

# 行政院國家科學委員會專題研究計畫成果報告

計畫名稱：主動脈在左側之先天性心臟畸型的冠狀動脈類型之分類

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† 八十六年度及以前的一般國科會專題計畫(不含產學合作研究計畫)亦可選擇適用，惟較特殊的計畫如國科會規劃案等，請先洽得國科會各學術處同意。

## 中英文摘要

## Abstract

**背景：**在目前針對先天已矯正型大動脈轉位(CCT)施行心房和大動脈雙轉換手術的時代，我們有系統的分析其冠狀動脈類型，並探究其與主動脈肺動脈幹旋轉之相關性。

**方法：**我們在本院收集了65個CCT，分成心房正位(CCTS)和心房反位(CCTI)兩組。並與大動脈完全轉位合併心房正位(CTS)和心房反位(CTI)之冠狀動脈分類做比較。冠狀動脈解剖學依照房位、心尖部位、心室環和主動脈和肺動脈幹旋轉來分析。根據心表面靠近心臟基部之相類似之中央冠狀動脈主要類型及其變異型分成四個類型(O, I, II和IV)。

**結果：**在CCTS右冠狀動脈至左房室溝(同CTI)，而在CCTI至右房室溝(同CTS)；但在這兩組CCT無心尖下腔靜脈毗連者右冠狀動脈皆到較後側之房室溝。但是如果合併心尖和下腔靜脈毗連，左旋冠狀動脈可後移至右冠狀動脈後側。否則在相同之主動脈肺動脈幹旋轉，四組心臟皆有相類似之中央冠狀動脈類型；而且依照反時鐘反向旋轉由類型IV(n=1)，類型II(n=9)，類型I(n=18)和類型O(n=37)依序演變(p<0.00000)。

**結論：**在房室溝之周邊冠狀動脈類型依照心尖和下腔靜脈毗連之程度決定其前後類型，而依據心室環決定其左右類型；不論其房位如何。靠近主動脈竇之中央冠狀動脈，因兩者依方便而結合，故中央冠狀動脈類型依據主動脈肺動脈幹旋轉而定；因此可由兩條大動脈關係位置預知冠狀動脈之可能類型，不論其心臟畸型為何。

**關鍵詞：**先天已矯正型大動脈轉位，冠狀動脈類型，主動脈肺動脈幹旋轉，房位，心尖部位，心室環，心尖下腔靜脈毗連。

**Background** In the current era of combined atrial and arterial switch operations for congenitally corrected transposition (CCT), we analyze its coronary artery (CA) anatomy systematically and correlate them with aortopulmonary rotation (APR).

**Methods.** We collected 65 cases of CCT with situs solitus (CCTS) or inversus (CCTI) at our hospital and compared with the previous classifications of CA in complete transposition with situs solitus (CTS) or inversus (CTI). CA anatomy was analyzed relating to atrial situs, apical position, ventricular looping and APR. Four main types and their similar variants of epicardial configuration at the base of the heart were categorized into four patterns (Pattern O, I, II and IV).

**Findings** The right CA (RCA) coursed to the left in CCTS (as CTI), and to the right in CCTI (as CTS); but to the more posterior atrioventricular groove in both without apicocaval juxtaposition (concordance of atrial situs and apical position). However, in CCT with more juxtaposition, the left circumflex may shift posterior to the RCA. Otherwise with the same APR, all above four groups have similar central pattern; and the evolution from pattern IV (n=1), II (n=9), I (n=18) to O (n=37) is dependent on counterclockwise APR.

**Interpretation** Peripheral CA pattern in the atrioventricular groove is dictated by apicocaval juxtaposition anterioposteriorly and ventricular looping dextrosinistrally, irrespective of atrial situs. Central CA pattern near the aortic sinus depends on APR due to marriage of convenience between them, and thus predictable from arterial relations.

**Keywords:** Coronary artery pattern,

aortopulmonary rotation, congenitally corrected transposition, atrial situs, ventricular looping and apicocaval juxtaposition.

## **Introduction**

Precise CA anatomy in congenitally corrected transposition (CCT), which is less well understood as that in complete transposition (CT)<sup>1,2</sup>, is surgically demanding in the current era of increasing combined atrial and arterial switch operations for this setting<sup>3,4</sup>. We proposed a systemic way to analyze the coronary artery (CA) anatomy in CCT, in an effort to improve the present random and incomplete classifications<sup>5-8</sup>. The impact of atrial situs, apical position, ventricular looping and aortic root position were analyzed sequentially. We also studied the correlation between the CA pattern and aortopulmonary rotation in CCT.

## **Methods**

### *Study patients*

Data from all patients who presented between 1981 and 1999 with congenitally corrected transposition of the great arteries with a documented coronary artery type and relations of the great arteries were collected for analysis. Coronary artery type and aortopulmonary relation were identified at angiography, ultrafast computed tomography, two-dimensional echocardiography, surgical intervention or autopsy. The patients were divided into two groups according to atrial arrangement with situs solitus (CCTS) or inversus (CCTI). The position of cardiac apex and direction of ventricular looping were also recorded in each case, and correlated with the peripheral coronary arterial pattern in the atrioventricular (AV) groove. Only patients with AV discordance and ventriculoarterial discordance (with or without pulmonary stenosis), single outlet of the heart (pulmonary atresia), or double-outlet right ventricle were included. All univentricular hearts and those with isomeric atrial situs were excluded. In each case, the ventricular septum was either intact or had a defect.

### *Coronary artery types (central pattern)*

The previously reported coronary artery types<sup>5-12</sup> were reorganized into five patterns (X, O, I, II, and IV) according to similarities of epicardial configuration at the base of the heart (figure 1). Our previous classification of coronary artery types in complete transposition<sup>2</sup> (CT), with either situs solitus (CTS) or inversus (CTI), was put together for convenience of comparison. In the present study, type 0 was BA-1 of the de Groot classification,<sup>13,14</sup> and types 1, 2, and 4 were those reported by Shaher and Puddu<sup>15</sup> (also designated by Yacoub and Smith<sup>16</sup> as A, D, and E). Type 10, previously reported in otherwise normal heart<sup>17</sup> and tetralogy of Fallot<sup>18-20</sup>, was also documented in congenitally corrected transposition<sup>7</sup>. The coronary artery that coursed to the lateral AV groove in CCTI and CTS is reversed from that in CCTS and CTI. Type 3c was grouped together with type 0 to pattern O, type 3d to pattern I, type 10a to pattern X, and type 3a was transitional between pattern I and II and grouped to pattern II.

Previous reports with documented coronary artery type and arterial relations were reviewed. Eta-square (correlation ratio) analysis was used to test statistical significance.

### *Terminology*

The following terms are used throughout: right- or left-hand sinus is the sinus on either the right- or left-hand side if one is positioned according to the nonfacing cusp of the aortic valve<sup>13</sup>. Facing commissure is the commissure that faces the pulmonary trunk. Right- or left-hand nonfacing commissure indicates the other two commissures on the right- or left-hand side if one is positioned according to the nonfacing cusp of the aortic valve. Apicocaval juxtaposition means the discordance of atrial situs and apical position.

## **Results**

Sixty-five patients who met the selection criteria were identified (table 1) (mean age at

time of admission 5 years and 2 months, range 4 days to 46 years; 42 male, 23 female).

### *Peripheral pattern*

#### *Impact of atrial situs and apical position (figure 2)*

In CCT without apicocaval juxtaposition (concordance of atrial situs and apical position), irrespective of its situs, the coronary artery in the more posterior AV groove is always the RCA (CCTS, n=25, figure 2A; CCTI, n=9, figure 2D). In contrast, the CA in the more posterior AV groove is morphologic left circumflex artery (LCX) in CT. The LCX shifts posteriorly in cases with apicocaval juxtaposition (figures 2 B, 2C, 2E and 2F). The degree of posterior shifting of the LCX toward RCA can indicate the index of apicocaval juxtaposition. In those with less rotation of the cardiac apex (figures 2B and 2E), LCX is still located anterior to the RCA or at the same level (CCTS, n=8; CCTI, n=6). However, in extreme cases (figures 2C and 2F), LCX is shifted even posterior to the slightly anteriorly shifted RCA (CCTS, n=8; CCTI, n=9), resulting in a similar anteroposterior AV groove arterial pattern as in CT.

#### *Impact of ventricular looping*

The left-sided AV groove artery in CCTS is the RCA (n=41) (figure 1, right column); while in CCTI with dextrolooping of the ventricle, RCA is right-sided (n=24) (figure 1, second column from the left).

A similar peripheral coronary artery pattern can be recalled in CCT as that in CT with the same apical position and ventricular looping in all hearts studied irrespective of their atrial situs. For example, the usual peripheral pattern of CTS (figure 3A) can be seen in CCTI with extreme apicocaval juxtaposition (figure 3B, cardiac apex well to the left). Their lateral views (figures 3D and 3E) are also similar. In CCTI with less or without apicocaval juxtaposition (figures 3C and 3F) as seen in most cases (15/24=63%), the anteroposterior view (figure 3C) is similar, whereas in the lateral view the posterior AV groove artery is the RCA (figure 3F), instead

of the LCX.

The usual peripheral coronary pattern in CCTS with extreme apicocaval juxtaposition (levolooping and apex to the right, figures 4B and 4E) is the same as CTI (levolooping and also apex to the right, figures 4A and 4D). With less or no apicocaval juxtaposition (figures 4C and 4F, apex to the left; 33/41=80%), the posterior AV groove artery is the RCA (figure 4F), while in anteroposterior view (figure 4C) its pattern is still the same as figures 4A and 4B.

### *Central pattern*

#### *Impact of aortic root position and its rotation*

Coronary arterial pattern near the aortic sinus depends on aortic root position irrespective of the disease category. From the aortic root position one can recall a similar coronary pattern near the aortic sinus in CCT as that in CT with the same ventricular looping. With similar aortopulmonary relation, the usual pattern of central coronary artery in CTS (figures 3A and 3D) can be anticipated in CCTI with (figures 3B and 3E) or without (figures 3C and 3F) apicocaval juxtaposition. For CCTS, one may also recall a similar central coronary pattern as CTI based on the relation of the great arteries. In CTI the most frequent pattern is type 0 (figures 4A and 4D); with the same aortopulmonary rotation one can recall in CCTS a similar central pattern with (figures 4B and 4E) or without (figures 4C and 4F) apicocaval juxtaposition. In one case of CCTS, a small infundibular branch entering the right hand sinus is noted in addition to the type 3c pattern.

The number of cases in each group is listed in table 1. Types 2<sub>ccts</sub>, 3d<sub>ccts</sub> and pattern X are not observed in this study. Type 2 of pattern II coronary artery, Yacoub type D or Shaher type 2 in CTS, can also be seen in CCTI with a more right anterior aorta (figures 5A and 5C). Type 4 of pattern IV has been reported<sup>5</sup> and seen in this series. In contrast, type 3c seen in CTS with a left anterior aorta can be seen in CCTS with a left anterior aorta (figures 5B and 5D). The aortic root may be left anterior to the pulmonary trunk in CTS<sup>2</sup>

<sup>14</sup> or right anterior to the pulmonary trunk in CCTS.<sup>5,7,8</sup> For a right anterior aorta in CCTS, types 2 and 3a coronary artery are reported,<sup>5-8</sup> type 1 and 3a are seen in this series (table 1); inasmuch types 3c and 0 coronary artery are documented in CTS with a left anterior aorta.<sup>2,14</sup> Thus CA pattern near the aortic sinus depends on aortic position irrespective of CT, CCT or their atrial situs. There is no pattern X in this series and the aortopulmonary rotation of pattern X is not specified<sup>7</sup>. Otherwise the evolution in the rest patterns from pattern IV to O is dependent on clockwise aortopulmonary rotation in all groups (eta-square 0.840,  $p < 0.00000$ ).

### Discussion

Coronary artery-ventricular concordance described in CCT frequently misleads that the left circumflex artery courses through the right AV groove in CCTS to reach the posterior surface of the heart,<sup>6,12</sup> especially in view of subaortic right ventricle is usually leftward and anteriorly located in CCT according to the loop rule. From this study it is clear that usually the RCA coursed to the posterior AV groove in CCT with less or without apicocaval juxtaposition. Because its ventricle connects with the left atrium that is more posteriorly located than the right atrium. The originally anteriorly located LCX in CCT shifts posteriorly in cases with apicocaval juxtaposition. The degree of posterior shifting of the LCX toward RCA can indicate the index of apicocaval juxtaposition that is of great surgical significance during double switch operation.<sup>4,21</sup> Namely, smaller right atrium as a result of apical rotation in apicocaval juxtaposition might need Mustard instead of Senning or augmentation patch for a Senning. On the other hand, through the larger left atrium one may facilitate the exposure of right ventricular aspect of the ventricular septal defect, thus decrease the chance of surgical heart block. Furthermore, when right heart overloading is obvious in cases with apicocaval juxtaposition, especially for those need Rastelli type repair, one and half ventricular repair is helpful.

The most frequent coronary artery pattern seen in CCTS is type 0,<sup>5-12</sup> the coronary artery courses to the lateral AV groove is reversed to BA-1 of the de Groot classification,<sup>13</sup> or mirror image of usual pattern in CTS.<sup>7</sup> In CCTS with type 0 coronary artery, the RCA is concordant with the right-hand sinus, and left coronary artery (LAD + LCX) with the left-hand sinus. In usual pattern of CTS (type 1), RCA is discordant with the right-hand sinus and also left coronary artery is discordant with the left-hand sinus. In this sense, discordance between coronary artery and aortic sinus can be used to define the transposition and concordance between them means corrected transposition. However, this definition only works in CCTS and CTS. In situs inversus of both situations, i.e. CCTI and CTI, this definition fails. The usual pattern in CCTI is the same as the usual pattern in CTS. Therefore, we would agree with McKay et al.<sup>8,22</sup> that it is not necessary to assume a stance in the "nonfacing" pulmonary sinus as some suggested<sup>7</sup>, especially in CCT to avoid confusion. Because it is the loop rule that determines the ventricle and hence its appropriate coronary artery in the lateral AV groove. In levolooped hearts such as CCTS and CTI, the RCA courses to the left AV groove; and vice versa.

Finally aortic root position dictates the types of coronary artery in CCT as in CT because marriage of convenience between aortic sinus and coronary arteries. It is demonstrated by embryologic study and previous analysis that coronary artery, pierced the nearest site of the aortic sinus after aortopulmonary rotation.<sup>2,23,24</sup> Similar coronary artery type, both central and peripheral, can be anticipated in face of the same apical position, ventricular looping and aortopulmonary rotation irrespective of their atrial situs. We demonstrated the correlation between coronary artery type and aortopulmonary rotation in complete transposition<sup>2</sup> and tetralogy Fallot<sup>20</sup>. We now establish CCT occupied a more left anterior circle of six basic patterns (figure 6). Pattern

X, and its subtype with single coronary artery were reported in congenitally corrected transposition with situs solitus<sup>7</sup>. The aorta was left to the pulmonary trunk in Pattern X, but their aortopulmonary rotation was not specified. The dependence of coronary artery types on aortopulmonary rotation makes it possible to anticipate coronary artery types on the basis of relation of the great arteries. In this communication we analyzed the coronary artery anatomy in a segmental sequence, that a systematic way to diagnose the coronary artery anatomy in CCT is realized.

### **Conclusions.**

**1. Peripheral pattern** One can recall a similar peripheral coronary pattern in CCT as in CT with the same apical position and ventricular looping.

#### **a. Impact of atrial situs and apical position**

In CCT without apicocaval juxtaposition (concordance of atrial situs and apical position), irrespective of its situs, the CA in the more posterior atrioventricular groove is always the RCA instead of the LCX as in CT, due to its ventricle connects with the left atrium that is more posteriorly located than the right atrium. The LCX shifts posteriorly in cases with apicocaval juxtaposition. The degree of posterior shifting of the LCX toward RCA can indicate the index of apicocaval juxtaposition that is of great surgical importance. In extreme cases, LCX is shifted even posterior to the slightly anteriorly shifted RCA, resulting in a similar anteroposterior atrioventricular groove arterial pattern as in CT.

#### **b. Impact of ventricular looping**

The left-sided atrioventricular groove artery in CCTS and CTI is the RCA, and vice versa; as the loop rule determines the ventricle and hence its appropriate coronary artery.

**2. Central pattern** Central pattern is dictated by the aortic root position irrespective of CT or CCT.

#### **Impact of aortic root position and its**

### **rotation**

Coronary pattern near the aortic sinus depends on aortic root position because of marriage of convenience between coronary artery and aorta. One can recall a similar central coronary pattern in CCT as that in CT with the same ventricular looping on the basis of the relation of the great arteries.

Dependence of coronary artery pattern on aortopulmonary rotation made it possible to predict the coronary pattern in CCT on the basis of the relation of the great arteries.

**As a whole**, apical position dictates the peripheral AV groove artery anteroposteriorly, ventricular looping dextrosiristrally; while aortic root position dictates the central coronary artery pattern irrespective of their atrial situs or disease category. One can recall a similar coronary pattern, both central and peripheral, in CCT as in CT with the ventricular looping, apical position and APR (CCTS with dextrocardia as in CTI, CCTI with levocardia as in CTS; both with extreme apicocaval juxtaposition). However, in CCT with lesser or without juxtaposition (CCTS with meso- or levocardia, CCTI with meso- or dextrocardia) as seen in most cases, the central pattern is still similar to CT with the same APR; but the more posterior AV groove artery is the RCA instead of the LCX as it is in CT and the lateral AV groove artery is dictated by ventricular looping.

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