行政院國家科學委員會專題研究計畫　成果報告

心尖部位在先天已矯正的大動脈轉位對左心室出口阻塞之影響

計畫類別：個別型計畫
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執行期間：92 年 08 月 01 日至 93 年 10 月 31 日
執行單位：國立臺灣大學醫學院外科

計畫主持人：邱英世
共同主持人：王主科，陳世杰，曾春典

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曾春典 國立台灣大學醫學院 內科

陳世杰 國立台灣大學醫學院 放射線科

一、中英文摘要

心尖部位一直被認為與依序分節診斷無關，其臨床意義只停留在心尖部位之觸摸，心電圖或超音波檢查需要在右位心或左位心之上進行檢查之階段而已。但實際上複雜性先天性心臟病常伴隨心尖部位之異常位置，其含義不為人所知。尤其在針對先天已矯正之大動脈轉位之雙轉換手術日益流行的今天，兩心室出口並沒有被轉換重建；徹底了解在心尖旋轉至與下腔靜脈同側時右心室有無包圍位於其後之左心室出口有其必要性。我們利用極快速電腦斷層掃描分析比較 65 例先天已矯正的大動脈轉位合併心尖部位異常之病例，配合臨床及血管造影之觀察，確定心尖部位旋轉對左心室出口阻塞之影響。我們發現心尖旋轉至與下腔靜脈同側時右心室會包圍位於其後之左心室出口 (n = 31)，其中 93.5% (n = 29)有左心室出口阻塞。但是在心尖沒有旋轉至與下腔靜脈同側時 (n = 34)右心室沒有包圍位於其後之左心室出口，其中 73.5% (n = 25)有左心室出口阻塞。這個差異在統計上有意義(p = 0.03)；在心房正位時 (p = 0.01)比心房反位(p = N.S.) 更加明顯。我們認為心尖旋轉至與下腔靜脈同側時高壓力的右心室會包圍位於其後之左心室出口，造成左心室出口阻塞；在心房正位時特別容易發生。施行雙轉換手術時兩心室出口並沒有被轉換重建，在這種病例有無必要施行雙轉換手術應重新考慮。

關鍵詞：心尖部位，房室連接反常，心室環，心室大動脈連接反常，心尖與下腔靜脈同側。。

Abstract

Apical position (Apex) is alleged to be independent of sequential segmental analysis, thus its clinical role was neglected. The right ventricle in congenitally corrected transposition with apicocaval ipsilaterality might wrap around the left ventricular outflow tract (LVOT). To delineate LVOT was compressed or not by the systematic ventricle in this setting, we carried out the following study. We analyzed 65 patients of congenital congenitally corrected transposition with various apical positions from 1981 to 2000. They were divided into 4 Groups according to the ventricular looping (Loop) and Apex: Group A, D-loop with levocardia; Group B, L-loop with dextrocardia, the direction of Loop and Apex are opposite to each other; Group C, D-loop with dextrocardia; Group D, L-loop with levocardia, the direction of Loop and Apex are to the same direction. Electron beam computed tomography was used to define left-right and ventro-dorsal relationship of left and right ventricular outflow tracts. Apicocaval ipsilaterality was found in 31 patients. (Group A, n = 15; Group B, n = 16), the right ventricle wrapped around LVOT in all cases. Among them, 93.5% (n = 29)

had LVOT obstruction (pulmonary atresia or stenosis). Whereas 73.5% of 34 patients without apicocaval ipsilaterality (Group C, n = 9; Group D, n = 25) had LVOT obstruction. The difference was statistically significant (p = 0.03).Further analysis found that this difference is more significant in the setting of situs solitus (Group B & D, p = 0.01) than situs inversus (Group A & C, p = N.S.). We concluded that LVOT obstruction is prone to occur in congenitally corrected transposition with situs solitus and (Group B) and apicocaval ipsilaterality, whereas in Group D, LVOT was not wrapped around by the systemic-pressured right ventricle had less incidence of LVOT obstruction. In the latter setting, to perform a double switch without exchanging ventricular outflow tract should be reconsidered.

Keywords: Apical position, Atrioventricular discordance, Ventricular looping, Ventriculo-arterial discordance, Situs solitus, Apicocaval ipsilaterality, Left Ventricular Outflow tract obstruction. .

二、Introduction

Apical position has been considered to be independent of sequential segmental analysis of congenital cardiac defect;[1] thus its clinical role was neglected, except during palpation of apical impulse and examining EKG[2] or echocardiography. For increasing complex congenital cardiac procedures performed at this tertiary referring center, we found it is useful to emphasize the clinical role of the cardiac apex in terms of not only defining the coronary artery (CA) in the posterior atrioventricular groove preoperatively,[3] but also helps in surgical planning to approach the intracardiac structures. Furthermore, the ventricular outflow tract was not switched in the double switch operation for congenitally corrected transposition,[4] in those with apicocaval ipsilaterality the systematic right ventricle might wrap around the left ventricle outflow tract.

To delineate the left ventricle outflow tract was compressed or not by the systematic ventricle in congenitally corrected transposition with apicocaval ipsilaterality. We carried out the following study.

三、Patients and Methods

From 1981 to 2000, we collected 65 patients with congenital cardiac defects with various apical position either pivoted dextrally (dextrocardia) or sinistrally (levocardia) in the setting of situs solitus or inversus, atrioventricular discordance and ventriculoarterial discordance. All patients with ambiguous atrial situs and/or univentricular atrioventricular connection were excluded from this study. The patients with single outlet or stenotic pulmonary outflow were included. The patients were divided into 4 Groups according to the type of ventricular looping relative to the apical position (Loop-Apex): in Group A (D-loop with levocardia) and Group B (L-loop with dextrocardia), Loop-Apex is opposite to each other (upper row in Fig. 1); in Group C (D-loop with dextrocardia) and Group D (L-loop with levocardia), Loop-Apex is to the same direction (lower row in Fig. 1). Fig.1 depicts the heart atrioventricular discordance and ventriculoarterial discordance. Apicocaval ipsilaterality represents dextrocardia in situs solitus or levocardia in situs inversus, in which the ventricular apical position and the inferior vena cava are on the same side to the midline spine, as shown on the upper row (A and B) on Fig. 1.

We used electron beam computed tomography imaging (Imatron C 150-L, South San Francisco, CA),[5] with electrocardiogram 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 was 1.5 mm (for neonates) or 3 mm. Computed tomography was performed beginning at the level of the great arteries, to the cardiac apex, without gaps. Nonionic iodinated contrast medium (2–3 mL/kg; Ultravist 370; Schering, Berlin) was delivered by a power injector. Patients under 5 years of age were routinely sedated with chloral hydrate 50 mg/kg before imaging.

Cardiac catheterization and angiocardiogram were used to evaluate the left ventricle tract obstruction (pulmonary stenosis or atresia) and the relationship between two ventricular outflow tracts. The incidence of obstruction in each group will be compared by Chi-square or Fischer exact test as appropriate.

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四、Results

Apicocaval ipsilaterality was found in 31 patients. (Group A, n = 15; Group B, n = 16), the right ventricle wrapped around LVOT in all cases (Fig. 2). Among them, 93.5% (n = 29) had LVOT obstruction (pulmonary atresia or stenosis) (Table 1). Whereas 73.5% of 34 patients without apicocaval ipsilaterality (Group C, n = 9; Group D, n = 25) had LVOT obstruction. The difference was statistically significant (p = 0.03).Further analysis found that this difference is more significant in the setting of situs solitus (Group B & D, p = 0.01) than situs inversus (Group A & C, p = N.S.) (Table 2).

Table 1. Number of cases in each group with and without apicocaval ipsilaterality

LVOT	Apicocaval ipsilaterality	No Apicocaval ipsilaterality
Obstrction	29	25
No obstruction	2	9
n	31	34

P = 0.03

Table 2. Number of cases in each group specified by atrial situs

LVOT	Apicocaval ipsilaterality			No Apicocaval ipsilaterality		
	Situs Inversus# Group A	Situs Solitus* Group B	Subtotal	Situs Inversus# Group C	Situs Solitus* GroupD	Subtotal
Obstruction	13	16	29	8	17	25
No obstruction	2	0	2	1	8	9
n	15	16	31	9	25	34

*P= 0.01 when comparing group B and D, situs solitus.
#P= N.S. when comparing group A and C, situs inversus.

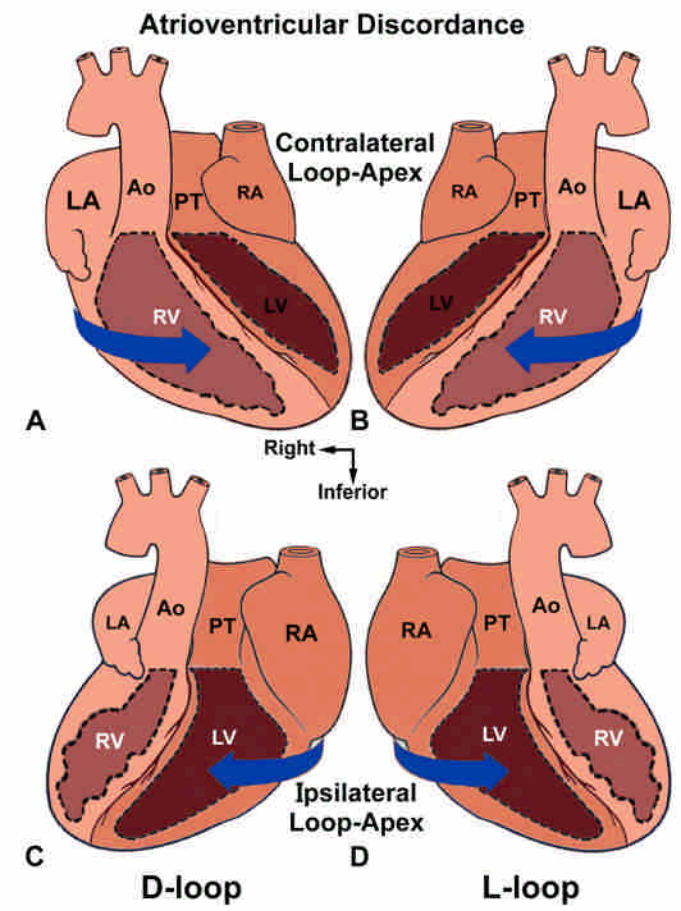

Fig. 1 Four possibilities of apical position in atrioventricular discordance as viewed frontally; the left column is situs inversus and D-loop, while the right column is situs solitus and L-loop. The upper panel has apicocaval ipsilaterality. (Ao=Aorta, LA=Left Atrium, Loop-Apex= the type of ventricular Loop relative to the ventricular Apical orientation, LV=Left Ventricle, PT=Pulmonary Trunk, RA=Right Atrium, RV=Right ventricle)

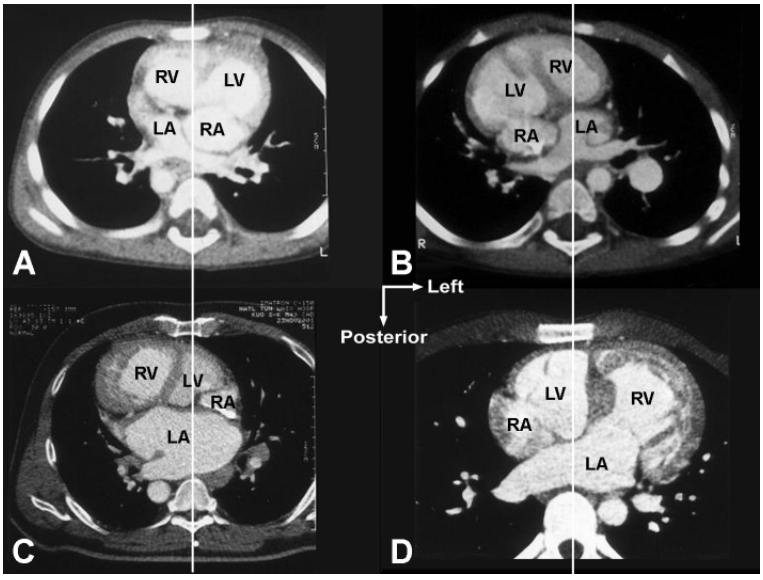

Fig. 2 Four possibilities of apical position in atrioventricular discordance as viewed axially on computerized tomogram; the left column is situs inversus and D-loop, while the right column is situs solitus and L-loop. The upper panel has the opposite apical orientation to the type of the ventricular loop that is present; in contrast, the lower panel both to the same direction. In the upper panel, the left ventricular outflow tract is wrapped around by the systemic-pressured right ventricle. The upper panel has apicocaval ipsilaterality. Whereas in the lower panel, the left ventricular outflow tract is free from compression. (Abbreviations see Fig. 1)

五、Discussion

Dextrocardia has been reported as a risk factor for mortality in congenitally corrected transposition.[6] Rastelli type double switch operation has been reported to prolong postoperative course than those with arterial switch type double switch that was performed on cases without uncorrectable left ventricle outflow obstruction.[7] But the association of dextrocardia and left ventricle outflow obstruction in congenitally corrected transposition has never been reported.

The choice of operation for cases of congenitally corrected transposition is often debated. The anatomical basis for selecting cases of congenitally corrected transposition that is justified for conventional repair or anatomical repair (double switch) is poorly understood. Our study found that LVOT was wrapped around by the systemic–pressured morphological right ventricle in congenitally corrected transposition with apicocaval ipsilaterality, thus the LVOT obstruction is prone to occur. Whereas in Group D, LVOT was not wrapped around by the systemic-pressured right ventricle had less incidence of LVOT obstruction. In those who are prone to be compressed (congenitally corrected transposition with apicocaval ipsilaterality), we should leave the right ventricle as subpulmonary low pressure outflow tract and carry out the anatomical repair (double switch); and vice versa in those who are not, one should choose a conventional repair thus the left ventricle outflow tract will remain as subpulmonary low pressure one to avoid compression on the right ventricle outflow tract.

In addition to the-above mentioned, apical rotation is of great surgical significance during double switch operation.[8-10] Firstly, smaller right atrium as a result of apical rotation in apicocaval ipsilaterality (Figs. 1A and 1B) might need Mustard instead of Senning or augmentation patch for a Senning.[8] Secondly, through the anterior and left larger atrium one may facilitate the exposures of the ventricular septal defect through the tricuspid valve from the morphological right ventricular aspect.[9] As shown on Fig. 1A and 1B, the conduction axis is further away from endocardial aspect of the morphological right ventricle than the morphological left ventricle, thus decrease the chance of surgical heart block.[10] Thirdly, when right heart overloading is obvious in cases with apicocaval ipsilaterality, especially for those need Rastelli type repair,[7] one and half ventricular repair is helpful.[11]

It may be argued that unusual apical position (dextrocardia) exists more frequently in ambiguous atrial situs and/or univentricular atrioventricular connections, all these malformations were ruled out in this study for more definitive elucidation and briefer discussion. The number of cases with situs inversus may be limited, but we have collected as many cases as possible. The electron beam computed tomography was super to define the anatomical details of complex congenital cardiac defects. We categorized our patients systemically into 4 groups and defined left-right and ventro-dorsal relationship of ventricular outflow tracts.

In conclusion, LVOT obstruction is prone to occur in congenitally corrected transposition with situs solitus and (Group B) and apicocaval ipsilaterality, whereas in Group D, LVOT was not wrapped around by the systemic-pressured right ventricle had less incidence of LVOT obstruction. In the latter setting, to perform a double switch without exchanging ventricular outflow tract should be reconsidered.

六、Self-Evaluation

The result of this one-year project revealed consistent with the original proposal. Investigators have demonstrated that contralateral Loop-Apex dictates LVOT was wrapped around by the systemic–pressured morphological right ventricle in congenitally corrected transposition. We found patients in congenitally corrected transposition with apicocaval ipsilaterality had their morphologically right ventricle wrapped around the left ventricular outflow tract. The incidence of left ventricular outflow tract obstruction (pulmonary atresia or stenosis) in each group was defined. For patients with unusual apical position, better surgical planning can be carried out. Hopefully one can identify the best candidate for double switch in complex congenital cardiac defects.

七、References

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