Original Article

Spectrum of Congenital Cardiac Malformations in Dextrocardia: A Single Institute Study

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Abstract

Introduction: Complex congenital heart malformations are associated with dextrocardia depending on the situs. The objective is to present the spectrum of cardiac malformations and the surgical interventions done in patients with dextrocardia at our tertiary care institute over 5 years. **Material and Methods:** A retrospective study of patients with dextrocardia undergoing some form of cardiac surgery at our institute from January 2012 to December 2016 was done. Seventeen cases were identified, and their records were reviewed. **Results:** Of 2231 cardiac surgeries done for congenital heart disease, there were 17 (0.76%) patients with dextrocardia with male predominance (70.58%). The mean age was 5.69 ± 6.66 years. Of 17 patients, 9 had situs inversus, 7 had situs solitus, and 1 patient had situs ambiguous. Thirteen patients (76.47%) had functional single-ventricle physiology. Six of them (46.15%) had situs solitus, 6 (46.15%) had situs inversus, and situs ambiguous in 1 (7.69%). Biventricular repair was possible in 4 patients (23.52%), of which 3 had situs inversus and 1 had situs solitus. Three patients had ventriculoarterial (VA) discordance and 2 had atrioventricular and VA discordance. There were two early mortalities and three late mortalities, all of them had single-ventricle physiology. **Discussion and Conclusion:** Dextrocardia is a rare congenital anomaly with wide variations of complex congenital cardiac malformations. It is associated with all forms of spatial relationships of visceral organs with situs inversus being the most common.

Keywords: *Congenital heart disease, dextrocardia, single ventricle, situs*

Introduction

Dextrocardia refers to the location of the heart in the right hemithorax, resulting from abnormal coordination among genes responsible for lateralization of organs in the right-left axis during embryonic development.[1] It is a rare congenital cardiac anomaly with incidence reported between 1 in 10,000 and 12,000 live births.[2,3] Dextrocardia occurs with all the three configurations, i.e., situs solitus, situs inversus, and situs ambiguous. It is more commonly associated with situs inversus which being mirror image of situs solitus with levocardia. The incidence of additional congenital cardiac malformations, however, is higher in dextrocardia with situs solitus than with situsinversus, i.e., 5% with situs inversus and 90% with situs solitus.[3-5] The spectrum of cardiac anomalies associated with dextrocardia is wide. We, in this retrospective study, present the range of cardiac anomalies and surgical interventions done in patients with dextrocardia of different age groups presenting at our tertiary care center, Post Graduate Institute of Medical Education and Research (PGIMER), Chandigarh, from January 1, 2012, to December 31, 2016.

Material and Methods

The study was approved by the institutional ethics committee. Our institute's database containing 2231 surgeries done for congenital heart disease in the Department of Cardiothoracic and Vascular Surgery, PGIMER, Chandigarh, between January 1, 2012, and December 31, 2016, was searched for the patients with dextrocardia. There were 17 patients with dextrocardia who had undergone some form of cardiac surgery for cardiac anomalies at our institute. The records of these 17 patients (0.76%) were reviewed for their demographic data (age, sex, weight, height, and body surface area), diagnosis from echocardiography/catheterization data/computed tomography angiography/intraoperative findings, surgery done whether emergency or elective,

How to cite this article: Kumar V, Singh RS, Mishra AK. Spectrum of congenital cardiac malformations in dextrocardia: A single institute study. J Anat Soc India 2020;68:279-84.

Article Info

Received: 16 June 2019
Accepted: 08 January 2020
Available online: 28 February 2020

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Access this article online

Website: www.jasi.org.in
DOI: 10.4103/JASI.JASI_69_19
Quick Response Code:

postoperative hospital stay, and intensive care unit (ICU) stay.

Dextrocardia was defined as the location of greater mass of the heart in the right hemithorax with its base to apex axis pointing toward right. Visceroatrial arrangement was defined situs solitus (which is normal position of the heart) when the inferior vena cava (IVC) was on the right side of spine draining into systemic morphological right atrium which was present to the right of morphological left atrium, morphological right bronchus was concordant with trilobed right lung, morphological left bronchus was concordant with bilobed left lung, stomach on the left side and liver on the right side [Figure 1]. Mirror image of situs solitus was defined situs inversus [Figure 2]. Any visceroatrial arrangement not fitting into these two categories was defined situs ambiguous.

Statistical analysis

The analysis was performed using SPSS version 21 (IBM, Armonk, New York, USA). Categorical data were reported as percentage (%) and frequency and data for continuous variables were summarized using mean ± standard deviation.

Results

Demographic data

Table 1 lists the morphological details of the patient. The mean age was 5.69 ± 6.66 years ranging from 37 days preterm child (gestation age 35 weeks) to 18 years. Twelve out of 17 patients were male (70.58%) and 5 were female (29.42%). The mean body surface area was 0.66 ± 0.44 m^2. Of 17 patients, 9 had situs inversus, 7 had situs solitus, and 1 patient had situs ambiguous.

Cardiac anomalies

Table 2 lists the congenital cardiac anomalies according to the dominant diagnosis associated with dextrocardia. Coronaries were normal in all cases.

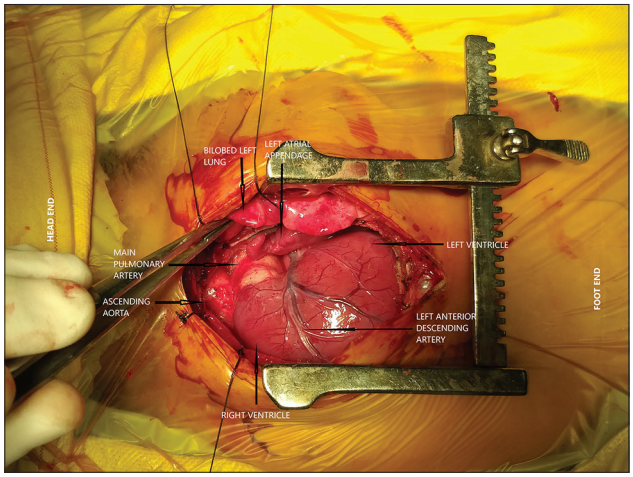

Figure 1: Intraoperative picture of dextrocardia with situs solitus showing morphological left ventricle anterior and to the left of the morphological right ventricle with normally related great arteries. Left lung is bilobed

Univentricular hearts

Thirteen of 17 (76.47%) patients had functionally single-ventricle physiology. Six of those 13 (46.15%) had situs solitus, 6 (46.15%) had situs inversus, and 1 case of situs ambiguous (7.69%). Three of those 13 patients had ventriculoarterial (VA) discordance, i.e., transposition of great arteries (2 with situs inversus and 1 with situs solitus) and 2 had atrioventricular (AV) and VA discordance (i.e., congenitally corrected transposition of great arteries) with situs inversus. Rest of the 8 patients had AV and VA concordance (i.e., normal relation). Three cases had malposed great arteries, four patients had bilateral superior vena cava (SVC) and IVC drained into the right atrium normally in all cases. There was no case of isomerism. There was one case of bilateral patent ductus arteriosus (PDA) associated with tricuspid atresia. Ventricular septal defect (VSD) was present in 16 cases, whereas ostium secundum atrial septal defect (OS-ASD) was present in 6 patients and ostium primum (OP)-ASD was present in two patients. The common atrium was present in one case of double-inlet left ventricle with double-outlet left ventricle.

Biventricular hearts

Four of 17 (23.52%) patients had two functional ventricles of which three patients had situs inversus and 1 had situs solitus. The definitive intracardiac repair was done in all the patients. Three patients had tetralogy of Fallot (TOF) with pulmonary stenosis (PS) and 1 had coarctation of the aorta (CoA) with hypoplastic distal arch with PDA. Bilateral SVC was present in one case of TOF with PS. PDA was present in one case of CoA only. There was AV and VA concordance in all the cases. AV valves were normal and were separate. VSD was present in all the cases; though, it was small muscular in a patient of CoA. Small OS-ASD was associated with CoA only.

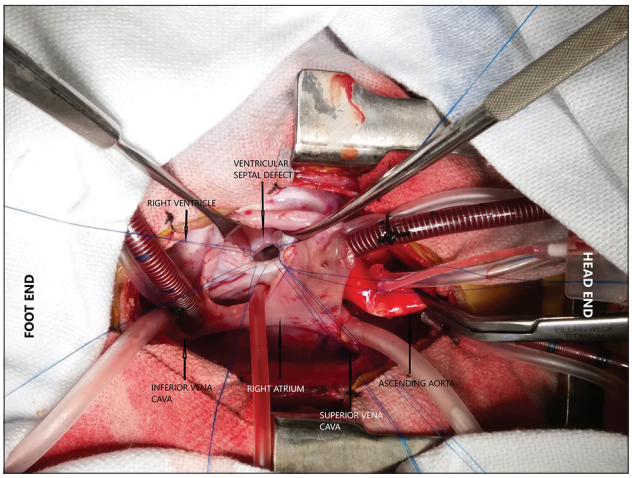

Figure 2: Intraoperative picture of dextrocardia with situs inversus showing morphological right atrium and right ventricle on the left side. Vena cavae (cannulated) are draining into the morphological right atrium. Ascending aorta (cannulated and cross clamped) is on the left side. Ventricular septal defect is visible after retraction of tricuspid leaflets. Small part of morphological right ventricle is also visible. Left ventricle is lying posterior and to the right of the morphological right ventricle is not visible

Table 1: Morphological characteristics of the patients

Characteristics	Situs inversus, *n* (%)	Situs solitus, *n* (%)	Situs ambiguous, *n* (%)	Total, *n* (%)
Number of patients	9 (52.94)	7 (41.17)	1 (5.88)	17
Age (years) (mean)	5.27±6.62	7.73±7.23	2/12	5.69±6.66
Gender				
Male	4 (33.33)	7 (58.33)	1 (8.33)	12 (70.5)
Female	5 (100)	0	0	5 (29.41)
Dominant ventricle				
Right	6 (66.67)	3 (33.33)	0	9 (52.94)
Left	0	3 (75)	1 (25)	4 (23.52)
Biventricle	3 (75)	1 (25)	0	4 (23.52)
SVC				
Single	7 (53.85)	5 (38.46)	1 (7.69)	13 (76.47)
Bilateral	2 (50)	2 (50)	0	4 (23.52)
AV valve				
Common	1 (25)	2 (50)	1 (25)	4 (23.52)
Atresia	0	1 (100)	0	1 (5.88)
AV discordance	2 (100)	0	0	2 (11.76)
VA discordance	2 (66.67)	1 (33.33)	0	3 (17.65)
Pulmonary valve				
Stenosis	8 (57.14)	5 (35.71)	1 (7.14)	14 (82.35)
Atresia	1 (50)	1 (50)	0	2 (11.76)
PDA				
Unilateral	3 (60)	2 (40)	0	5 (29.41)
Bilateral	0	1 (100)	0	1 (5.88)
VSD	9 (56.25)	6 (37.5)	1 (6.25)	16 (94.11)
ASD	3 (37.5)	4 (50)	1 (12.5)	8 (47.06)
Malposed great arteries	0	3 (100)	0	3 (17.65)

ASD: Atrial septal defect, AV: Atrioventricular, PDA: Patentductusarteriosus, SVC: Superior vena cava, VA: Ventriculoarterial, VSD: Ventricular septal defect

Table 2: Dominant caridac anomaly according to situs

	Situs inversus	Situs solitus	Situs ambiguous
TOF	3	1	0
Double-outlet right ventricle	1	2	0
dTGA	2	1	0
ccTGA	2	0	0
AV canal defect	1	0	1
Tricuspid atresia with pulmonary atresia	0	1	0
CoA with PDA	0	1	0
DILV	0	1	0
DOLV	0	1	0

ccTGA: Congenitally corrected transposition of great arteries, CoA: coarctation of aorta, dTGA: d-transposition of great arteries, PDA: Patent ductusarteriosus, TOF: Tetralogy of fallot, DILV: Double-inlet left ventricle, DOLV: Double-outlet left ventricle

Surgical interventions

Table 3 lists the surgical interventions done in all the 17 cases of dextrocardia, according to situs.

Three of 17 patients had previously undergone modified Blalock-Taussig (BT) shunt, (i.e., connecting left sublcavian artery to the left pulmonary artery through a polytetrafluoroethylene (PTFE) tube graft). Out of those three two underwent second stage Bidirectional (BD) Glenn (anastomosis of SVC to the right pulmonary artery), and one was converted to fenestrated lateral tunnel Fontan procedure. Two of those previous modified BT shunts were not patent at the time of surgery. Five of 17 patients underwent primary modified BT shunt and 1 central shunt using PTFE tube grafts (Gore-Tex, W. L. Gore and Associates, Inc., Flagstaff, AZ) of which 2 modified BT shunts and central shunt were done in emergency. Three out of five modified BT shunts were performed via median sternotomy one of them also required PDA clipping and the rest of the two shunts were done through posterolateral thoracotomy. BD Glenn shunt on cardiopulmonary bypass without arresting heart was done in five cases and in one of them left pulmonary artery origin stenosis was augmented with pericardial patch. Two out of 17 patients underwent fenestrated lateral tunnel Fontan procedure.

Four (23.52%) patients underwent definitive biventricular repair. Three of them underwent intracardiac repair with transannular autologous pericardial patch augmentation of right ventricle outflow tract for TOF with PS and one had resection of coarct segment with extended end-to-end

Table 3: Cardiac surgical interventions done in the patients in cohort

Serial number	Age	Sex	Situs	Diagnosis	Previous surgery	Surgical procedure	Result
				Two ventricles			
1	4.5 years	Female	Inversus	TOF, severe PS, small pulmonary annulus and LPA	Nil	TOF repair with TAPP extending into LPA	Alive
2	1 year	Female	Inversus	TOF, severe PS, small pulmonary annulus	Nil	TOF repair with transannular patch	Alive
3	15 years	Female	Inversus	TOF, severe PS small pulmonary annulus, bilateral SVC	Nil	TOF repair with transannular patch	Alive
4	10 days	Male	Solitus	CoA with hypoplastic distal arch with PDA, small ASD, small VSD	Nil	Resection of coarct segment with extended EEA with PDA ligation	Alive
				Single ventricle			
5	2 months	Male	Inversus	dTGA, VSD, severe PS, hypoplastic LV, small PDA, bilateral SVC, recurrent cyanotic spells	Nil	Emergency left modified BT shunt with PDA clipping via median sternotomy	Died after 2 months
6	1 year	Female	Inversus	dTGA, VSD, severe PS, hypoplastic LV	Nil	BD glenn under CPB without cardiac arrest	Alive
7	18 years	Male	Inversus	DORV, noncommitted VSD, severe PS	Left modified BT shunt	BD glenn under CPB without cardiac arrest	Alive
8	2 years	Male	Inversus	ccTGA, VSD, severe PS, hypoplastic LV	Right modified BT shunt	BD glenn under CPB without cardiac arrest	Alive
9	5 years	Female	Inversus	Complete unbalanced AVCD, DORV, LV hypoplastic, small PDA, LPA origin stenosis	Nil	BD glenn with pericardial patch augmentation of LPA under CPB without cardiac arrest	Alive
10	8 months	Male	Inversus	ccTGA, VSD, pulmonary atresia, small PDA, hypoplastic LV	Nil	Right modified BT shunt via right posterolateral thoracotomy	Alive
11	15 years	Male	Solitus	DORV, noncommitted VSD, PS, small ASD, malposed great arteries	Modified left BT shunt	Fenestrated lateral tunnel fontan	Alive
12	1 year	Male	Solitus	TOF, pulmonary atresia, hypoplasticrv, small central pulmonary arteries	Nil	Modified left BT shunt via modified posterolateral thoracotomy	Died after 2 years
13	17 years	Male	Solitus	DORV, VSD, PS, malposed great arteries, LV hypoplastic, recurrent cyanotic spells and recurrent VT	Nil	Emergency central shunt between ascending aorta and LPA via median sternotomy	Died after 7 months
14	37 days (preterm 35 weeks)	Male	Solitus	TA, PA, VSD, ASD, bilateral PDA, malposed great arteries on PGE1 infusion	Nil	Emergency modified right BT shunt through median sternotomy	Died on pod 4 due to sepsis and LCOS
15	10 years	Male	Solitus	dTGA, VSD, PS, hypoplastic LV	Nil	BD glenn under CPB without cardiac arrest	Alive
16	2 months	Male	Ambiguous	Complete unbalanced AVCD, hypoplastic RV, severe PS, bilateral SVC, recurrent cyanotic spells	Nil	Emergency modified right BT shunt through median sternotomy	Died on POD 2 due to LCOS

Contd...

Table 3: Contd...

Serial number	Age	Sex	Situs	Diagnosis	Previous surgery	Surgical procedure	Result
17	11 years	Male	Solitus	DILV, DOLV, common AV valve, hypoplastic RV, bilateral SVC	Nil	Fenestrated lateral tunnel fontan	Alive

ASD: Atrial septal defect, AV: Atrioventricular, AVCD: AV canal defect; BD: Bidirectional, BT: Blalock-Taussig, ccTGA: Congenitally corrected transposition of great arteries, CoA: Coarctation of aorta, CPB: Cardiopulmonary bypass, DILV: Double-inlet left ventricle, DOLV: Double outlet left ventricle, DORV: Double-outlet right ventricle, dTGA: d-transposition of great arteries, EEA: End-to-end anastomosis, LCOS: Low cardiac output syndrome, LPA: Left pulmonary artery, LV: Left ventricle, PDA: Patent ductusarteriosus, PGE1: Prostaglandin E1, POD: Postoperative day, PA: Pulmonary atresia, PS: Pulmonary stenosis, RV: Right ventricle, SVC: Superior vena cava, TA: Tricuspid atresia; TAPP: Transannular pericardial patch, TOF: Tetralogy of Fallot, VSD: Ventricular septal defect, VT: Ventricular tachycardia

anastomosis with PDA ligation. Surgical revisions were not required in any patient.

Mortality

There were two mortalities of 17 (11.76%) patients with dextrocardia. One was a preterm (35 weeks gestation) 37 days male child diagnosed with situs solitus, dextrocardia, tricuspid atresia, pulmonary atresia, small confluent pulmonary arteries, bilateral PDA, malposed great vessels on prostaglandin E1 infusion with recurrent cyanotic spells. Emergency right modified BT shunt was done with 3-mm PTFE tube graft through median sternotomy. However, the patient died on the 3rd postoperative day due to low cardiac output syndrome and septicemia. Another was a 2 months male child with situs ambiguous, dextrocardia, complete unbalanced AV septal defect with severe PS, hypoplastic right ventricle, recurrent cyanotic spells without PDA. Modified right BT shunt was done with a 3.5-mm PTFE tube graft. The child died of low cardiac output syndrome on the 2nd postoperative day.

Postoperative hospital stay

The mean postoperative hospital stay for the alive 15 cases was 10.93 ± 3.76 days. Table 4 lists the mean postoperative hospital stay according to the situs and functionality of the ventricles. For patients with univentricular physiology mean stay was 12 ± 3.79 days, while for the patients with biventricular repair, it was 8 ± 1.41 days.

Intensive care unit stay

Table 5 lists the mean ICU stay according to the situs and functionality of the ventricles. Mean ICU stay for the alive 15 cases was 5.06 ± 2.55 days. Biventricular hearts had mean ICU stay of 4.5 ± 1.73 days, while for univentricular hearts, it was 5.27 ± 2.83 days.

Discussion

Dextrocardia is a rare congenital cardiac anomaly characterized by the location of the heart in the right side of the chest cavity. Dextrocardia is more commonly associated with situs inversus than situs solitus or situs ambiguous as reported in many series. However, cardiac malformations are more commonly associated with situs solitus.[1,3,6-8] Situs inversus with dextrocardia is more likely to occur with structurally normal heart with normal life expectancy and thus may escape recognition.[1] However, in our study, 52.94% of the patients that were operated for congenital heart disease had situs inversus as compared to 41.18% of patients with situs solitus and 5.88% with situs ambiguous. This is in contrast to the retrospective study by Bohun *et al*.,[6] in which out of 43 patients who were surgically treated, 46.5% had situssolitus, 23.25% situs inversus and 30.23% had situs ambiguous. Similarly, offen *et al*.[9] also found situs solitus (74%) more commonly associated with cardiac malformations in the adult patients with dextrocardia. This difference is may be because our study deals with the patients who were referred to our institute and underwent some form of cardiac surgery and therefore may have selection bias for the patients who can be surgically treated.

Dextrocardia is frequently associated with univentricular hearts with its prevalence reported from 11% to 25% in different series.[2,3,6,10] In our study, functionally univentricular hearts were present in 13 of 17 (76.47%) patients. Six of those 13 (46.15%) had situssolitus, 6 (46.15%) had situsinversus, and 1 case of situs ambiguous (7.69%). Similar findings were seen in many other studies.[6,7,11] In a study by Bohun *et al*.[6] of 81 patients with dextrocardia, 17 had univentricular hearts (20.98%). Of those 17 patients, 9 were associated with situssolitus only 1 had situsinversus and rest 7 had situs ambiguous. Similarly, among adult patients with univentricular physiology in Offen *et al*.'s[9] study, 81.81% had situs solitus and 18.18% had situs inversus. Poh *et al*.[11] also reported situs solitus in 41.5% of patients with dextrocardia in functionally single-ventricle hearts as compared to only 12.2% with situs inversus.

Biventricular repair was possible in 4 patients of 17 (23.52%) in our cohort. Three (75%) of them had situs inversus and 1 (25%) had situs solitus, which is consistent with the study of Offen *et al*.,[9] in which 8 of 19 (42.1%) patients underwent two ventricle repair and had situs inversus in 5 of those 8 (62.5%) patients as compared to 3 (37.5%) with situs solitus.

Most of the patients in our study underwent primary shunt procedure (modified BT shunt in five and central

Table 4: Postoperative hospital stay (mean days±standard deviation)

	Situs solitus	Situs inversus	Total
Univentricular repair	12±5.54	11.33±1.75	12±3.79
Biventricular repair	7	8.33±1.53	8±1.41
Total	11.83±5.49	10.33±2.18	

Table 5: Intensive care unit stay (mean days±standard deviation)

	Situs solitus	Situs inversus	Total
Univentricular repair	6.8±3.6	4±1.26	5.27±2.83
Biventricular repair	3	5±1.73	4.5±1.73
Total	6.17±3.54	4.33±1.33	

shunt in 1), five underwent BD Glenn, 3 underwent intracardiac repair for TOF, 2 underwent Fontan operation, and 1 had CoA repair. All patients but one had decreased pulmonary blood flow with stenosis in 13 (76.47%) and pulmonary atresia in 3 (17.64%). Three patients had previously undergone BT shunts which were converted to BD Glenn in two patients and one had Fontan operation. Only 1 patient underwent primary Fontan operation. Most studies, however, report an increased number of Fontan procedures in patients with dextrocardia with functional single ventricle physiology.[6,7,11] This discrepancy can be explained by the fact that our study is over 5 years only and most of the patients with univentricular hearts have presented at age <2 years (n = 8). Other retrospective studies had their data spanned over >10 years and most of those Fontan operations were completion procedures.

Limitations

The study is limited by its being a retrospective study of a single center with the small number of patients included over a small period of 5 years. It includes only those patients which were referred to our center which could undergo some form of cardiac surgery, therefore, having selection bias. Hence, our study cannot be generalized to patients with isolated dextrocardia.

Conclusion

Our study supported the fact that dextrocardia is a rare congenital anomaly with wide variations of complex congenital cardiac malformations. It is associated with all forms of spatial relationships of visceral organs with situs inversus being the most common. Univentricular physiology is more commonly associated with dextrocardia and thus, most of the surgeries performed are palliative in nature; however, if possible, biventricular repair can also be performed. Considering such a wide spectrum of congenital cardiac anomalies and that too complex, dextrocardia can be called Pandora's box. This retrospective study is from a single center with the small number of patients, larger collaborative study spanned over longer period including multiple centers may be required to further characterize the patients and their outcome.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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