the study population into appropriate birth weight (>2.5kg) and low birth weight (<2.5kg). They found that mortality in LBW neonates was significantly higher than appropriate birth weight (51.4% Vs 15%).[7] Similar results were found in a study conducted by Chirdan et al. with significantly higher mortality in the LBW group (p=<0.05).[5] Chowdhary et al. calculated 40% mortality in LBW neonates with ARM as compared to 2.9% in neonates with weight >2.5 kg (P=<0.01).[8] The overall estimated prevalence of low birth weight worldwide is 16%. The estimated prevalence of low birth weight in the least developed and developing countries is higher (19%) than in developed countries (17%). The main cause of low birth weight in Asian developing countries is intrauterine growth restriction which is influenced by maternal and socioeconomic factors before and after pregnancy. Pakistan has one of the highest prevalence of LBW ranging from 19% in urban areas to 32% in rural areas.[9] In our study 36.4% (16/44) patients had LBW. In Pakistan, neonatal mortality secondary to LBW can be reduced by creating public awareness by organizing Nutrition Education programs for pregnant women by lady health workers and public health specialists. Hence, improvements in antenatal care can reduce overall mortality in neonates with ARM.

Septicemia has a significant association with poor outcomes in our study. Bima et al. studied risk factors for mortality of ARM patients in Palembang and found sepsis to be the significant risk factor of neonatal mortality (P=0.049).[10] In another Nigerian study, sepsis was found to be the major contributor to neonatal mortality in ARM.[11] In the developing world, due to the lack of resources, trained midwives/LHVs, and adequate medical facilities, neonatal sepsis is the major cause of morbidity and mortality. Activation of a cascade of inflammatory processes in neonatal sepsis leads to tissue damage and organ dysfunction such as respiratory failure, cardiovascular failure, hepatic and renal failure eventually leading to death.[12] In our study, all the patients with neonatal sepsis were managed in the pediatric surgical ward in collaboration with neonatologists. Availability of adequate medical facilities such as surgical NICU, specialized and dedicated nursing staff, and neonatal intensivist can dramatically improve the outcome of surgical patients with neonatal sepsis.[3]

Age at presentation did not affect the outcome of our study. However, in a Nigerian study, the delayed presentation was associated with a poor outcome.[11] The delayed presentation was also a risk factor of mortality in an Indian study.[7] However, in both studies, the mean age of presentation was not mentioned. The difference in results in our study may be due to the mean age of presentation. In our study, the mean age of presentation is 2.1 days with a maximum age at presentation of 3 days. Higher mean age at presentation may be the reason for significant association with poor outcomes in these studies. Another factor that may cause differences in results with these studies is the sample size. The sample size in both studies is almost double that of our study.

There are various other factors affecting outcome in neonates with ARM which were not included in our study but investigated by other authors. Dastamuar et al. found a significant relationship between the type of ARM and mortality. According to their study, high ARM had a significant association with mortality (P=0.036).[10] According to Cassina et al., presence of two or more additional major congenital defects were significantly associated with poor outcome (HR, 7.9; 95% CI, 2.2–27.8; p=0.001).[6] An Indian study concluded that apart from low birth weight, major associated anomalies and delay in referral were the important factors that influenced the outcomes of babies with ARM during the neonatal period.[7] Another Indian study showed significant mortality in ARM cases associated with esophageal Atresia.[2] Mirza et al. found a statistically significant relationship

of associated congenital anomalies with mortality in neonates with ARM.[4]

A limitation of our study is a small sample size. A large sample size with a longer duration of study may help in better understanding of the correlation of different variables such as delayed presentation and postoperative complications with the outcome. Non confirmation of cardiac defects through echo-cardiography and non-evaluation of its effects on the outcome is another limitation of our study.

CONCLUSION

Low birth weight and sepsis are proven as factors of poor prognosis in our cohort of patients. In poor countries, the lack of resources, lack of trained midwives/Lady Health Visitors, and intensive care are contributing factors to sepsis and delayed presentation, and ultimately mortality. Good antenatal care, awareness of the midwives/LHVs to refer such patients in time, and provision of adequate intensive care can improve the outcome of surgery in ARMs.

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