Original Article

Frequency of Cardiac Anomalies in Patients of High and Low Variety Anorectal Malformation (ARM)

Cardiac Anomalies

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ABSTRACT

Objective: To study the frequency of cardiac anomalies in patients of high and low variety anorectal malformation (ARM)

Study Design: Prospective Observational study

Place and Duration of Study: This study was conducted at the Inpatient Department of Pediatric Surgery, Nishtar Hospital, Multan and Children Hospital and Institute of Child Health Multan for 06 months from July, 2019 to January, 2020.

Materials and Methods: After the approval of ethical committee, parental informed consent was taken from the parents before the commencement of study. All the neonates that presented with absent anus, had undergone Invertogram study and were classified as low or high ARM. Echocardiography was performed in all patients to diagnose cardiac anomalies. Data was collected on a predesigned performa and analyzed statistically on SPSS version 20.

Results: Out of 121 patients, 83 (68.6 %) were male patients while 38 (31.4 %) were female patients. Mean age of our study cases was 1.23 ± 0.513 days. Those who belonged to rural areas were 74 (61.2 %) and 47 (38.8 %) belonged to urban areas. Mean gestational age of these children was 38.14 ± 1.29 weeks and 106 (87.6%) had gestational age more than 36 weeks. Regarding mode of delivery, 99 (81.8%) were born vaginally and 22 (18.2%) through cesarean section. Positive family history of anorectal malformations was noted in 9 (7.4%) whereas 89 (73.6%) mothers were illiterate. Low variety ARM was found in 59 (48.8%) cases and high variety ARM in 62 (51.2%) while cardiac anomalies were noted in 40 (33.8%).

Conclusion: Very high frequency of cardiac anomalies was noted in our study among neonates presenting with anorectal malformation. Cardiac anomalies were significantly associated with gender, family history of anorectal malformation and high variety of ARM. Such patients can be helped by early diagnosis and timely management to decrease burden of related morbidity and mortality.

Key Words: Anorectal malformation, Cardiac anomalies, Congenital anomalies.

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INTRODUCTION

In pediatric surgical practice Anorectal Malformation (ARM) is one of the most frequently presenting congenital anomaly.[1] The neonate is born without a normal anal opening.

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ARM is responsible to cause neonatal intestinal obstruction, which is usually diagnosed by the absence, or ectopic location of the anus and delayed passage of meconium.[2] One of every 4000 to 5000 infants is affected by ARM although this condition is more common in certain geographic areas and it is slightly more common in males[3] while annual report of international Clearing house for Birth Defects Surveillance (2011) revealed incidence ARM is approximately 1 to 3 in every 5000 live births.[4]

There are several forms of imperforate anus and anorectal malformations. The new classification is in relation to the type of associated fistula. The classical Wingspread classification was in low and high anomalies: A low variety ARM, in which the colon remains close to the skin. A high variety ARM, in which the colon is higher up in the pelvis and there is a fistula connecting the rectum and the bladder, urethra or the vagina.

Patients with these malformations often have other associated congenital anomalies. About 20-80% patient of ARM have different anomalies including cardiovascular, genitourinary, gastrointestinal, neurological and musculoskeletal anomalies.[5] Although it is difficult to directly compare the types of associated anomalies because of their varied nature. The diagnosis of these anomalies on early stage is very important, because correct operative approach may be planned and prognosis for the infant may be assessed, second reason is that the mortality and morbidity among infants with cardiac anomalies is directly related to the nature and severity of the other malformations.[6] Associated anomalies such as congenital heart disease, are present in a high percentage of patients with ARM. These associated anomalies are serious and have long term prognosis.[7] Different studies reported that prognosis of child with ARM more often depend on extent of these associated anomalies than on ARM itself.[7] The frequency and types of such associated anomalies different among various reported studies, ranging among 20% to 70%.[5] A study conducted in India by Ratan et al[8] has reported 10% cardiac anomalies in ARM while cardiac anomalies proportion in high variety was 85 % (n = 34 /40) and in low variety it was 15 % (n= 6/40) showing cardiac anomalies were significantly more prevalent in high variety (p=0.01).

Owing to the scarcity of local data from Pakistan on this topic, this study has been proposed to ascertain current magnitude of the problem to give baseline information which will help clinicians to diagnose cardiac anomalies at early stage, followed by timely management. This will improve quality of life of such patients in terms of disease related morbidities leading to several hospital admissions. It will also help hospital authorities in terms of extra financial burden.

MATERIALS AND METHODS

It was a prospective observational study, carried out at Department of Pediatric Surgery, Nishtar Medical University Hospital, Multan during July 2019 to January 2020. Sampling technique was non – Probability Consecutive sampling. In this study 121 cases of both gender (male and female) having age up to 28 days of life presented with ARM, were included. If parents are not giving consent, it was the only exclusion criteria.

Sample size was n= 121, P1 = 85 % (cardiac anomalies with high velocity), P2 = 15 % (cardiac anomalies with low variety), d = 0.09 and confidence level = 95 %

The study was started after approval from ethical committee of Nishtar Medical University and children hospital and institute of child health (CHICH) Multan. Consecutive (n = 121) patients meeting inclusion criteria of our study were recruited from the emergency Department of Pediatric Surgery, Nishtar Hospital and CHICH Multan. Informed consent was taken from parents after briefing them about the objectives and procedures of this study. Baseline characteristics including age, gender, gestational age, mode of delivery (Vaginal / Cesarean section), previous sibling with ARM and maternal education were noted. All the neonates had undergone Cross-table lateral radiograph study and were classified as low or high variety ARM. Echocardiography was performed by pediatric cardiologist in all patients included in study to diagnose cardiac anomalies and type of cardiac anomaly as defined in operational definitions.

Data was entered and analyzed by SPSS version 22. Mean and standard deviation were calculated for quantitative data like age of the patient and gestational age. Frequency and percentage was calculated for gender, age groups, residential status, Mode of delivery (Vaginal / Cesarean section), Variety of ARM, family history, maternal education and cardiac anomalies.

Frequency of cardiac anomalies in high and low variety ARM was compared by chi-square test and p-value of ≤ 0.05 was taken as significant.

Effect modifiers like age, gender, residential status, Mode of delivery (Vaginal / Cesarean section) Variety of ARM, family history of cardiac diseases, maternal education and gestational age were controlled by stratification. Post stratification Chi-Square test was applied to see the effect on frequency of cardiac anomalies in high and low variety ARM and p-value of ≤ 0.05 was taken as significant.

RESULTS

Our study comprised of a total of 121 patients meeting inclusion criteria of our study. Of these 121 study cases, 83 (68.6 %) were male patients while 38 (31.4 %) were female patients. (Figure No. 1). Mean age was 1.23 ± 0.513 days (with minimum age of 1 day to 3 days). Majority of the cases i.e. 93 (81.0 %) were aged up to 24 hours.

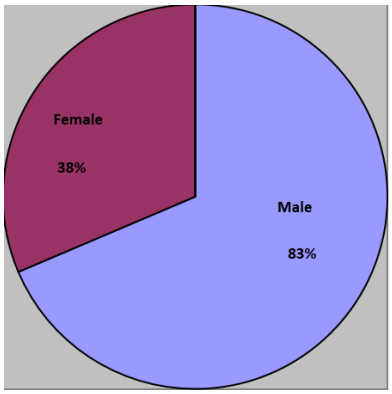

Figure No.1: Gender distribution of ARM Patients

Mean gestational age of these children was 38.14 ± 1.29 weeks and 106 (87.6%) had gestational age more than 36 weeks. Majority of these 99 (81.8%) were born vaginally and 22 (18.2%) through cesarean section. Positive family history of anorectal malformations was

noted in 9 (7.4%) whereas 89 (73.6%) mothers were illiterate. It was found that 74 (61.2 %) belonged to rural areas and 47 (38.8 %) belonged to urban areas.

Low variety anorectal malformations were noted in 59 (48.8%) and high variety anorectal malformation was noted in 62 (51.2%). Cardiac anomalies were noted in 40 (33.8%) and these cardiac anomalies were stratified with regards to gender, age, residential status, gestational age, mode of delivery, family history, maternal literacy and variety of ARM. (Tables 1 - 2).

Table No.1: Stratification of Cardiac Anomalies with regards to gender

Gender	Cardiac Anomalies (n=121)		P – value
	Yes(n=40)	No(n=81)	
Male (n=83)	22	61	0.036
Female (n=38)	18	20	

Table No.2: Stratification of Cardiac Anomalies with regards to variety of ARM

Variety	Cardiac Anomalies (n=121)		P – value
	Yes(n=40)	No(n=81)	
Low variety (n=59)	12	47	0.004
High Variety (n=62)	28	34	

DISCUSSION

Anorectal malformations occur approximately in 1/1,500 to 1/5,000 live births. They may occur alone, but they can commonly have other associated anomalies or occur as a part of the combined anomaly. The treatment involved may be complicated by a need to address the associated anomalies, in addition to the anorectal malformations. Furthermore, the problems of these associated anomalies could have more of an impact on the morbidity and mortality before and after surgical treatment. The surgical and medical management of patients with these malformations can be complex, especially when one considers the high frequency of concomitant anomalies that occur with ARMs. This frequency of additional anomalies in patients with ARMs ranges from 40% to 70%. Thorough evaluation of patients with ARMs is essential because it is these coexisting anomalies that account for most of the morbidity and mortality that is associated with this condition.[9]

The frequency of associated anomalies in other organs is known to be approximately 40-70% in the decreasing order of urogenital system, musculoskeletal system, and cardiovascular system. In addition, anomaly in other parts of the gastrointestinal system can occur concomitantly, and a prompt treatment is required in such situations when the neonate's life is threatened. In particular, when an esophageal atresia accompanies the anorectal malformations, the complex of surgical procedure and the difficulty of situational postoperative management must be considered.[10]

Our study found that 83 (68.6 %) were male patients while 38 (31.4 %) were female patients. A study conducted by Khawaja et al) from Lahore had found 79 % male gender predominance.[11] Qazi et al had reported 68 % male gender predominance[12] and study conducted by Byun et al reported 12 : 1 male to female ratio in ARM[13], were found in compliance with our study results. Similar results were also found by Mirza et al and by Cho et al with 67 % male gender predominance.[14,15]

Mean age of our study cases was 1.23 ± 0.513 days years (with minimum age of our study cases was 1 day to 3 days). It was also indicated that majority of our study cases i.e. 93 (81.0 %) were aged up to 24 hours. A study conducted by Qazi et al had also reported that majority of patients presenting within 24 hours[12] while study by Mirza et al had reported 3.4 days mean age.[14]

Mean gestational age of these children was 38.14± 1.29 weeks and 106 (87.6%) had gestational age more than 36 weeks which coincides with the study conducted by Byun et al.[13]

It was noted that patients with low variety anorectal malformations were 59 (48.8%) and high variety anorectal malformation was noted in 62 (51.2%). Byun et al has indicated 54.9 % high variety ARM[13] which is close to our results. A study conducted in USA by Cho et al has also reported 58 % high variety ARM.[15]

In this study, cardiac anomalies were found in 40 (33.8%). Qazi et al and Byun et al had described 38 % and 39.6% cardiac anomalies in anorectal malformation respectively[12,13], while Mirza et al had reported 8 % cardiac anomalies[14] which is quite lower frequency than our study. Cho et al had reported 27% cardiac anomalies in USA.[15]

Our study had few limitations. Individual cardiac anomalies were not studied which may furthur elaborate the severity of cardiac anomalies in terms of survival and mortality. There are many studies which classify the cardiac anomalies associated with GI malformations. A study done by Goroko et al in 2004 indicated that The most common CHD was isolated atrial septal defect (ASD) (73%), followed by ASD + ventricular septal defect (VSD) + patent ductus arteriosus (PDA) (7.6%), ASD + VSD (3.8%), ASD + PDA (3.8%), VSD (3.8%), PDA (3.8%), and coarctation of the aorta (3.8%). (16) Similar study was also done by Teixeira et al to find the prevelance of cardiac anomalies with low variety anorectal malformation.[17] Similar studies performed in Saudi Arabia showed the cardiac anomalies with type of ARM associated. Other GI anomalies were also discussed in this study.[18]

Anorectal malformations are associated with VACTERAL which provides a vast scope of research. Regarding Cardiac anomalies the survival of the

patients can be assessed to plan single or staged procedure for correction of ARM.[19,20].

CONCLUSION

Very high frequency of cardiac anomalies was noted in our study among neonates presenting with anorectal malformation. Cardiac anomalies were significantly associated with gender, family history of anorectal malformation and high variety of ARM. All clinicians treating ARM patients should anticipate cardiac anomalies in these patients for early diagnosis and timely management which will decrease burden of related morbidities and mortalities.

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Author's Contribution:

Concept & Design of Study:	Abdul Latif
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Data Analysis:	Shakeel Ahmed, Matee-Ullah, Muhammad Kashif
Revisiting Critically:	Abdul Latif, Mahreen Zahra
Final Approval of version:	Abdul Latif

Conflict of Interest: The study has no conflict of interest to declare by any author.

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