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Congenital Heart Defects in Children with Dextrocardia: A Ten-Year Study

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Abstract

Background

Dextrocardia is a malposition of the heart defined as the right-sided development of the heart. It can increase the likelihood of congenital heart defects or diseases (CHD) and the risk of related morbidities and mortalities. We aimed to determine the frequency of CHDs among Dextrocardia patients.

Materials and Methods

In a retrospective cross-sectional study the records of patients with Dextrocardia who referred to Imam Reza Hospital (tertiary referral center) of Mashhad between 2006 and 2016 were studied. Data were analyzed using SPSS software version 16.0.

Results

In total 163 patients, 85 of whom (54%) were males were studied. Their mean age was 11.41± 0.326 years. The frequency of major Dextrocardia subtypes were: Situs inversus in 77 (47%), Situs Solitus in 59 (36%), and Situs ambiguous in 28 (17%) patients. The frequency of associated complex congenital heart defects (CHD) was 55% in Situs inversus, 77% in Situs solitus and 100% in Situs ambiguous. Isolated associated CHD had a greater frequency in the Situs inversus and Situs solitus groups. The most common isolated associated CHD was septal defects.

Conclusion

More than 90% of all patients with Dextrocardia, had CHD whereas all patients with Dextrocardia and Situs ambiguous had complex CHD.

Key Words: Children, Congenital Heart Defects, Cardiac Malposition, Dextrocardia, Visceral situs.

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1- INTRODUCTION

Congenital malformations are one of the major causes of infant mortality (1, 2). Previous studies on congenital heart disease (CHD) have reported congenital malformations as six to eight cases per 1,000 live births (3 ,4), whereas in more recent studies, this rate has been reported as about one percent of live births; so CHDs are the most common congenital defects (5, 6). Currently, different incidents have been reported for Dextrocardia regarding the sampling method (7-10). This study was performed on Dextrocardia, the major subtype of cardiac malposition. Dextrocardia is defined as the right-sided development of the heart, with the axis from the base of the heart to the apex pointed to the right side of the chest (from the Latin term "dexter", meaning "right," and the Greek term "kardia", meaning "heart") (7).

Normal positioning of the abdominal viscera is defined as the superior and inferior vena cava positioned in the right side of the abdomen entering the right atrium, while the liver is placed on the right and the stomach on the left (6). If the viscera are in their normal position it is named as Situs solitus, with a mirror position, Situs inversus, and if they have an unspecified position or are positioned in any other way, it is called Situs ambiguous. Complex heart diseases are indeed sever malformations that cause an abnormal and complex physiological state for the patient throughout all of his/her lifetime (11). Therefore, except for complex cases, the isolated cardiac defects that are often related to a specific part of the arteries and the heart and are categorized separately and include: septal defects, great vessels abnormalities, arterial or venous abnormalities, pulmonary arteries abnormalities, etc. The purpose of this study was to further increase our knowledge about the congenital heart defects (CHD) associated with Dextrocardia in patients referring to Imam Reza Hospital (tertiary referral center) in Mashhad city, Iran.

2- MATERIALS AND METHODS

2-1. Study design and population

The purpose of this retrospective cross sectional study was to describe the frequency of heart anomalies associated with Dextrocardia and various visceral Situs. The records of children with Dextrocardia who referred to Imam Reza Hospital in Mashhad city (North East of Iran), during the period from 2006 to 2016, were studied. The epidemiological data of patients with Dextrocardia who were examined or admitted to the outpatient clinic at any age and gender were also enrolled. In total, 163 patients' records that had Dextrocardia were implied.

2-2. Measuring tools / Laboratory measurements

The data regarding the cardiovascular anatomy was systematically collected via echocardiography (VIVID 7 GE and Resona 7 MINDRAY with multifrequency convex probes, 2-5 MHZ) based on the segmental approach. The superior and inferior vena-cava according to their placement, aorta, veins and pulmonary arteries, atrioventricular and ventriculoarterial communications and ventricular and atrial defects were carefully considered. The location and method of connecting the arteries and cardiac veins and collateral arteries are described. Grouping of patients was done in the above mentioned categories as patients with complex heart disease versus isolated defects.

2-3. Ethical consideration

This research was approved by the Regional Ethics Committee for Medical Research in Mashhad University of Medical Sciences, Mashhad, Iran.

2-4. Data Analyses

Data were analyzed using SPSS software (version 16.0). P-value less than 0.05 were considered statistically significant.

3- RESULTS

In this study, randomized sampling was used from outpatient clients and patients admitted to a referral center for pediatric heart. In total 85 (54%) males with a mean age of 133± 0.329 months and 75 (46%) females with a mean age of 141± 0.387 months were studied (**Table.1**). Among the studied cases Situs inversus was diagnosed in 77 (47.2%), Situs solitus in 58 (35.5%), and situs ambiguous in 28 (17.1%). The prevalence of complex congenital heart disease in patients with Dextrocardia categorized in terms of their situs was as follows: 55% in Situs inversus, 77% in Situs solitus and 100% in Situs ambiguous in 100%. Moreover, twelve patients with Situs inversus totalis and 3 patients with Situs solitus were not involved with any anomalies (**Table.2**).

The prevalence of isolated CHD (noncomplex CHD) is presented in **Table.3**. In Situs inversus the most common cardiac defects were septal defects and pulmonary artery abnormality was more frequent in Situs solitus group. In the Situs inversus group, 45% of the cases had ASD followed by VSD (36%), PS (22%), PH (18%), AVSD (13%), and PDA (1%), respectively. PDA was reported in 50% of the cases in the Situs solitus group whereas 40% had ASD and/or VSD (**Table.3**).

Table-1: Demographic characteristics of children in control and interventional groups

Variables	Interventional Group	Control Group	Test	P- value
	N (%) or Mean (SD)	N (%) or Mean (SD)		
Gender				
Male	20 (46.5%)	23 (53.4%)	X^2=0.453	0.501
Female	20 (54%)	17 (46%)		
Age				
3 years	18 (64.2%)	10 (35.8%)	X^2=4.119	0.249
4 years	9 (37.5%)	15 (62.5%)		
5 years	8 (50%)	8 (50%)		
6 years	5 (41.6%)	7 (58.4%)		
Weight	14.40±1.676	14.02±1.625	t=-1.016	0.313
Time of venipuncture	4.92+1.071	4.70+0.966	t=-0.986	0.327

X2: Chi-square test; SD: Standard deviation.

Table-2: Comparison the pain mean during the intervention in control and interventional groups

Variable	Mean (SD)	Independent t-test	P- value
Control group	7.95 (1.084)	17.505	0.001
Interventional group	2.65 (1.577)		

SD: Standard deviation.

Table-3: Comparison the categorical pain intensity during the intervention in control and interventional groups by fisher test

Variable	Control group Number (%)	Interventional group Number (%)	P- value
Low (0-3)	0	31 (77.5%)	0.001
Average (4-7)	11 (30%)	9 (22.5%)	
High (8-10)	29 (70%)	0	

4- DISCUSSION

The purpose of this study was to determine the frequency and types of CHD associated with Dextrocardia. It is not reasonable to compare this study with other studies. In this study, randomized sampling was used on the outpatient clients and on those admitted to a referral center for pediatric heart. In other reports, the incidence or prevalence of Dextrocardia was reported in a live birth or pregnancy population (7-10). Bohun et al. (7) reported the incidence of Dextrocardia in pregnancy as 0.8 per 10,000, Claudine et al. (12) reported one out of twelve thousands of pregnancies, and Kidd et al. (9) evaluated this incidence 0.4 for every 10,000 live births. In the present study 163 echocardiographies have been performed on patients with Dextrocardia, which is more than the study by Garg et al. (14) in which 125 cases of echocardiography were analyzed. In several other studies, the number of studied patients with Dextrocardia was also less than the present study (7-14).

However, the proportion of Dextrocardia cases with Situs inversus (47%) compared to Situs ambiguous (17%) in our study was in contrast to that of Garg et al.'s report (14); they found Situs inversus in 39%, Situs solitus in 34%, and Situs ambiguous in 26% showing a roughly small difference. This difference could be justified by demographic variations or due to the fact that the incidence of cardiovascular malformations in Situs inversus is less and patients with Situs ambiguous often have complex heart diseases, Situs inversus and Situs solitus have better survival rates. In Bohun et al.'s study (7), Situs solitus 33%, Situs inversus 37%, and Situs ambiguous 30% were reported to be more common, in line with the study of Garg et al. (14). The mean age of Garg et al.'s study (14) was 9 years, 7.5 years in the Bohun et al. (7), and in this study, the mean age of patients was 11.4 years which was higher than the two similar studies. According to the two recent findings, it appears that the prevalence of different Situs varies from embryonic to school age, which can be attributed to various abnormalities associated with different Situs. Of course, it should be noted that the probability of referencing and marking on the Situs inversus is less than the others, so a different sampling type can cause these variations. In fact, Dextrocardia with Situs inversus is usually associated with fewer cardiac malformations, often diagnosis is coincidental or with cardiac compliant. In Dextrocardia-related abnormalities, except for complex cases, as in the general population (15-20), ventricular septal defect and pulmonary artery abnormality is more prevalent in Situs inversus and Situs solitus, and in the case of Situs ambiguous, according to the study complex CHDs are always possible. The presence of complex cardiac diseases in all forms of Dextrocardia is very high and their relative prevalence in each Situs shows Situs ambiguous with Dextrocardia needs more urgent or emergent treatment and has a noticeably poor diagnosis.

Figure.1 shows over 90 percent of Dextrocardia patients had CHD and **Figure.2** compares the risk of CHD in each patient with Dextrocardia that will appear through the visceral Situs. Dextrocardia and Situs inversus cases that had no defects were only 15.8% of that group. Patients with Situs solitus and Dextrocardia that had CHD were 95%. All cases with Situs ambiguous in this study had complex CHD. Frequency of complex CHD in Situs solitus was 77% and in Situs inversus was 55% that implies low prognosis for them. These statics show the risk of CHD in Dextrocardia is very high and urgent or emergent cardiologist consultation should be considered in all situations of Dextrocardia.

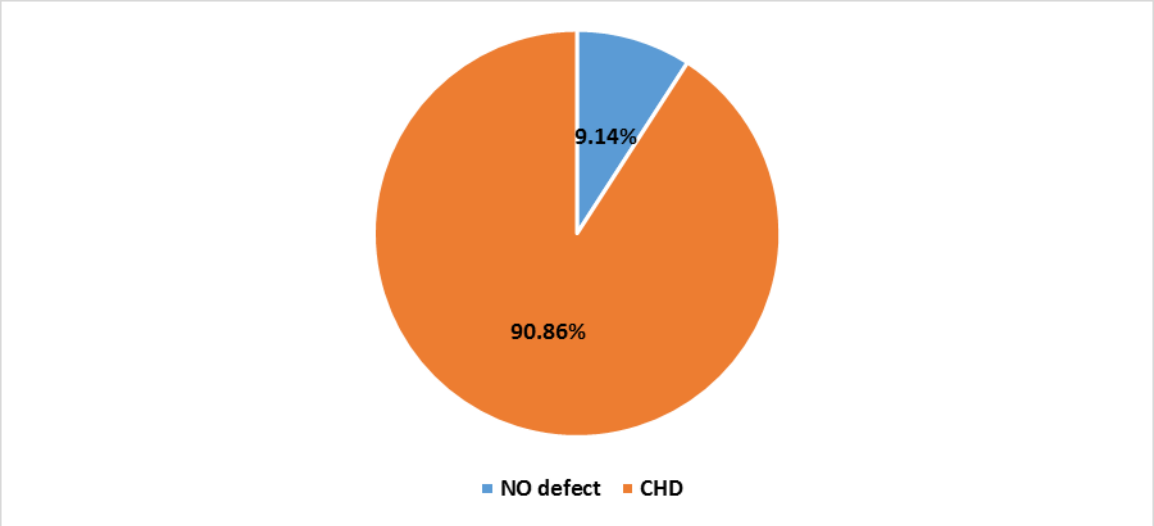

Fig.1: CHD in Dextrocardia Patients.

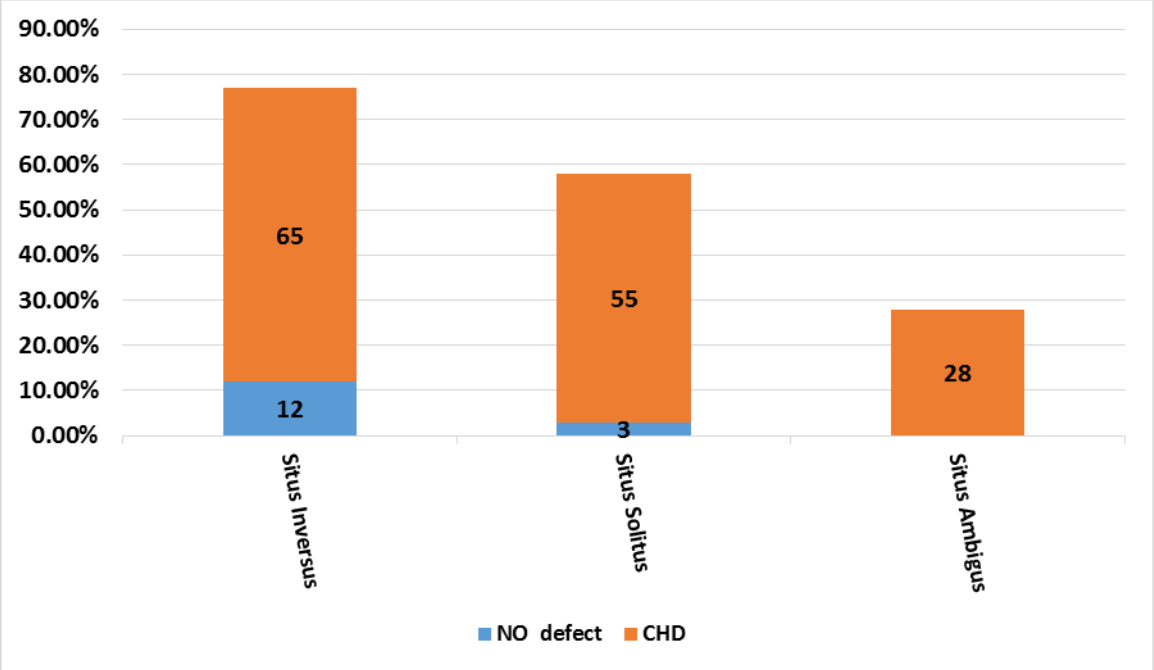

Fig.2: Frequency of CHDs vs. no-defects in Dextrocardia categorized by Situs.

5- CONCLUSION

The purpose of this study was to determine the frequency and types of CHD associated with Dextrocardia. More than ninety percent of all patients with Dextrocardia had CHD and all of the patients with Dextrocardia and Situs ambiguous had complex CHD. Complex CHDs was more common in Situs ambiguous and then Situs solitus.

6- ABBREVIATIONS

ASD: Autism spectrum disorder, VSD: Ventricular septal defect, PS: Performance status, AVSD: Atrioventricular septal defect, CHD: Congenital heart disease, PDA: Patent ductus arteriosus.

7- CONFLICT OF INTEREST: None.

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