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Original article

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

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ABSTRACT

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

Introduction

The apical position has been considered inconsequential to the segmental analysis of congenital cardiac defects [1]; 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 ipsilaterality, 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 ipsilaterality. 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 ipsilaterality.

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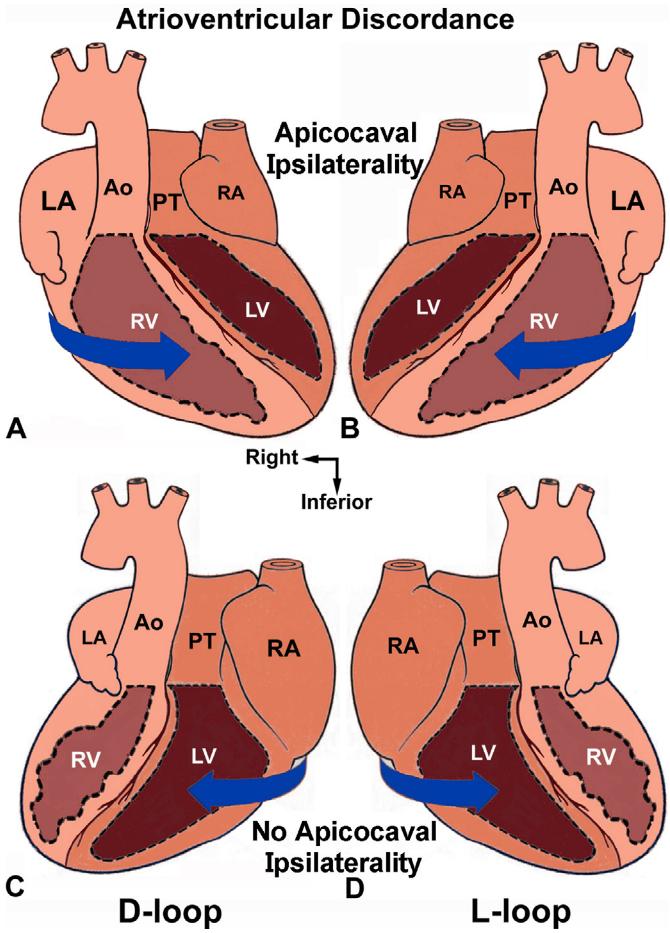

Fig. 1. As an evolutionary result of ventricular looping (D-loop vs. L-loop) and apical rotation (dextrocardia vs. levocardia), four possibilities of CCT exist. The upper panel shows that the ventricular loop and the apex are disposed to the opposite side of the spine, rendering an anatomical landmark of ipsilateral position of the apex and the inferior vena cava, i.e. ACI. In contrast, the lower panel depicts that the ventricular loop and the apex are disposed to the same side of the spine, resulting in absence of ACI. 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 indicates CCT with ACI, and the lower panel indicates CCT without ACI. The systemic right ventricle is in a proclivity to wrap around the LVOT in patients with ACI. LVOT is less likely to be compressed by the systemic right ventricle in patients without ACI. 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.

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 evolutional 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 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 $\pm$ 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 85.96% 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 "posterior" 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 "anterior" 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