Original article

Influence of apical position on the left ventricular outflow tract obstruction in congenitally corrected transposition

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A B S T R A C T

Background: The right ventricle has a proclivity to wrap around the left ventricle outflow tract (LVOT) in congenitally corrected transposition (CCT) patients with apicocaval ipsilaterality, which may influence the outcome of the double switch operation (DSO). The goal of this study was to determine if the LVOT is compressed by the right ventricle in this setting.

Methods: A total of 103 patients with CCT were divided into four groups according to ventricular looping and apical position, including Group A (D-loop and levocardia), Group B (L-loop and dextrocardia), Group C (D-loop and dextrocardia), and Group D (L-loop and levocardia). Computed tomography was used to define left-right laterality and ventro-dorsal relationship of the LVOT.

Results: Apicocaval ipsilateral was found in 57 patients (Group A, n = 25; Group B, n = 32), in whom the right ventricle was found to wrap around the LVOT. Among them, 49 (86%) had LVOT obstruction. In 46 patients without apicocaval ipsilaterality (Group C, n = 10; Group D, n = 36), 31 had LVOT obstruction (67.4%). LVOT obstruction was more prone to occur in patients with apicocaval ipsilaterality compared with those without (p = 0.025), and was more significant in the situs solitus (p = 0.058) than in situs inversus (p = 0.547).

Conclusions: LVOT obstruction was more prone to occur in CCT patients with situs solitus and apicocaval ipsilaterality (Group B). The ventricular outflow tract was influenced by apical position, which should be considered to avoid a posterior ventricular outflow tract from compression after DSO.

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Introduction

The apical position has been considered inconsequential to the segmental analysis of congenital cardiac defects [11]; thus, its clinical significance has been neglected, with the exception of its role in the physical examination, and interpretation of the electrocardiogram (ECG) [2] and echocardiography. Based on the diversity of procedures for complex congenital heart disease performed at our tertiary referral center, the apical position appeared to be of crucial importance not only in defining the coronary artery pattern in the posterior atrioventricular groove [3], but also in planning the preoperative surgical approach to the intracardiac structures.

The double switch operation (DSO) has been advocated as an anatomical repair for congenitally corrected transposition (CCT); however, the ventricular outflow tracts are not spatially switched simultaneously [4–12]. The systemic right ventricle has a proclivity to wrap around the left ventricle outflow tract (LVOT) in CCT patients with apicocaval ipsilateral, which may influence the options and outcomes of DSO. To perform DSO on a normal heart in animal models to test this hypothesis is feasible but rather complicated, and it is impossible to imitate apicocaval ipsilateral. So we chose to observe the incidence of LVOT obstruction in various settings of CCT patients created naturally. Thus, the goal of this study was to assess to what extent the LVOT was compressed by the systemic right ventricle in CCT patients with and without apicocaval ipsilateral.

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upper panel of Fig. 1. In Group C (D-loop with dextrocardia) and Group D (L-loop with levocardia) patients, after ventricular looping and apical rotation, the apex and the inferior vena cava are positioned contralaterally. Hence, apicocaval ipsilaterality will not present in Group C and Group D patients (as shown in the lower panel of Fig. 1). The other synonym of apicocaval ipsilaterality is discordance of atrial situs and position of the apex [12].

Electron beam computed tomography (Imatron C 150-L, South San Francisco, CA, USA) [13] was used with electrocardiographic gating to define the left-right and ventro-dorsal relationship of the ventricular outflow tracts. All images were taken at the end-diastolic phase of the cardiac cycle. The slice thickness chosen was either 1.5 mm (for neonates) or 3 mm.

Computed tomography was performed from the level of the great arteries to the cardiac apex. Nonionic iodinated contrast medium (2–3 ml/kg; Ultravist 370; Schering, Berlin, Germany) was delivered by a power injector. Patients under 5 years of age were sedated with chloral hydrate (50 mg/kg) beforehand.

Cardiac catheterization and angiogram were used to evaluate the LVOT obstruction (pulmonary stenosis or atresia) and the relationship between the two ventricular outflow tracts. The incidence of obstruction in each group was compared by chi-square or Fischer exact test, as appropriate.

Results

Age was 15.49 ± 13.41 years, with a median of 11.91 years. There was no statistical difference among the four groups of patients. Apicocaval ipsilaterality was found in 57 patients (Group A, n = 25; Group B, n = 32), in whom the systemic right ventricle was found to wrap around LVOT (Fig. 2). Among these 57 patients, 49 patients (86%) had LVOT obstruction (pulmonary atresia or stenosis) (Table 1). In contrast, 67.4% of 46 patients without apicocaval ipsilaterality (Group C, n = 10; Group D, n = 36) had LVOT obstruction and the difference was significant (p = 0.025). In other words, patients with apicocaval ipsilaterality had a higher incidence of LVOT obstruction than those without apicocaval ipsilaterality (p = 0.025). This difference was even more significant in patients with situs solitus (Group B vs. Group D, p = 0.058) compared with those patients with situs inversus (Group A vs. Group C, p = 0.547) (Table 1). The positive predictive value was 83.96% and the positive likelihood ratio was 1.76 (Table 1). Although ventricular septal defect “alone” did not incur LVOT obstruction significantly (p = 0.606, patients with VSD vs. patients without VSD), the influence of apicocaval ipsilaterality on the LVOT obstruction was slightly modified by the presence of “concomitant” ventricular septal defect (Table 2).

Gross anatomy of autopsy in each patient with and without apicocaval ipsilaterality is shown in Fig. 3A and B. It is noteworthy that the “narrow” and “posterolateral” morphologically left ventricle was directed toward a hypoplastic pulmonary trunk in a case with apicocaval ipsilaterality (Fig. 3A, situs inversus and levocardia, Group A), and the inferior vena cava was close to the cardiac apex. In contrast, in a case without apicocaval ipsilaterality (Fig. 3B, situs solitus and levocardia, Group D), the “anterolateral” LVOT is more widely “open”.

Discussion

Anatomical repair is the procedure of choice for CCT patients with significant tricuspid regurgitation and conventional repair is usually satisfactory provided that there is no significant tricuspid regurgitation [5]. Hiramatsu et al. reported that the incidence of mortality, re-operation, and arrhythmia were lower in patients treated by arterial DSO compared with those treated by Rastelli DSO [6]. In contrast, Murtuza et al. reported a significantly higher

Methods

Our Institutional Review Board approved our study protocol and waived informed consent due to the retrospective nature of our study.

From 1981 to 2015, 103 consecutive patients with CCT, with their apex pivoted toward the right (dextrocardia) or toward the left (levocardia) in situs solitus or situs inversus, were included in this retrospective study. Patients with ambiguous atrial situs, univentricular atrioventricular connection, criss-cross hearts, or single outlet ventricles were excluded from this analysis.

As an evolution result of ventricular looping (D-loop vs. L-loop) and apical rotation (dextrocardia vs. levocardia), there are four subgroups of CCT (Fig. 1). In Group A (D-loop with levocardia) and Group B (L-loop with dextrocardia) patients, as a result of ventricular looping and apical rotation, the apex and the inferior vena cava are positioned ipsilaterally. Thus, apicocaval ipsilaterality will present in Group A and Group B patients (as shown in the
Fig. 2. Relationship of apical rotation (dextrocardia vs. levocardia) and ventricular looping (D-loop vs. L-loop), in patients with CCT, as viewed axially on computed tomography. The left column denotes CCT with situs inversus and D-loop ventricle, while the right column denotes CCT with situs solitus and L-loop ventricle. The upper panel denotes CCT with ACI, and the lower panel denotes CCT without ACI. ACI, apicocaval ipsilaterality; CCT, congenitally corrected transposition; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Table 1
Influence of apicocaval ipsilaterality on the left ventricular outflow tract obstruction in congenitally corrected transposition.

<table>
<thead>
<tr>
<th>Anatomical landmarks (N=103)</th>
<th>LVOTO (n=80)</th>
<th>No LVOTO (n=23)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACI (n=57)</td>
<td>49</td>
<td>8</td>
</tr>
<tr>
<td>Group A, situs inversus (n=25)</td>
<td>22</td>
<td>3</td>
</tr>
<tr>
<td>Group B, situs solitus (n=32)</td>
<td>27</td>
<td>5</td>
</tr>
<tr>
<td>No ACI (n=46)</td>
<td>31</td>
<td>15</td>
</tr>
<tr>
<td>Group C, situs inversus (n=10)</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Group D, situs solitus (n=36)</td>
<td>23</td>
<td>13</td>
</tr>
</tbody>
</table>

ACI, apicocaval ipsilaterality; LVOTO, left ventricular outflow tract obstruction. The Chi-square test:
\[ p = 0.025, \text{Group A and Group B with ACI vs. Group C and Group D without ACI}. \]
\[ p = 0.547, \text{Group A (D-loop and levocardia) vs. Group C (D-loop and dextrocardia)}. \]
\[ p = 0.058, \text{Group B (L-loop and dextrocardia) vs. Group D (L-loop and levocardia)}. \]

Table 2
Influence of apicocaval ipsilaterality on the left ventricular outflow tract obstruction could be modified by the presence of ventricular septal defect in congenitally corrected transposition.

<table>
<thead>
<tr>
<th>Anatomical landmarks (N=103)</th>
<th>(LVOTO, VSD)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>(+, +) (n=30)</td>
</tr>
<tr>
<td>ACI (n=57)</td>
<td>24</td>
</tr>
<tr>
<td>Group A, situs inversus (n=25)</td>
<td>13</td>
</tr>
<tr>
<td>Group B, situs solitus (n=32)</td>
<td>11</td>
</tr>
<tr>
<td>No ACI (n=46)</td>
<td>6</td>
</tr>
<tr>
<td>Group C, situs inversus (n=10)</td>
<td>1</td>
</tr>
<tr>
<td>Group D, situs solitus (n=36)</td>
<td>5</td>
</tr>
</tbody>
</table>

ACI, apicocaval ipsilaterality; LVOTO, left ventricular outflow tract obstruction; VSD, ventricular septal defect; +, presence; -, absence. The Chi-square test:
\[ p = 0.004, \text{Group A and Group B with ACI vs. Group C and Group D without ACI}. \]
\[ p = 0.100, \text{Group A (D-loop and levocardia) vs. Group C (D-loop and dextrocardia)}. \]
\[ p = 0.052, \text{Group B (L-loop and dextrocardia) vs. Group D (L-loop and levocardia)}. \]

The morbidity rate in patients treated by arterial DSO compared with those treated by Rastelli DSO [7].

Not all such data are contradictory, for there are many factors involved in patient selection which may influence the surgical options at different institutions and render the treatment of a large cohort of homogeneous patients impossible to accomplish using the same surgical repair procedure. The surgical options for CCT are often individualized [8–11], albeit there is a continual quest for the optimal surgical repair for CCT [14–17]. However, no studies have mentioned the influence of apical position, rotation (dextrocardia vs. levocardia) or atrial situs (solitus vs. inversus) on the LVOT obstruction in CCT.

Dextrocardia has been reported as a risk factor for CCT [18]; however, the influence of dextrocardia on LVOT obstruction in CCT remains unresolved. Our study found that the right ventricle has a proclivity to wrap around the LVOT in patients with apicocaval
Ipsilaterality (Groups A and B), in whom LVOT obstruction is prone to occur. It is justified to restore the posterior low-pressure ventricular outflow to become the systemic high-pressure ventricular outflow tract, and to free LVOT from anterior compression by DSO. In patients without apicocaval ipsilaterality (Groups C and D. Fig. 3B), although a DSO restored both atrioventricular and ventriculoarterial discordance, the persistent spatial arrangement of the ventricles, however, might render the posterior low-pressure right ventricular outflow to be compressed by the anterior high-pressure systemic ventricle potentially in the post-operative follow-up. As far as this disadvantage is concerned, the choice of a DSO in this anatomic setting should be reconsidered. Another risk factor of death in CCT was the length between the ventricular septal defect and the aortic valve, which may incur LVOT obstruction potentially. Under such circumstances, a univentricular repair would be less risky than a biventricular repair.

If one recalls the situation in normal heart with concordance of atrial situs and position of the apex, the low-pressure right ventricle was found to wrap around the systemic LVOT. The functional implication of this normal cardiac anatomy has never been revealed. Under the influence of apical pivoting, the spatial arrangement of the LVOT in patients with apicocaval ipsilaterality (Groups A and B), born with discordance of atrial situs and position of the apex, atrioventricular discordance, and ventriculoarterial discordance (so-called “triple” discordance), is in a proclivity to be wrapped around by the systemic right ventricle before surgery (upper panel in Fig. 1); thus, a DSO to restore both the atrioventricular and ventriculoarterial discordance is justified and reasonable. On the contrary, DSO may “initially” restore the atrioventricular and ventriculoarterial discordance (so-called double discordance) in patients without apicocaval ipsilaterality (Groups C and D), but may also “subsequently” cause post-operative right ventricular outflow tract obstruction or subpulmonary stenosis in the follow-up. Henceforth, in this setting of double discordance (without the 3rd discordance), the indication of a DSO is questionable. To them, DSO should less likely be regarded as an anatomical repair, since the positional anomalies, which surgeons cannot switch, persisted after DSO, and were not reversed like the CCT patients born with the 3rd discordance (Groups A and B).

In addition to LVOT obstruction, the anatomical position of the right atrium, under the influence of ventricular looping and apical rotation, is also of surgical importance in DSO [12,19,20]. Basically, the right atria in patients with apicocaval ipsilaterality (Groups A and B) are smaller than the right atria in patients without apicocaval ipsilaterality (Groups C and D).

Three vital points merit our attention. First, in patients with apicocaval ipsilaterality (upper panel in Fig. 1), including those with atrial/situs inversus and levocardia (Fig. 1A), and with atrial/situs solitus and dextrocardia (Fig. 1B), the posteriorly located right atrium is small. Thus, a Mustard (instead of a Senning or a modified Senning procedure with patch augmentation) would be indicated for better long-term outcome [19]. Second, a larger left atrium, which is anteriorly located, may facilitate surgical exposure of the ventricular septal defect through the tricuspid valve from the morphological right ventricle [12]. As shown in Fig. 1A and B, the axis of the conduction system is distributed farther away from the endocardium of the morphological right ventricle than from that of the morphological left ventricle, which may decrease the chance of complete atrioventricular block during exposure and repair of the ventricular septal defect from the morphological right ventricle [20]. Third, the volume overload of the smaller right atrium and right ventricle may become obvious with time during follow-up of patients with apicocaval ipsilaterality (Groups A and B) if treated by Rastelli DSO [11]. Thus, for those patients with apicocaval ipsilaterality (Groups A and B), one and a half ventricular repair may be an alternative [14–17].

Fig. 3. (A) A specimen of CCT in Group A in the setting of situs inversus and levocardia. The cardiac apex is to the left and ipsilateral to the near-by IVC, which drained into the I’t MRA. Note that the anterior wide-open RV was found to wrap around the posterior narrow LVOT, which directed toward an atritic and hypoplastic PT. (B) A specimen of CCT in Group D without ACI. Note that the patent posterior subaortic right ventricular outflow tract was wrapped around by the anterior subpulmonary LVOT in the setting of situs solitus and levocardia. A so-called anatomical repair, like double switch operation to restore the double discordance, might render the posterior low-pressure right ventricular outflow (committed to pulmonary artery) more susceptible to be compressed by the anterior systemic, or high-pressure, LV after surgery. ACI, apicocaval ipsilaterality; Ao, aorta; CCT, congenitally corrected transposition; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; PT, pulmonary trunk; RA, right atrium; RV, right ventricle. desc. Ao, descending aorta; IVC, inferior vena cava; l’t MRA, left-sided morphologically right atrium; r’t MRA, right-sided morphologically right atrium; SVC, superior vena cava.
Study limitations

First, the number of CCT with situs inversus was limited. Second, patients with ambiguous atrial situs and univentricular atrioventricular connections were excluded from this retrospective study. Third, only electron beam computed tomography was used to define the anatomical details of CCT in this study. However, cardiac magnetic resonance imaging is considered superior to computed tomography in assessing the functional and structural abnormalities of the right ventricle after DSO during long-term follow-up. It may be argued that an unusual apical position, or dextrocardia, exists more frequently in ambiguous atrial situs and univentricular atrioventricular connections, which were excluded from this retrospective study for the sake of definitive elucidation and discussion. Electron beam computed tomography was superior to other imaging modalities in defining the anatomical details of complex congenital cardiac defects. Our patients were categorized systemically into four groups and defined left-right and ventro-dorsal relationship of ventricular outflow tracts. The incidence of apicocaval ipsilaterality in CCT was 55.3% (57/103) in our study, thus permitting this observation.

Conclusion

LVOT obstruction is prone to occur in patients of CCT with apicocaval ipsilaterality. The fact that there exists a potential influence of apical position on the development of LVOT obstruction in patients with CCT should be taken into consideration when planning anatomical repair.

Conflict of interest

All authors declare that there is no conflict of interest.

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