行政院國家科學委員會專題研究計畫　成果報告

心尖部位的臨床含義

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計畫主持人：邱英世

計畫參與人員：陳世杰

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共同主持人：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。我們利用極快速電腦斷層掃描分析比較各種複雜性先天性心臟病合併心尖部位異常，配合臨床及血管造影之觀察，發現心尖部位與冠狀動脈類型及外科手術由那個部位進行切開術有關連。在先天已矯正之大動脈轉位之雙轉換手術日益流行的今天，這些結果在臨床上有實際上的應用價值。

關鍵詞：心尖部位，心房位，房室連接反常，心室環，外科手術路徑，心室大動脈連接反常，後側房室溝冠狀動脈。

Abstract

To delineate the role of apical position (Apex) in diagnosis of coronary artery (CA) and surgical approach to the heart.

We analyzed 873 patients of various congenital cardiac defects from 1995 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; Group C, D-loop with dextrocardia; Group D, L-loop with levocardia. Electron beam computed tomography was used to define left-right and ventro-dorsal relationship of CA in the atrioventricular grooves and surgical access to the cardiac septa and valves. Either Group C (n=21) or D (n=33) had their morphologically right ventricle (mRV) and right CA pivoted posteriorly, irrespective of their atrial situs, atrioventricular or ventriculoarterial connection. Surgical access into the heart is easier via the anteriorly located morphologically left ventricle (mLV), left atrium in atrioventricular concordance or right atrium in atrioventricular discordance than the posterior one. Whereas in Group A (n=802) or B (n=17), mRV is pivoted anteriorly and the-above-mentioned are reversed.

Loop-Apex determines the ventricle (mLV if contralateral, mRV if ipsilateral) that is pivoted posteriorly, and hence its appropriate CA in the posterior atrioventricular groove. A surgeon should stand on the opposite side of the Apex to obtain the best-exposed operation field. Meanwhile, the anteriorly pivoted ventricle or its appropriate atrium is most accessible and can be considered as entry route into the heart when favorable.

Keywords: Apical position, Atrial situs, Atrioventricular discordance, Ventricular looping, Surgical approach, Ventriculo-arterial discordance, Posterior atrioventricular groove coronary artery.

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, but also helps in surgical planning to approach the intracardiac structures.

三、Patients and Methods

From 1995 to 2000, we collected 873 patients with various congenital cardiac defects with either apical position pivoted dextrally (dextrocardia) or sinistrally (levocardia) in the setting of situs solitus or inversus, atrioventricular concordance or 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. Those with mesocardia were also excluded for clear demonstration and brief discussion. 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 Figs 1 and 2); in Group C (D-loop with dextrocardia) and Group D (L-loop with levocardia), Loop-Apex is to the same direction (lower row in Figs. 1 and 2). Fig.1 depicts the heart with atrioventricular concordance, while Fig. 2 atrioventricular discordance. In Figs 1 and 2 only those with ventriculoarterial discordance are shown. The associated anomalies in patients of Groups A to D other than ventriculoarterial discordance (Table 1 and 2) are not depicted. 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 lower row (C and D) on Figs. 1 and 3, and upper row on Figs. 2 and 4.

We used electron beam computed tomography imaging (Imatron C 150-L, South San Francisco, CA),[3] with electrocardiogram gating to define the left-right and ventro-dorsal relationship of the coronary arteries in the atrioventricular grooves (Figs. 3 and 4). 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.

The coronary filling of aortogram or ventriculogram (Fig. 5) was used to define its course. Surgical accesses to the internal cardiac structures were compared in different groups from axial view of computed tomogram (Figs. 3 and 4) and frontal diagram (Figs. 1 and 2).

四、Results

The number of patients in each group was listed in Fig. 1 (atrioventricular concordance), and Fig. 2 (atrioventricular discordance) (405 female and 468 male; aged 2 days to 24 year-10 month; mean 5 year-2 month).

Coronary Artery (CA)

All patients in groups C and D had their morphologically right ventricle (mRV) pivoted posteriorly and hence its right CA is posterior to the anteriorly located left circumflex CA,

irrespective of a situs solitus (Figs. 3C and 4D), a situs inversus (Figs. 3D and 4C), atrioventricular concordance (Figs. 3C and 3D), atrioventricular discordance (Figs. 4C and 4D), ventriculoarterial concordance or ventriculoarterial discordance (Figs. 1 and 2). Fig. 5 illustrated the angiogrphic findings of a concordant transposition (Fig. 1C) and discordant transposition (Fig. 2D) in the setting of situs solitus. That a D-loop ventricle with dextrocardia (upper panel, Group C) or a L-loop ventricle with levocardia (lower panel, Group D,) had their morphologically right CA located posterior to the left circumflex CA.

In contrast, for Groups A and B the posterior ventricle is the morphologically left ventricle (mLV) and the left circumflex CA is posterior to the morphologically right CA.

Surgical Approach

In all Groups, a surgeon stands on the side of patient opposite to the apical position to look into the intracardiac structures can get the best-exposed operation field.

For the heart in Groups C and D, mLV and its appropriate atrium are pivoted anteriorly. The posterior mRV and its appropriate atrium are often smaller, as shown in Fig.3C. To repair multiple muscular ventricular septal defects or parachute mitral valve through the anterior mLV is easier. In atrioventricular concordance with apicocaval ipsilaterality (Fig. 1C, 1D, 3C and 3D), left atrial approach to either the atrial septum or mitral valve is easier. However, in atrioventricular discordance without apicocaval ipsilaterality (Figs. 2C, 2D, 4C and 4D), right atrial approach to these two structures in easier. But isolated ventricular septal defect is approached without incising the systemic ventricle; when the posterior atrium is too small, transarterial approach to repair the subpulmonic or subaortic ventricular septal defect is performed.

In contrast, for Groups A and B, the mRV and its appropriate atrium are pivoted anteriorly (Figs. 3A, 3B, 4A and 4B); surgical access to intracardiac structure via these chambers is easier. In atrioventricular discordance with apicocaval ipsilaterality (Figs. 2A and 2B), surgical access to repair the ventricular septal defect through the left atrium is as easy as it is through the right atrium in atrioventricular concordance (Figs. 1A and 1B).

五、Discussion

Surgical access to the heart from the left side in a dextrocardia case with situs solitus has been reported, either to replace the mitral valve[4] or to reroof the coronary sinus.[5] In one case associated with juxtaposition of atrial appendages, reattachment of the left superior vena cava to the left-sided right atrial appendage from the left side was also facilitated by dextrocardia.[6]

Embryogenetically ventricular loop formation occurs in two steps. First, the heart tube loops in one direction (normally to the right, forming a D-loop; or abnormally to the left, forming an L-loop). Second, the ventricular apex “should” swing in the opposite direction (normally to the left, resulting in levocarida; or abnormally to the right, resulting in dextrocardia). This is to say: (1) All D-loop ventricles “should” have levocardia (Group A); and (2) All L-loop ventricles “should” have dextrocardia (Group B). If only the first step is completed (looping), but not the contralateral apical swing, then the mRV is posterior to the mLV and the right CA is posterior to the left circumflex CA. Hence, the situation (posterior mRV and right CA) occurs in two situations: (1) D-loop ventricle with dextrocardia (Group C); and (2) L-loop ventricle with levocardia (Group D). Figure 5 illustrated the angiographic findings of ipsilateral Loop-Apex, that dextroversion of a D-looped ventricle (upper panel) or levoversion of a L-looped ventricle (lower panel) will have their morphologically right CA located posterior to the left circumflex CA.

This general rule is true irrespective of situs solitus or inversus (Figs. 1 to 4, left vs. right column), atrioventricular concordance or atrioventricular discordance (Figs. 1 and 3 vs. Figs.

2 and 4). The coronary angiographic finding has been demonstrated in dextrocardia,[7] but its implication with apical position has never been pointed out before.[2] The above rule helps not only preoperative angiographic or echographic CA interpretation, but also intraoperative CA identification and transfer during an arterial switch operation. The accurate diagnosis of the CA anatomy is the cornerstone of a successful CA transfer. Fig. 5 illustrated the frequently misinterpreted angiogram before an arterial switch operation. In contrast to Fig. 5 (ipsilateral Loop-Apex), only those transpositions with contralateral Loop-Apex will have their morphologically left circumflex CA located posterior to the right CA as usually impressed.

Besides helping identification of CA pre- and intra-operatively in the present era of increasing double switch operation for congenitally corrected transposition,[8-11] our study is also useful to clarify the clinical implications of apicocaval ipsilaterality. Apicocaval ipsilaterality is known to occur more frequently in congenitally corrected transposition. Another way of saying the above rule incorporating apicocaval relationship is: Complete transposition without apicocaval ipsilaterality (Fig. 1A, 1B, 3A and 3B) and congenitally corrected transposition with apicocaval ipsilaterality (Fig. 2A, 2B, 4A and 4B) will have their morphologically left circumflex CA located posterior to the right CA as usually impressed, and vice versa. The degree of posterior shifting of the left circumflex CA toward the right CA can indicate the index of apicocaval ipsilaterality.

Apicocaval ipsilaterality is of great surgical significance[8,10,11] during double switch operation. Namely, smaller right atrium as a result of apical rotation in apicocaval ipsilaterality (Figs. 2A, 2B, 4A and 4B) might need Mustard instead of Senning or augmentation patch for a Senning.[8] On the other hand, through the anterior and left larger atrium one may facilitate the exposures of the ventricular septal defect through the tricuspid valve from the mRV aspect.[10] As shown on Fig. 2A and 2B, the conduction axis is further away from endocardial aspect of mRV than mLV, thus decrease the chance of surgical heart block.[11] Furthermore, when right heart overloading is obvious in cases with apicocaval ipsilaterality, especially for those need Rastelli type repair, one and half ventricular repair is helpful.[12]

It may be argued that unusual apical position (dextrocardia) exists more frequently in ambiguous atrial situs and/or univentricular atrioventricular connections, but all these malformations were ruled out in this study. When repairing these congenital defects, univentricular repair is often the choice instead of biventricular repair. For univentricular repair it is not necessary to identify the cardiac chambers and CA or to expose intracardiac structure, especially for extracardiac total cavopulmonary connection, except concomitant atrioventricular valve repair is necessary. Some cases with ambiguous atrioventricular connections are fit for biventricular repair, such as those who had less morbid association in left isomerism than right isomerism.[13] They still can be analyzed according to the Loop-Apex as in this communication. In those with mesocardia usually two atrioventricular groove CA's overlap at true lateral view, and can be analyzed in either direction yielding to which outweigh the other, although it was excluded in this study for briefer discussion and clear demonstration.

Although anterior chamber is more easily accessible, sometimes one has to incise a posterior chamber. For those who need Rastelli operation, an incision on the right ventricular infundibulum is inevitable, despite its posterior location. The other unfavorable example to incise the anterioly more accessible chamber is isolated ventricular septal defect in Group C and D; one would be reluctant to incise the anterior systemic mLV. In such case when the posterior atrium is too small, transarterial approach to repair the subpulmonic or subaortic ventricular septal defect is an alternative.

六、Self-Evaluation

The result of this one-year project revealed consistent with the original proposal. Investigators have demonstrated that contralateral Loop-Apex dictates the mLV and left circumflex CA to be located posteriorly; while ipsilateral Loop-Apex has posterior mRV and right CA, irrespective of their atrial situs, atrioventricular connection or ventriculoarterial connection. A surgeon stands on the side of patient that is opposite to the apical position can get the best-exposed operation field. Meanwhile, the anteriorly pivoted ventricle or its appropriate atrium is most accessible and can be considered as entry route into the heart when favorable. We would recommend left atrial approach to repair the ventricular septal defect through the tricuspid valve during anatomical repair of Group A and B patients with atrioventricular discordance (Figs. 2A and 2B) as though they were approached through the right atrium in atrioventricular concordance (Figs. 1A and 1B).

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Table 1—Associated anomalies of the patients other than ventriculoarterial discordance in the setting of atrioventricular concordance

Anomalies	No. of cases			
	1A	1B	1C	1D
Right lung agenesis or hypoplasia	0	0	3	0
Left pulmonary artery sling	0	0	1	0
Scimitar syndrome	0	0	2	0
Atrial septal defect	0	1	0	0
Ventricular septal defect	0	0	2	1
Tetralogy of Fallot	0	2	0	0
Anatomically corrected malposition	4	0	1	0
Isolated ventricular inversion	4	0	0	0
Isolated infundibuloarterial inversion	1	0	0	0

Table 2—Associated anomalies of the patients other than ventriculoarterial discordance in the setting of atrioventricular discordance

Anomalies	No. of cases			
	2A	2B	2C	2D
Right lung agenesis or hypoplasia	0	3	0	0
Left pulmonary artery sling	0	1	0	0
Scimitar syndrome	0	2	0	0
Atrial septal defect	0	0	1	0
Ventricular septal defect	1	2	0	0
Tetralogy of Fallot	0	0	2	0
Anatomically corrected malposition	0	1	0	4
Isolated ventricular inversion	0	0	0	4
Isolated infundibuloarterial inversion	0	0	0	1

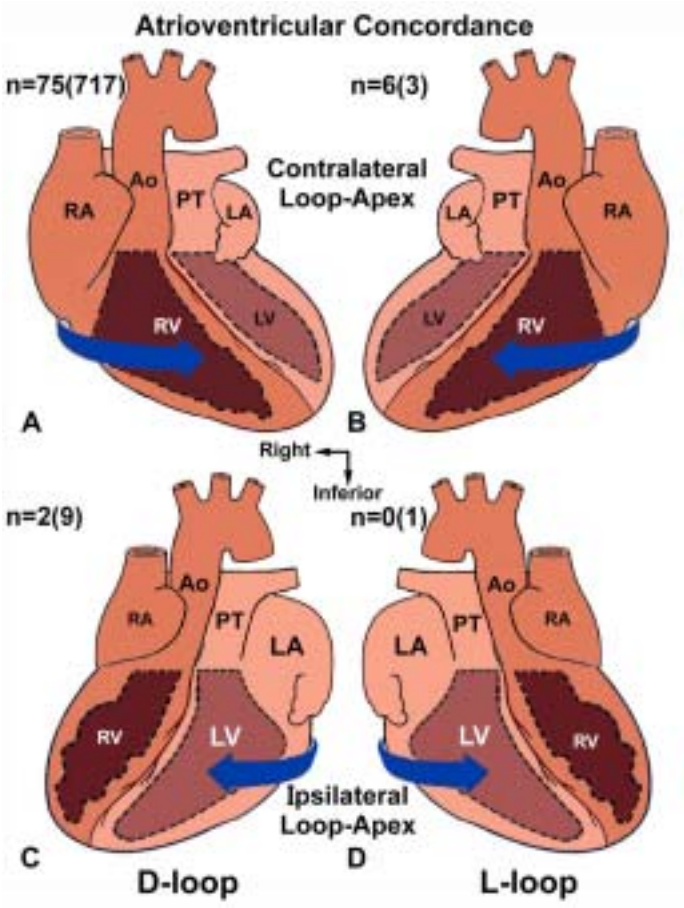

Fig. 1 Four possibilities of apical position in atrioventricular concordance as viewed frontally; the left column is situs solitus and D-loop, while the right column is situs inversus and L-loop. Numbers in parentheses indicate number of cases other than ventriculoarterial discordance. That the conduction axis is closer to the endocardial aspect of the morphologically left ventricle is depicted. The lower row 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)

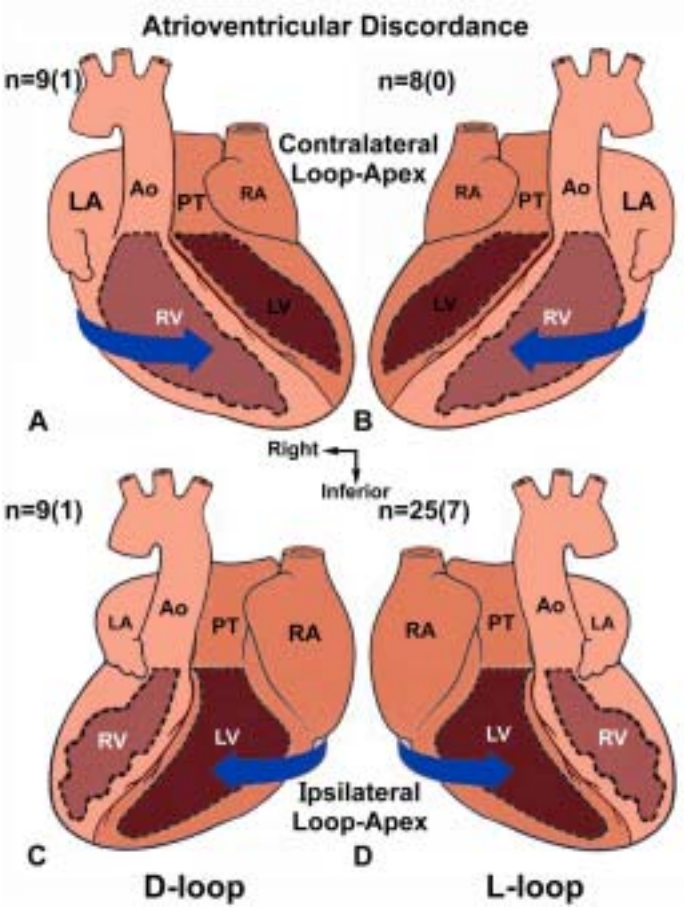

Fig. 2 Four possibilities of apical position in atrioventricular discordance as viewed frontally; the left column is situs inversus and D-loop, while the right column is situs solitus and L-loop. Numbers in parentheses indicate number of cases other than ventriculoarterial discordance. That the conduction axis is closer to the endocardial aspect of the morphologically left ventricle is depicted. The upper row has apicocaval ipsilaterality. (Abbreviations see Fig. 1)

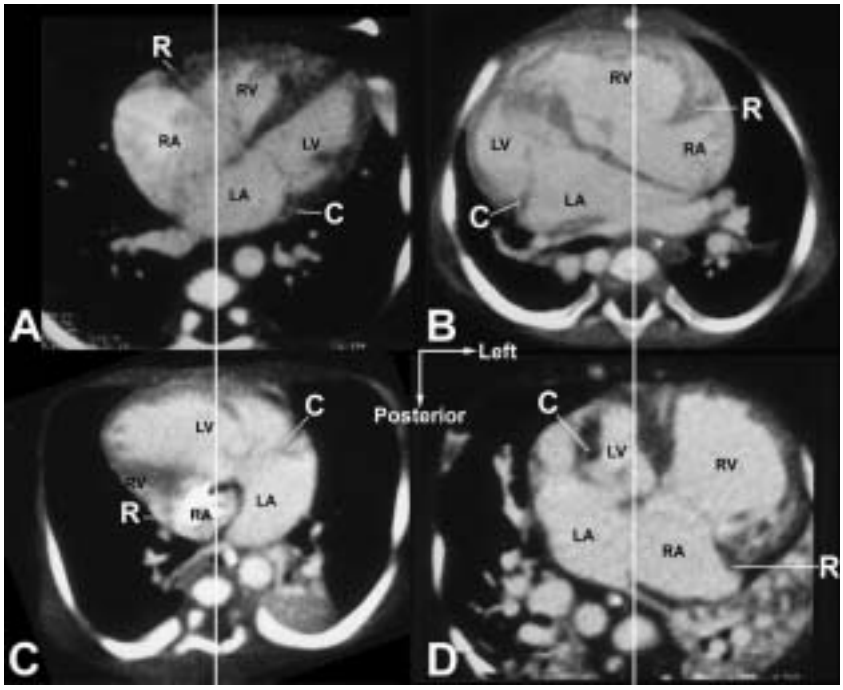

Fig. 3 Four possibilities of apical position in atrioventricular concordance as viewed axially; the left column is situs solitus and D-loop, while the right column is situs inversus and L-loop. The upper panel has the opposite apical orientation to the type of the ventricular loop that is present; in contrast, the lower panel both to the same direction. In the lower panel, the left circumflex artery (C) is in front of the right coronary artery (R), while in A and B, R is in front of C. The lower panel has apicocaval ipsilaterality. (Abbreviations see Fig. 1)

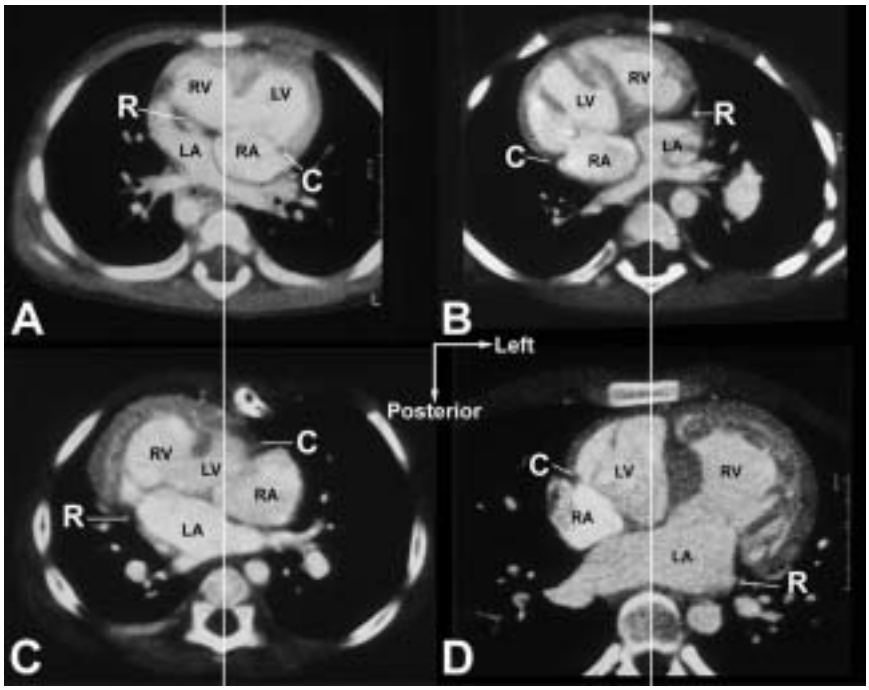

Fig. 4 Four possibilities of apical position in atrioventricular discordance as viewed axially; the left column is situs inversus and D-loop, while the right column is situs solitus and L-loop. The upper panel has the opposite apical orientation to the type of the ventricular loop that is present; in contrast, the lower panel both to the same direction. In the lower panel, the left circumflex artery (C) is in front of the right coronary artery (R), while in A and B, R is in front of C. The upper panel has apicocaval ipsilaterality. (Abbreviations see Fig. 1)

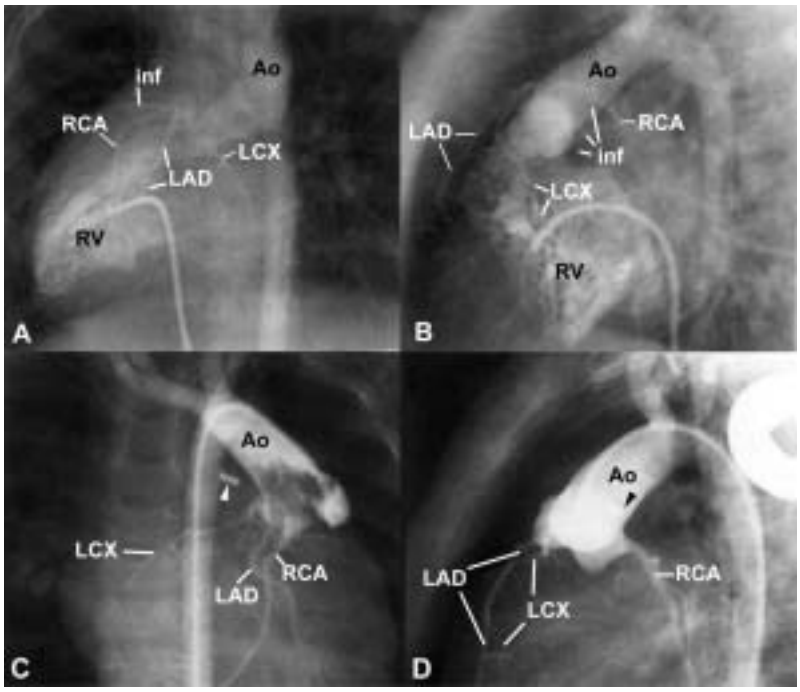

Fig. 5 Coronary angiogram in one patient of complete transposition with situs solitus and dextrocardia (A and B, Fig. 1C), and another of congenitally corrected transposition with situs solitus and levocardia (C and D, Fig. 2D). Clip (arrowhead) indicated site of pulmonary trunk banding. A and C are frontal projections, while B and D, left lateral view. Both patients had their morphologically right coronary artery (RCA) located posterior to the left circumflex artery (LCX) on lateral view. In the upper panel the coronary artery with the infundibular branch (inf) is the RCA. In the lower panel, RCA is the largest in diameter, while the LCX is the smallest. (Ao = aorta, RV = right ventricle, LAD = left anterior descending)