Cardiovascular Topics

A retrospective review of right-sided hearts at a South African tertiary hospital

Nadia Beringer, Antoinette Cilliers

Abstract

Background: A right-sided heart (RSH) has three main causes: dextrocardia, dextroposition and dextroversion. It may be associated with cardiac malformation, extra-cardiac abnormalities and adverse patient outcomes. The aim of this study was to determine the prevalence, demographics, associated cardiac malformations (ACM) and outcome of paediatric patients diagnosed with a RSH at a South African tertiary hospital.

Methods: A retrospective review was performed over a 22-year period.

Results: RSH comprised 1% of the paediatric cardiology referrals. Dextrocardia was the most frequent cause (58.1%) and the majority of these patients had ACM (81.5%). More than a third (40.9%) were diagnosed with dextroposition, secondary to extra-cardiac factors. Dextroversion was the least common cause (1.1%). Over a quarter of all patients were confirmed dead at the time of the study; most had been diagnosed with dextrocardia. Two-thirds of the patients were lost to follow up.

Conclusion: A RSH is an unusual occurrence. Dextrocardia, the most common cause, is frequently associated with ACM and extra-cardiac abnormalities. It is therefore important to timeously elucidate the cause of a RSH.

Keywords: right-sided hearts, dextrocardia, dextroposition, dextroversion

Submitted 25/5/17, accepted 22/10/18
Cardiovasc J Afr 2018; 29: online publication www.cvja.co.za
DOI: 10.5830/CVJA-2018-051

A right-sided heart (RSH) is a cardiac malposition that is predominantly caused by dextrocardia, dextroposition or dextroversion. Each cause has a different aetiology, associated cardiac malformations (ACM) and outcome. ACM range from severe cyanotic congenital heart defects to sub-clinical ductal lesions.

Dextrocardia is the most common cause of a RSH. The reported incidence ranges from 0.01% of all live births in high-income countries to a slightly higher incidence of 0.35% in low-income countries. It occurs when the base-apex of the heart is displaced to the right. Factors intrinsic to the heart are responsible for this malformation, which occurs during foetal embryogenesis. However, no specific genetic cause or inheritance pattern for dextrocardia has been identified.

Dextroposition is the displacement of the heart to the right hemi-thorax, secondary to extra-cardiac causes. It is not usually associated with ACM.

Dextroversion results from a congenital malrotation of the ventricular part of the heart about its long axis with the atria remaining in the normal position. It is essentially dextrocardia with normally related atria and viscera. ACM are very common and mostly involve the cono-truncal region of the heart.

The term situs is not only used to describe the atrial position, but also the anatomical position of the abdominal and thoracic organs. There are three types of situs described associated with a RSH: situs solitus, situs inversus and situs ambiguous. Situs solitus refers to the normal position of the atria, thoracic and abdominal organs. The complete mirror image of this relationship is situs inversus.

When situs cannot be determined anatomically, it is referred to as situs ambiguous. This is also known as heterotaxy or isomerism and is invariably coupled with severe cardiac and non-cardiac malformations. Situs ambiguous is further divided into right atrial isomerism (RAI) and left atrial isomerism (LAI). Non-cardiac malformations include: asplenia in RAI and polysplenia and biliary atresia in LAI. Malrotation of the gut can occur in both.

The incidence and severity of complex cardiac anomalies are directly proportional to the failure of shift of the cardiac apex with regard to the abdominal and thoracic organs. In other words, situs solitus dextrocardia is more likely to be associated with cardiac malformations, compared to situs inversus dextrocardia.

Failure to diagnose a RSH and its associations may impact on patient outcomes. To date, there are no publications from the African continent describing RSH: its overall prevalence,
underlying causes, ACM, therapeutic interventions, and morbidity and mortality rates. Thus follows a retrospective review of all the children with RSH presenting to a large tertiary-care hospital in southern Africa.

Methods

A retrospective review of children diagnosed with RSH at the Chris Hani Baragwanath Academic Hospital (CHBAH) was undertaken. The study spanned a 22-year period and patient records were obtained from the electronic database of the CHBAH cardiology department.

Data collected included age at diagnosis, gender, underlying cause of the RSH, prevalence, ACM, extra-cardiac abnormalities, situs, interventions and patient outcome at the time of the study. We used chest roentgenogram, echocardiography, electrocardiogram, cardiac catheterisation and foetal ultrasonography either alone or in combination to diagnose the RSH. ACM were grouped according to the diagnostic categories described by DC Fyler and published in the New England Regional Infant Cardiac Program in 1980.15

Permission to conduct retrospective analyses was obtained from the Human Research Ethics Committee of the University of the Witwatersrand.

Statistical analysis

Descriptive statistical analysis was performed. The Chi-squared test, unpaired Student’s t-test and Mann–Whitney U-test were employed. A p-value < 0.05 was used as the level of significance. Data were collected and managed using REDCap (Research Electronic Data Capture)16 and were analysed using Microsoft Excel and Graphpad Prism. REDCap is a secure, web-based application designed to support data capture for research studies, hosted at the University of the Witwatersrand.16

Results

There were 18 870 paediatric patients referred for cardiac assessment between 1 January 1991 and 2 November 2012. One hundred and eighty-six children were found to have RSH. This comprised 1% of the total paediatric cardiology referrals seen during the study period.

Of the 186 patients with RSH, 108 were diagnosed with dextrocardia as the underlying cause. A further 76 patients had dextroposition and only two had a confirmed diagnosis of dextroversion. The extra-cardiac causes of dextroposition are described in Fig. 1.

The median age at diagnosis of a RSH was two months (range, prenatal to 16 years). The majority of diagnoses were made before one year of age (144 out of 186 patients, 77.4%).

There were 97 male patients and 83 female patients. The gender of six neonates was not documented. The male-to-female ratio (180 patients) was 1:0.86 (53.9% male, 46.1% female).

ACM

Eighty-eight out of the 108 patients (81.5%) with dextrocardia demonstrated ACM. The various ACM are listed in Table 1.

Of the 76 patients with dextroposition, four demonstrated ACM (5.3%). An atrial septal defect secundum was diagnosed in one patient and a large patent ductus arteriosus (PDA) in another. A further two patients in this subgroup demonstrated a small PDA in association with a hypoplastic right lung and Scimitar syndrome. There were 11 patients with dextroposition secondary to congenital diaphragmatic hernias (14.5%), none of whom had documented ACM. One out of the two patients in this subgroup had transposition of the great arteries (TGA).

Situs

Fifty-two patients with dextrocardia (Fig. 2) exhibited situs inversus (48.1%) and ACM was diagnosed in 32 of them (61.5%). Situs solitus was found in a further 24 patients (22.2%), 15 of whom demonstrated ACM (62.5%). There was

<table>
<thead>
<tr>
<th>Table 1. Dextrocardia: associated cardiac malformations</th>
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<tr>
<td>Associated cardiac malformations</td>
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<tr>
<td>Heterotaxias</td>
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<tr>
<td>Single ventricle</td>
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<tr>
<td>Hypoplastic left ventricle</td>
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<td>Tricuspid atresia</td>
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<td>Double-outlet right ventricle</td>
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<tr>
<td>D-transposition of great arteries</td>
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<tr>
<td>L-transposition of great arteries</td>
</tr>
<tr>
<td>Endocardial cushion defect</td>
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<tr>
<td>Total anomalous pulmonary venous return</td>
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<tr>
<td>Tetralogy of Fallot</td>
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<tr>
<td>Coarctation of the aorta</td>
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<tr>
<td>Ventricular septal defect</td>
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<tr>
<td>Pulmonary stenosis</td>
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<tr>
<td>Atrial septal defect secundum</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
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<tr>
<td>No significant heart disease</td>
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<tr>
<td>Lung disease</td>
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<tr>
<td>Other (all other diagnoses)</td>
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</tbody>
</table>

Most patients demonstrated more than one ACM at the time of echocardiography.

Fig. 1. Extra-cardiac causes of dextroposition.
no statistical difference between the two situs arrangements and their associated cardiac malformations \( (p = 1.000) \).

Eighteen patients demonstrated situs ambiguous, with an incidence of 16.7%. In this subgroup, 17 had documented ACM (94.4%), five were found to have RAI and eight to have left LAI. The remaining five patients with situs ambiguous did not have their atrial isomerism defined. Fourteen patients in the dextrocardia subgroup had unknown situs arrangements (13%).

All of the 76 patients with dextroposition demonstrated situs solitus. This is in keeping with the underlying aetiology of dextroposition and involves external forces that may either be compressive or pulling in nature.

Of the two patients with dextroversion, one had situs solitus and one had situs inversus with TGA.

**Ventricular arrangements**

The majority of patients with dextrocardia had two ventricles (84.3%), while 17 had a univentricular heart (15.7%). Over two-thirds of the patients demonstrated atroventricular (AV) concordance, 10 had AV discordance, and the AV relationship of 25 patients was not recorded. Sixty-three patients demonstrated ventriculo-arterial (VA) concordance, 11 had VA discordance, and the VA relationship of 22 patients was not recorded.

All the patients with dextroposition had normal AV and VA relationships.

Both the patients diagnosed with dextroversion had two ventricles and AV concordance. One had VA discordance (TGA) while the other patient had a normal heart.

**Extra-cardiac abnormalities**

A total of 48 patients with dextrocardia had associated extra-cardiac abnormalities (44.4%), eight of whom (16.7%) were diagnosed with Kartagener syndrome (situs inversus totalis, chronic sinusitis and bronchiectasis). Two out of the five patients diagnosed with dextrocardia and RAI were confirmed to have asplenia on computerised tomography of the abdomen, and one patient had gastric outlet obstruction. There were eight patients diagnosed with LAI, three had an interrupted inferior vena cava, one patient had heart block, while duodenal atresia was diagnosed in one other.

**Surgery**

Cardiac surgery was performed in 25 of the patients with dextrocardia (23.1%). Eight of these procedures were corrective (32%), while 17 were palliative (68%). Corrective surgeries included ligation of a PDA, closure of a ventricular septal defect (VSD) and/or pulmonary artery (PA) banding. Palliative procedures included Blalock–Taussig or Glenn shunts. Nine patients with dextrocardia underwent general gastrointestinal (GIT) surgeries (Table 2). Four patients in this subgroup demonstrated isomerism (44.4%).

Twenty-nine patients with dextroposition underwent surgical procedures. Seven were diagnosed with Scimitar syndrome and had percutaneous occlusion of the anomalous feeder arteries from the descending aorta. A wide variety of non-cardiac surgeries were undertaken in the remaining patients with dextroposition (Table 3).

**Patient outcomes**

Twenty-one (19.4%) of the patients with dextrocardia were reported dead at the time of the study. All had complex ACM, which included: hypoplastic left ventricle, tricuspid atresia, double-outflow right ventricle, and single atrium and ventricle. Five had previously undergone cardiac surgery.

Nine out of the 21 patients had situs inversus (42.9%), six had situs solitus (28.6%) and five had situs ambiguous (23.8%). In two patients the situs was undefined.

Infection was the documented cause of death in seven patients (33.3%). Twenty-two patients (20.4%) were confirmed alive, and 65 (60.2%) had been lost to follow up.

Eight patients with dextroposition had died at the time of the study (10.5%), two of whom had Scimitar syndrome. Eleven were found to be alive (14.5%), while the majority of patients were lost to follow up (75%).

In the dextroversion subgroup, neither of the patients underwent surgery and both were alive at the time of the study.
Discussion
The study results are in keeping with published literature that describes a RSH as an uncommon entity.1,3 RSH comprised only 1% of the total number of paediatric patients referred for cardiac assessment at CHBAH over a 22-year period.

The majority of patients were diagnosed with dextrocardia. There was no statistical significance between dextrocardia, the two most common situs arrangements and ACM. This differs from other published data that have demonstrated an association between dextrocardia, situs arrangements and ACM. As this was a retrospective audit, pertinent patient information was not obtained in some cases. Therefore it is possible that some of the study patients had undocumentated ACM.

Eight of the study patients with situs inversus dextrocardia without ACM were confirmed to have Kartagener syndrome.2,9 This highlights the importance of including a ciliopathy in the differential diagnosis of patients with recurrent respiratory infections and dextrocardia.

In this study, 7% of patients with situs inversus dextrocardia and 22% of patients with situs ambiguous dextrocardia underwent GIT surgery. Patients with situs inversus totalis can present with abdominal signs and symptoms that differ from patients with situs solitus. For example, appendicitis in patients with situs inversus totalis and midgut rotation can present with left lower-quadrant pain. Patients with situs ambiguous are at risk of malrotation of the bowel and consequently bowel ischaemia.12 This highlights the importance of timeous recognition of different situs arrangements. Furthermore, as seen in the study, RAI is associated with asplenia, placing these patients at an increased risk for serious bacterial infections.12

The prevalence of situs ambiguous dextrocardia (16.7%) was found to be similar to that previously reported.4,4,7 In the study, both the RAI and LAI groups demonstrated some of the more common serious cardiac and non-cardiac anomalies (in particular GIT defects) requiring specialised paediatric surgical services. These anomalies are in keeping with previously published literature.12 The mortality rate was also in keeping with published data, and was higher in the RAI group compared to the LAI group.7,8,12

The risk for congenital heart lesions in patients with dextroposition is reportedly the same as for the general population.1,12,15 This was supported in the study results. Over a quarter of the patients were diagnosed with Scimitar syndrome, which is an unusual cause of dextroposition and is often associated with cardiac lesions.12,20

The diagnosis of dextroversion is sometimes missed and erroneously interchanged with dextroposition.

Conclusion
This study confirms the rarity of RSH, which are associated with both cardiac and non-cardiac anomalies and requires a high index of suspicion to make the diagnosis. Appropriate assessment will allow for a streamlined approach and correct management.

References