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Original Article

Clinical Profile and Outcome of Patients with Dextrocardia

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Abstract

Background: The term dextrocardia means position of heart in the right side of the chest. Heterotaxia is a clinical condition where position of heart and viscera is altered. There are 3 terminology that includes all the positions: situs solitus, situs inversus totalis, situs ambiguous. Depending upon the types, the clinical presentations and outcomes vary. So, this study was done to know the different clinical profile and outcome of the patients with dextrocardia.

Objective of study: To determine the different clinical profile and outcome of patients with dextrocardia.

Methods: This Hospital based prospective, cross-sectional study was carried out at outpatients and inpatients in the Department of Paediatrics, Medical College and Hospital, Kolkata from December 2018 to May 2019. Data was analysed using SPSS software version 20.0.

Results: In total 8 patients, 3 of whom (37.5%) were males were studied. Their median age of presentation was 2.25 months with an interquartile range of 4.86 months. Situs inversus in 3 (38%), situs solitus in 4 (50%), situs ambiguous 1 (12%) in patients were observed. The frequency of associated complex congenital heart defects (CHD) was 66% in Situs inversus, 75% in Situs solitus and 100% in Situs ambiguous. The most common isolated associated CHD was septal defects (66% of all CHD). Most common presentation was respiratory distress (75%), followed by central cyanosis (25%) associated with underlying congenital heart disease (75%), followed by 12.5% presented with neonatal cholestasis syndrome (biliary atresia) and 12.5% had associated tracheoesophageal fistula and 12.5% was physiologically completely normal without any underlying defect. Outcome was also varied. 50% of all cases died. 25% due to severe sepsis with MODS, 12.5% due to sepsis with ARDS, 12.5% after Kasai procedure, 25% recovered successfully from LRTI and 12.5% is completely normal.

Conclusions: 75% of all patients with Dextrocardia, had CHD whereas all patients with Dextrocardia and Situs ambiguous had complex CHD. Sepsis was most commonly associated complications and mortality was very high.

Keywords: Dextrocardia; Heterotaxia; Situs inversus; Situs ambiguous; Congenital heart disease; Cyanosis; Sepsis

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 6-8 cases per 1,000 live births [3,4], whereas in more recent studies, this rate has been

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reported as about one percent of live births; so CHDs are the most common congenital defects [5,6]. The term dextrocardia (heart in the right side of the chest) expresses the position of the heart as a whole but do not specify the segmental relationship of the heart. A normally formed heart can be in the right chest because of extracardiac abnormalities.

On the other hand, a heart in the right chest may be a sign of a serious congenital heart defect. There are 3 terminologies that include all the positions: situs solitus, situs inversus totalis, situs ambiguous. Classification and diagnosis of abnormal cardiac position is done in 3 segmental approach: [7]

1. Determination of atriovisceral situs
2. Determination of position of ventricles
3. Determination of position of great vessels.

The four most common types of dextrocardia are (1) classic mirror-image dextrocardia, (2) normal heart displaced to the right side of the chest (dextroversion), (3) congenitally corrected TGA, and (4) mal-differentiated ventricle such as seen with asplenia or polysplenia syndrome [8].

Most of the time the diagnosis is accidental and it causes parental anxiety as well as difficulty for the clinician, partly because of the need of varied investigations like ECG, echocardiography, USG and CT scan of whole abdomen, rarely CT-angiography of chest to delineate great vessels clearly.

Management is also very difficult and ranging from medical management to cardiac surgery and other surgeries (like Kasai operation and repair of tracheoesophageal fistula) and the management of post-operative complications.

The objective (purpose) of this study was to find out the different clinical presentations and subsequent outcomes of the patients with dextrocardia in patients referring to Medical College and Hospital, Kolkata (tertiary referral), India.

Methods and Materials

Study design and population

The purpose of this cross sectional, longitudinal study was to describe the clinical profile and outcome of patients with Dextrocardia. The records of children with Dextrocardia who referred to Department of Paediatrics, Medical College and Hospital, Kolkata, during the period from December 2018 to May 2019, were studied. The patients with Dextrocardia who were examined at OPD or admitted to the inpatient clinic at any age and gender were also enrolled. In total, 8 patients' records that had Dextrocardia were implied.

Inclusion criteria: Patients accidentally or incidentally was found to have dextrocardia.

Exclusion criteria: All causes of dextroversion.

Measuring tools / Laboratory measurements

The data regarding the cardiovascular anatomy was systematically collected via echocardiography (VIVID 7 GE 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.

A detailed and thorough clinical examination were done in all cases.

Following investigations were done according to patient's clinical profile: complete haemogram, c-reactive protein, blood culture, chest X-ray, electrocardiogram, echocardiography, ultrasonography of abdomen, CT scan of abdomen, HIDA scan, HRCT thorax.

Ethical consideration

This research was approved by the Institutional Ethics Committee for Medical Research in Medical College and Hospital, Kolkata.

Data Analyses

Data were analysed using SPSS software (version 20.0). P-value less than 0.05 were considered statistically significant.

Results

In this study, median age of presentation was 2.25 months with a interquartile range of 4.86 months, among them 38% were males with a median age of 2.5 months with a interquartile range of 2 months and 62% females with a median age of 1.5 months with a interquartile range of 5.94 months were studied. Among the studied cases, situs inversus was diagnosed in 3 (38%), situs solitus in 5 (50%), and situs ambiguous in 1 (12%). The prevalence of complex congenital heart disease in patients with Dextrocardia categorized in terms of their situs was as follows: 66% in Situs inversus, 75% in Situs solitus and 100% in Situs ambiguous. Moreover, 1 patient with Situs inversus totalis and 1 patient with Situs solitus were not involved with any anomalies. In Situs inversus and Situs solitus group the most common cardiac defects were septal defects. 50% of the cases had VSD followed by ASD (38%), AVSD (25%), PS (22%), PH (18%).

Out of 412 patients admitted due to respiratory distress only 6 has dextrocardia. In our study also the most common presentation was respiratory distress (87.5%), polycythaemia was associated with 37.5% cases, central cyanosis was associated with 25% cases, congenital heart disease was associated with 75% cases (among which 45% were cyanotic and 30% were acyanotic), biliary atresia was associated with 12.5% cases, Tracheoesophageal fistula was associated with 12.5% cases. Underlying cause remain unknown in 87.5% cases, associated dysmorphism was with 12.5% cases, abnormality of situs was associated with 50% cases, sepsis was found to be most common complication and cause of increased mortality and morbidity and was associated with 75% cases. Associated features depicted in Table 1.

Discussion

The purpose of this study was to find out the different clinical presentations and subsequent outcomes of the patients with dextrocardia. In other reports, the incidence or prevalence of Dextrocardia was reported in a live birth or pregnancy population [9-12]. Bohun et al. [9] 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. [11] evaluated this incidence 0.4 for every 10,000 live births. However, the proportion of Dextrocardia cases with Situs inversus (38%) compared to Situs ambiguous (13%) in our study was in contrast to that of Garg et al.'s report [13]; they found Situs inversus in 39%, Situs solitus in 34%, and Situs ambiguous in 26% showing

Table 1:

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8
Polycythaemia	Y	N	Y	N	N	N	Y	N
Central cyanosis	Y	N	Y	N	N	N	N	N
Heart defects	Y	Y	Y	Y	N	Y	Y	N
Biliary atresia	N	N	Y	N	N	N	N	N
Tracheo-esophageal fistula	N	N	N	Y	N	N	N	N
Sepsis	Y	Y	Y	Y	N	Y	Y	N
Possible cause	Unknown	Unknown	TORCH infection	Unknown	Unknown	Unknown	Unknown	Unknown
Dysmorphism	N	N	N	N	N	N	Y	N
Situs	Ambiguous (Asplenia)	Solitus	inversus	Solitus	Situs inversus	Situs inversus	Solitus	Solitus

Table 2: Different laboratory parameters are depicted

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8
Haemoglobin (g/dl)	18.5	12.3	17.8	11.5	12	11.8	18	13
Total leucocyte count (per mm^3)	43,700	34,670	19,110	17,780	6,770	15,110	13,340	10,880
C-reactive protein (mg/dl)	36	27	8.8	10.5	2.5	12.3	9.6	5.8
Urea/ creatinine	Normal	Raised	Raised	Normal	Normal	Raised	Normal	Normal
Procalcitonin (ng/ml)	55	557	489	27	1	112	218	3
Blood culture	Pneumococus	Klebsiella	Gram Negative	Negative	Not done	Negative	Not done	Not done

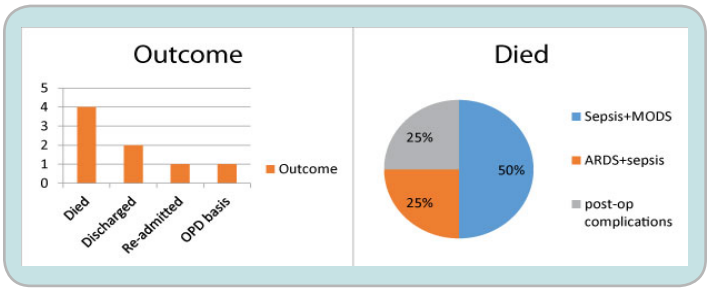
a roughly small difference. This difference could be justified by demographic variations or due to the fact that the incidence of cardiovascular malformations in Situs inversus is less and patients with Situs ambiguous often have complex heart diseases, Situs inversus and Situs solitus have better survival 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. [13]. The mean age of Garg et al.'s study [13] was 9 years, 7.5 years in the Bohun et al. [7], and in this study, the median age of patients was 2.25 months which was quite lower than the two similar studies. 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.

Situs inversus may be associated with other congenital anomalies such as duodenal atresia, asplenism, multiple spleens, ectopic kidney, horseshoe kidney, and various pulmonary and vascular abnormalities [5]. Situs inversus totalis that is associated with primary ciliary dyskinesia is known as Kartagener syndrome. Patients with primary ciliary dyskinesia have repeated sinus and pulmonary infections [14-15].

Many people with situs inversus totalis are unaware of their unusual anatomy until they seek medical attention for an unrelated condition [16]. Although the exact cause is unknown, dextrocardia has been linked with several factors including an autosomal recessive gene with incomplete penetrance, maternal diabetes, cocaine use, and conjoined twinning [17-19].

Girls have higher incidence as compared to boys in our study (37.5% were boys and 62.5% were girls. Median age of presentation is 2.25 months with a interquartile range of 4.86 months.

In our study most common presentation was respiratory distress (87.5%), followed by cyanosis (25%), followed by passing of clay coloured stool (12.5%). First two symptoms were equally distributed in both boys and girls whereas last symptom was seen in girl only.



The most common associated anomaly was heart disease (75%), followed by situs inversus (50%), followed by biliary atresia and tracheoesophageal fistula (each 12.5%)

The most common complication was sepsis (75%). ARDS were seen on 12.5% cases. AKI and multiple organ failure was seen in 12.5% cases.

50% cases required invasive ventilation whereas 12.5% cases required high flow oxygen.

50% of all cases died. Among them 50% died due to sepsis and MODS, 25% cases died due to sepsis with ARDS, 25% cases died due to acute renal failure as a post-operative (kasai) complications.

25% cases were discharged successfully and 12.5% cases required re-admission.

Conclusion

The purpose of this study was to determine the clinical presentations and outcome associated with Dextrocardia. More than 70% 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. One should always search for other anomalies whenever a case of dextrocardia is diagnosed because extracardiac anomalies are not uncommon [5]. Sepsis is almost always present and fatal pneumococcal infection is more common in patients with asplenia syndrome [6]. Doctors should encourage routine medical physical examination for their patients, which could help identify this anomaly, thereby preventing wrong diagnosis and possibly death due to delay in management.

Limitations

As incidence is very rare, a few cases were found, so the result of this study whether can be extrapolated to larger population or not still remain unclear.

Abbreviations

ASD: Atrial septal defect, VSD: Ventricular septal defect, PS: Pulmonary stenosis, AVSD: Atrioventricular septal defect, CHD: Congenital heart disease, LRTI: lower respiratory tract infection, ECG: electrocardiogram, USG: ultrasonography and CT scan: computed tomography scan, OPD: outpatient department, MODS: multi-organ dysfunction syndrome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information's to be reported in journal. The patients understand that their names and initials will not be published and due efforts

will be made to conceal their identity, but anonymity cannot be guaranteed.

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Ethical Approval: The study was approved by the Institutional Ethical Committee

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