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Congenital Heart Diseases with Ano-Rectal Anomalies: A local Experience

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Abstract— Background& objectives: Neonates with anorectal anomalies associated with congenital heart disease (CHD) pose a major challenge to cardiologists and surgeons. The aim of this study is toevaluate the frequency of patients with anorectal anomalies (ARA) associated with CHD at a tertiary care hospital in a5 years period. **Methods:** This retrospective study reviewed the filling system of all children who underwent surgical correction for ARA either of high or low type in a 5 years period from January 2012 until December 2017. Various assessment measures were evaluated and reported including; clinical examination, electrocardiography (ECG), chest radiography, and color Doppler echocardiography. **Results:**Sixty-three patients with ARA were studied. They were 44 males and 19 females with a male to female ratio of (2.3: 1). Out of the total patients, 41 showed CHD with a percentage of 65.1%. CHD types varied from atrial-septal defect (ASD) in 28 patients (68.3%), coarctation of the aorta in 2 patients (4.9%), mixed anomalies of ASD associated with ventricular-septal defect (VSD) and patent ductus arteriosus (PDA) in 8 patients (19.5%). Two patients (4.9%) showed multiple CHD that were ASD + PDA while the remaining one (2.4%) were having VSD. **Conclusion:**The frequency of CHD in patients with ARA was very high. The commonest CHD encountered presentation was ASD. The current reported data indicate the needs to establish larger scale studies at national or international levels for accuratelydetermine the frequency of CHD in ARA patients.

Key words: Congenital heart disease, Anorectal anomalies, CHD, ARE, ASD.

INTRODUCTION

Anorectal anomalies (ARA) isone of the major congenital anomalies in which a baby, in most of the cases, is born with no apparent normal anal opening. The incidence of ARA ranged fromtwo to 2.8 per 10,000 live births, with significant global variations in their prevalence[1-2]. However, the prevalence of anorectal anomalies in Saudi Arabia is approximately 1/1000 live births with a male-to-female ratio of 1.9:1[3].Other reported the incidence of ARMs in Madinah Kingdom of Saudi Arabia, is higher than the reported data.They may be single anomaly or associated with other congenital anomalies or syndromes [4]. ARA is not usually lethal;yet, managing patients with such anomaliesis mostly complex, especially with the high frequency of associated anomalies that ranges from 40% to 70%[5-6]. The morbidity and mortality among those patients areoften dueto the highincidence ofassociated anomalies thatnecessitate a compulsoryand comprehensive evaluation and management of theanomalous patients.Therefore, it is very imperative to diagnose any cardiac condition of the child before any surgery takes place to avoid any possible complications during anesthesia [7].Many other anomalies may associate with ARA. They include trachesophageal fistulae (TEF), urogenital anomalies, spinal defects, limbs and extremities, as well as renal and cardiovascular system.Mostof theassociated cardiac lesions are corrected post the initial repair of the ARA except for limited cases that present with cyanosis and high complex CHD[8-10].

To our knowledge, the frequency andnature of CHD associated with ARA was not clearly reported in Saudi Arabia. The current retrospective study aimedto report and evaluate thefrequency and nature of associated CHD in newborns with ARA.

SUBJECTS &METHODS

This retrospective study reviewed the filling system of all children who underwent surgical correction for ARA, either of high or low type in a 5 years period from January 2012 until December 2017. Various assessment measures were evaluated and reported including; clinical examination, electrocardiography (ECG), chest radiography, and color Doppler echocardiography.The findings were tabulated as demographic patient's data including, the gender of the patient, type of ARA and types of CHD associated anomalies. Data were statistically analyzed usingSPSS Statistics program, version 23. Obtained data were subjected to a descriptive analysis.

Ethical issues: This study was approved by the research ethical committee and Institutional research board (IRB) of King Faisal University (KFU) - College of Medicine.

RESULTS

Sixty-three patients were studied. They were 44 males and 19 females with a male to female ratio of 2.3: 1. The demographic data of the studied patients are shown in table 1.

Forty-one patients out of the total studied patients with ARA suffered of CHD. Most of them had ASD (28, 68.3%). Data are shown in table 2.

Figure 1 showed the electrocardiographic picture of ASD in a 3 months old boy.

DISCUSSION

Association between congenital heart diseases and gastrointestinal tract anomalies is well documented. Nevertheless, reports on the prevalence of association of CHD with anorectal anomalies are not obvious [11]. CHD frequency among these cases is high [12-14]. Theymay be associated with chromosomal anomalies and sometime non syndromic multiple anomalies[15-17].Such patients may undergo non-cardiac surgery and have higher perioperative risk. Hence, it is mandatory to clearly diagnose them, before the surgery.The advanced echocardiography with Doppler color flow measurements have made it easy to screen and diagnose CHD[18].

The current study encompassed 63 infants with ARA. The incidence of significant cardiac malformations was 65.1%, indicating a strong and true association. Our data showedan incidence close to a previously published data[19]. They concluded that the frequency of CHD in gastrointestinal malformations patients was very high[19]. Moreover, the current studyexpressed different types of CHD in association with ARA. ASD was the most common CHD with incidence of 68.3 % (28 cases out of 41). These data coincide with another study[20] that reported the frequency of cardiac defects in 242 patients with GI malformations to be 31 (44.9%) with ASD, 17 (24.6%) with VSD, 5 (7.2%) with PDA, 3 with ASD + VSD + PDA, 2 with isolated dextrocardia, and 2 with aortic stenosis[20]. In addition,other study of 43 children with gastrointestinal malformation, reported ASD to be the commonest congenital cardiac anomalies diagnosed among these children[19].Some authors stated the presence of associated extra cardiac anomalies to be 21.4% [21], others reported a similar figure of association between CHD and gastrointestinal system malformation in a non-syndromic condition[22-25]. However, others documented to reach up to 65% in syndromic association[6].In our current study, we reported an overall higher CHD percentage (65.1%) this may be attributed to the variability of the prevalence of CDH with other gastrointestinal tract anomalies.

Many proposals have raised to explain the relationship between CHD and GISM in general and ARM in particular. The most reliable explanation maybe the failure in midline mesodermal embryogenesis[26].

CONCLUSION

The current study concluded that; the frequency of CHD in patients with ARA is significantly high. The commonest CHD encountered presentation is ASD. It recommends echocardiography screening for neonates with ARAfor early diagnosis of any associated cardiac anomalies, not only for proper preoperative assessment and postoperative management but also for successful follow up. This may avoid unfavorable or fatal outcome, as mortality and morbidity rate may be much higher in presence of such association.

The reported data also indicate the need to establish larger scale studies within the national or the international levels to determine the prevalence of CHD in ARA patients.

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Conflicts of Interest

None

Ethical Standards

The author assert that all procedures contributing to this work comply with the ethical standards of the guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the ethical committee and Institutional research board (IRB) of King Faisal University (KFU) - College of Medicine.

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Tables:

Table 1: Demographic data of the study population

Sex	Number = 63	%
Males	44	69.8
Females	19	30.2
Age	Less than 1 up 12 months	%
Less than 1 month	36	57.1
1-2 months	15	23.8
2-12 months	12	19.1

Table 2: Types of Congenital heart Diseases (CHD) associated anorectal anomalies (ARA)

ARA No	CHD Total No %	ASD	Aortic Coarctataion	ASD+VSD+PDA	ASD+ PDA	VSD
63	41 65.1%	28 68.3%	2 4.9%	8 19.5%	2 4.8%	1 2.4%

ASD =Atrial Septal Defect**, VSD** = Ventricular Septal Defect**, PDA**= Patent Ductus Arteriosus

Figures:

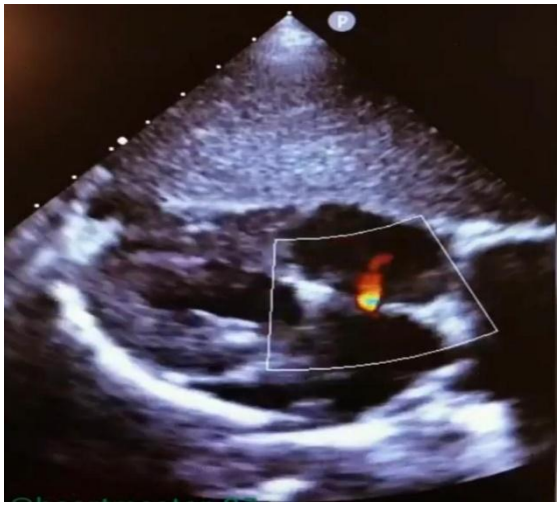

Fig 1: Echocardiography (subcostal view) showing Secundum ASD for a 3-months-old child.