Easy Category for Complex Congenital Cardiac Segmental Connections

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To clarify the variant complex congenital cardiac defects, Van Praagh introduced a system of segmental sets to classify the majority of congenital heart diseases, but the code system entails some confusion for complete understanding. We attempted to recategorize the variant sets into four subgroups according to the connection of the atrial–ventricular and ventricular–arterial segments. This complexity can simply be grouped into four subgroups with regularities. From a simple table so formed, we can quickly ascertain the hemodynamics and the circulatory physiology, and therefore quickly determine the treatment protocol for variant complex hearts.

Key Words: anatomically corrected malposition, cardiac sets, concordance, discordance
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The simplest method for the categorization of complex congenital cardiac anomalies is to group them into cyanotic and non-cyanotic categories. Cyanosis is usually either a result of systemic venous return bypassing the pulmonary circulation and re-entering the systemic circulation without oxygenation or a result of diminished pulmonary flow caused by pulmonary stenosis or atresia. The former condition is caused by discordant connection of the cardiac segments at the atrial–ventricular or the ventricular–arterial level. By assembling the three cardiac segments, we can simply classify variant complex hearts into four subgroups. According to the concordance or discordance of the three cardiac segments, we can simplify the variant sets into a simple table. From this simple classification, we can easily understand hemodynamic physiology and establish the best mode of treatment.

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Methods and Results

To clarify variant complex congenital heart diseases (CHDs), Van Praagh introduced segmental sets to classify the majority of CHDs [1]. He used the visceroatrial situs status including situs solitus (S) and situs inversus (I), ventricular looping status (D-loop and L-loop), and the two great arterial relationships (S-, I-, D-, and L-position) to code complex hearts using a simple symbol {--,--,--}, but this code still entails some confusion to clearly understand what it signifies. For cardiac anomalies, the most important influential factors are the circulatory status and the severity of structural anomalies. To understand the circulatory physiology, we attempted to categorize CHDs into four groups according to the atrial–ventricular and ventricular–arterial status. From the connections between the atrial–ventricular and the ventricular–arterial segments, we can assemble the complex cardiac anomalies into four subgroups: (1) atrial–ventricular (A–V) concordance with ventricular–arterial (V–A) concordance as no discordance is Group A; (2) atrial–ventricular discordance with ventricular–arterial concordance as A–V discordance is Group B; (3) atrial–ventricular concordance with ventricular–arterial discordance as

Table 1. Cardiac segmental relationship

	Ventricular–arterial concordance	Ventricular–arterial discordance
Atrial–ventricular concordance	{S,D,S}/{I,L,I} {S,D,L}-ACM/{I,L,D} {S,D,I}-IIAI/{I,L,S}	{S,D,D}-TGA/{I,L,L} {S,D,L}-TGA/{I,L,D}
Atrial–ventricular discordance	{S,L,S}-IVI/{I,D,I} {S,L,I}-IAI/{I,D,S} {S,L,D}-ACM/{I,D,L}	{S,L,L}-ccTGA/{I,D,D} {S,L,D}-ccTGA/{I,D,L}

ACM = anatomically corrected malposition; IIAI = isolated infundibulo-arterial inversion; IVI = isolated ventricular inversion; IAI = isolated atrial inversion; TGA = transposition of the great arteries; ccTGA = congenital corrected transposition of the great arteries.

V–A discordance is Group C; and (4) atrial–ventricular discordance with ventricular–arterial discordance as double discordance is Group D.

In CHDs with atrial situs solitus with ventricular D-loop ({S,D,--}), this combination will present the condition of atrial–ventricular concordance and four types of anatomies including {S,D,S}, {S,D,I}, {S,D,D}, and {S,D,L}. In {S,D,S}, the ventricular–arterial relationship is concordance, so this type of heart can be categorized as Group A. In {S,D,D}, the ventricular–arterial relationship is V–A discordance with transposition of the great arteries (TGA), so it can be categorized as Group C. In {S,D,I}, it is V–A concordance and is grouped as Group A but with the character of isolated infundibulo-arterial inversion [2]. In {S,D,L}, there are two conditions in existence. One is ventricular–arterial concordance, which is characterized by anatomically corrected malposition of the great arteries ({S,D,L}-ACM), and this can be categorized as Group A; the other is ventricular–arterial discordance with the character of TGA ({S,D,L}-TGA) [3], and this can be categorized as Group C. {S,D,S} is most frequently seen in the normal circulatory heart, but may exist in rare cases of posterior TGA in which the aorta originates from the right ventricle, and the aorta is located rightward and posteriorly to the pulmonary artery.

In atrial situs solitus with ventricular L-loop, this combination will present the physiology of atrial–ventricular discordance, and there are four combinations including {S,L,S}, {S,L,I}, {S,L,D}, and {S,L,L}. In {S,L,S} and {S,L,I} hearts, the ventricular–arterial relationship is V–A concordance, so the two groups can be categorized as Group B, but {S,L,S} means isolated ventricular inversion [4] and {S,L,I} means isolated atrial inversion [5,6]. In {S,L,L}, the ventricular–arterial relationship is V–A discordance with TGA, so it can be categorized as Group D and characterized as congenitally corrected TGA. In {S,L,D}, there exists two subgroups: one is V–A concordance with the character of anatomically corrected malposition of the great arteries ({S,L,D}-ACM) [5] and categorized as Group B, and the other is a rare type of V–A discordance with congenitally corrected TGA ({S,L,D}-ccTGA) [3], and categorized as Group D.

In the condition of atria situs inversus, it will be the mirror images of atrial situs solitus. The same conditions exist in ventricular L-looping versus ventricular D-looping. We summarize the variant cardiac sets below and in Tables 1 and 2.

- Group A (no discordance): serial circulation without cyanosis, no need for treatment.
- Group B (A–V discordance): parallel circulation with cyanosis, needing atrial switch operation.
- Group C (V–A discordance): parallel circulation with cyanosis, needing arterial switch operation.
- Group D (double discordance): serial circulation, needing double switch operation for total correction.

Some rare entities such as atrioventricular valvular dysplasia as in tricuspid atresia, the single ventricular heart as in hypoplastic left heart syndrome, and other outlet malconnection as in double outlet right or left ventricle are not suitable for these categories.

Discussion

The problem when attempting to group the variant cardiac codes into a simple table is how to categorize the types of {S,D,L} and {S,L,D}. Except for the two types, every set in the biventricular heart has its own unique circulatory physiology, such as {S,L,L} which stands for A–V discordance and V–A discordance (the so-called ccTGA) and {S,D,D} which stands for A–V concordance and V–A discordance (the so-called complete TGA), but in types of {S,D,L} and {S,L,D},

Table 2. Circulatory relationship*

	V–A concordance	V–A discordance
A–V concordance	No treatment (Group A) RA → RV → PA AO ← LV ← LA	Arterial switch (Group C) RA → RV → AO LA ← LV ← PA
A–V discordance	Atrial switch (Group B) RA → LV → AO LA ← RV ← PA	Double switch (Group D) RA → LV → PA AO ← RV ← LA

*The arrows indicate the direction of blood flow. A–V = atrial–ventricular; V–A = ventricular–arterial; RA = right atrium; RV = right ventricle; PA = pulmonary artery; AO = aorta; LV = left ventricle; LA = left atrium.

there are two different circulations in existence in the biventricular heart.

In {S,D,L}, two distinct circulatory physiologies are found: one is {S,D,L}-ACM and the other is {S,D,L}-TGA. Twisting of the conotruncus in one direction and of the ventricular loop in the opposite direction appears to be of central importance in the morphogenesis of all types of ACM. In {S,D,L}-ACM, the conotruncus twists rightward instead of leftward. Persistence and growth of the subaortic part of the conus results in aortic-atrioventricular separation and fibrous discontinuity, and absorption of the subpulmonary part of the conus permits pulmonary-atrioventricular proximation and fibrous continuity [7]. In {S,L,D}-ACM, the same theory appears to be able to explain this entity. However, if there is presence of extensive levoposition of conotruncus with persistence of subaortic conus, and the aorta is located anteriorly and leftward to the pulmonary artery, the physiology of {S,D,L}-TGA will present [3]. In contrast, if there is presence of extensive dextroposition of conotruncus with massive subaortic conus, and the aorta is located anteriorly and rightward to the pulmonary artery, {S,L,D}-ccTGA will present.

From these simple categories, we can quickly ascertain the cardiac hemodynamics and circulatory physiology, and therefore quickly determine the treatment protocol for variant complex hearts. The prime information we need to know is the situs status of the atrium, the ventricular position, and the relationship of the great arteries, all of which can easily be obtained by noninvasive echocardiography. Our system of classification may reduce the confusion of medical students and inexperienced physicians alike in diagnosis of congenital cardiac anomalies. The exceptions not suitable for this system are the conditions of single ventricle and some types of double outlet right/left ventricle, but almost all types of biventricular cardiac anomalies are suited to this system of classification.

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複雜的先天性心臟連接異常的簡易分群

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為了使複雜的先天性心臟連接異常有統一的規範及名稱，**Van Praagh** 根據左右心房的位置，左右心室的位置及兩條大血管之間的相互關係，制定了一套命名法。這樣子的命名雖然可以清楚的描繪出心房、心室、及大血管個別的立體關係、但是卻存在著同一名稱有不同的解剖異常及學習與判斷其所對應的血流動力學關係上的困難、因此我們希望可以透過群組簡化的方式來使心臟的命名更容易被瞭解。我們根據心房與心室、心室與大血管的連接關係而將心臟的異常連接分為四個群組：**A** 組為正常血循、不須處理；**B** 組及 **C** 組均為發紺組、須接受心房轉位或大動脈轉位的處置；而 **D** 組則為 **B**、**C** 二組異常的合併、須接受心房轉位及大動脈轉位的雙重處置。我們發覺透過群組簡化的方式使這個命名法更容易被瞭解。

關鍵詞：解剖矯正性異位，心臟節段代號，正相連接，異相連接

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