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Background—We investigated the coronary arterial origins and course and the position of the great arteries in hearts with discordant ventriculoarterial connections. At the same time, we sought to evaluate the practicality of alphanumeric classifications in accounting for surgically relevant features of the coronary arteries.

Methods and Results—We studied 200 postmortem hearts, noting the patterns of coronary arterial branching, the vertical and horizontal location of the arterial orifices within the aortic sinuses, the course of the proximal coronary arteries in relation to the aortic wall, and the relations of the great arteries and their respective commissures. All hearts examined had concordant atrioventricular and discordant ventriculoarterial connections. We found 7 of the 8 predicted patterns for sinusal origin of the 3 major coronary arteries and identified 5 different positions of the arterial trunks relative to each other. A correlation was found between less frequent relationships of the arterial trunks and unusual patterns of coronary arterial branching, as well as with mismatch between the valvar commissures.

Conclusions—The surgically relevant features of the coronary arteries in hearts with discordant ventriculoarterial connections are best described rather than classified. Correlations exist between certain, less frequent relations of the great arteries and unusual patterns of branching of the coronary arteries. The presence of unusual great arterial positions should alert the surgeon to potentially complicated arrangements of the origin and distribution of the coronary arteries. **(*Circulation*. 2002;106:1980-1984.)**

Key Words: transposition of great vessels ■ coronary disease ■ surgery ■ anatomy ■ arteries

The arterial switch operation is now accepted as the surgical treatment of choice for patients with concordant atrioventricular and discordant ventriculoarterial connections (transposition). In many centers, the mortality rate for this procedure is remarkably low, but certain patterns of origin and distribution of the coronary arteries are recognized as increasing the surgical risk.[1,2] Thus, in a recent multicenter trial,[1] an increased risk for death was associated with origin of the right coronary artery from sinus 1, juxtacommissural origin of both coronary arteries, or intramural origin of a coronary artery. These abnormal features can now be diagnosed with some accuracy in most cases using echocardiography, thus obviating the need for angiography.[3,4] The definition of intramural origin, nonetheless, remains problematic. First defined on the basis of histological examination,[5] its reported frequency has varied considerably, almost certainly reflecting the definitions used for diagnosis.[1,3,6] Furthermore, other features, such as retropulmonary course of the left coronary artery or origin of both the right coronary artery and the left anterior descending from sinus 1, have been shown to be important.[2] These various features, all of potential surgical significance, are not the basis of the popular alphanumeric classifications presently used for description of these hearts.[7,8] In the present study, therefore, we sought to establish the frequency of the potentially relevant surgical features in a large series of autopsied hearts.

Methods

We selected 200 formalin-fixed specimens with concordant atrioventricular and discordant ventriculoarterial connections from the cardiopathological museum at Great Ormond Street Hospital for Children. Specimens with other atrioventricular connections were excluded. The ventricular septum was intact in 122 hearts (61%), whereas 78 hearts (39%) had at least one ventricular septal defect.

The origins of the coronary arteries were identified from the luminal aspect of the aorta. They were described as seen from the stance of an observer in the nonadjacent aortic sinus looking toward the pulmonary trunk. One sinus is then always to the right hand of the observer—sinus 1.[9] The other sinus is to the left hand of the observer—sinus 2. These definitions hold good irrespective of the relationships of the arterial trunks to each other.

In each specimen, we recorded the number and position of the coronary arterial orifices relative to sinuses 1 and 2. We then assessed the vertical location of each orifice relative to the sinutubular junction. The radial position of the orifice was described as being in the middle of the sinus or in its left or right third. We took particular note of those arteries positioned close to the commissure.

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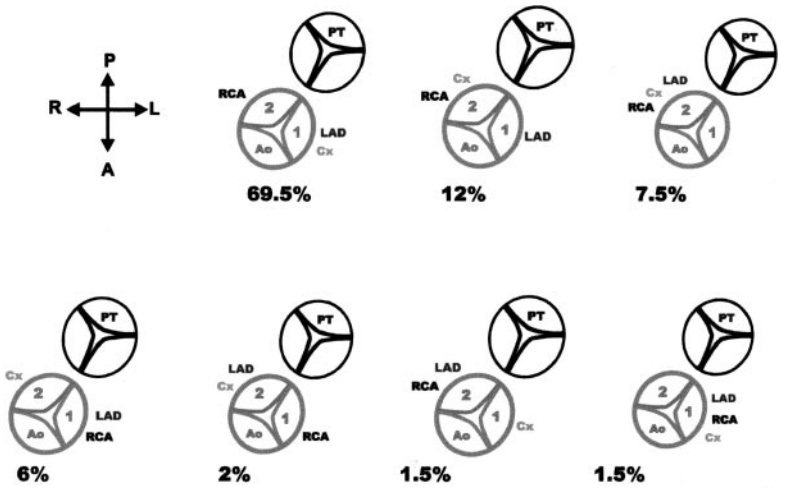

Figure 1. Schematic drawing showing the origins of the coronary arteries from the aortic sinuses. This cartoon is simplified to show the general arrangement and takes no cognizance, within each group, of other variables such as the relationship of the great arteries to each other, the position of the commissures, the position of the orifice within the sinus, and the proximal course of the coronary artery in relation to the aortic wall. All are shown as if the aorta is anterior and to the right, which is not always the case. Irrespective of the relationships of the arterial trunks, nonetheless, the arteries can always be described as taking origin from either sinus 1 or sinus 2. Ao indicates aorta; PT, pulmonary trunk; LAD, left anterior descending artery; CX, circumflex artery; RCA, right coronary artery; sinus 1, right-hand sinus; and sinus 2, left-hand sinus. The percentage of the total is shown beneath each schematic pattern.

By probing from the luminal aspect, we determined the orientation of the orifice of the coronary artery relative to the wall of the sinus and charted the initial course of the artery as it passed toward the interventricular or atrioventricular grooves. We found orthogonal, tangential, and intramural arrangements.[10]

We then examined the orientation of the sinuses of the aorta relative to those of the pulmonary trunk. In most cases, the commissures between the valvar leaflets faced each other at the sinutubular junctions. In a few cases, we discovered commissural mismatch. In addition to the orientation of the commissures, we established the relationships of the great arterial trunks, placing the heart as far as possible in the position it was presumed to have occupied during life. We recognized 5 categories. The aorta could be anterior and to the right of the pulmonary trunk, directly anterior, anterior and to the left, side by side with the aorta to the right, or posterior and to the right. The latter arrangement is also called normal relations, albeit with discordant ventriculoarterial connections.[11,12] The variations encountered were described using words but then analyzed in light of the commonly used alphanumeric classifications. Results are given in absolute numbers or in percentages. For statistical analysis, we used the Cochran-Mantel-Haenszel test based on table scores. A difference was considered statistically significant for $P<0.05$.

Results

All 200 hearts showed concordant atrioventricular and discordant ventriculoarterial connections. The oldest hearts dated from 1940. We found 7 different patterns of sinusal origin (Figure 1). The proportions varied in the hearts with and without a ventricular septal defect (Table 1). In all those deemed to have septal defects, the septum itself was otherwise well formed, and all hearts possessed concordant atrioventricular connections.

In 178 hearts, the anterior interventricular artery arose from sinus 1. The angle of origin was orthogonal in 170, tangential in 7, and intramural in 1 heart. The arterial orifice was in the mid third of the sinus in 127 hearts, in the left third in 28 hearts, and in the right third in 23 hearts.

The right coronary artery arose from sinus 1 in 14 hearts. In 12 of these hearts, its proximal course was orthogonal, whereas it was tangential in 2 hearts. Of these 14 hearts, 7 had the arterial orifice in the right third of the sinus, with 5 having orifices in the mid third and 2 in the left third.

In 174 hearts, the right coronary artery took its origin from sinus 2. In all but one of these hearts, the proximal course of the artery was orthogonal. There was tangential origin in the other. The orifice of the artery was in the middle third in 115 cases, in the left third in 42 cases, and in the right third in 17 cases.

In 22 hearts, the anterior interventricular artery arose from sinus 2, taking a retropulmonary course in 11 of these. In 8 hearts, it coursed anteriorly over the right ventricular outflow tract. In 2 hearts, it coursed between the great arterial trunks. An intramural origin was identified in both hearts with the latter pattern (Figure 2). In the remaining heart, a solitary artery ran behind the pulmonary trunk and branched to supply all three arteries. The right coronary artery continued anteriorly across the right ventricular outflow tract to reach the right atrioventricular groove. The proximal course of the anterior interventricular artery was orthogonal in 18 of these hearts and tangential in 2. In the other 2, as discussed, the artery had an intramural origin. In both of these hearts, the intramural segment was a main stem also feeding the circumflex artery, with its take-off above the sinutubular junction, juxtacommissural, and located in the right third of the sinus. In all cases, the main stem crossed the commissure as it coursed leftwards (Figure 2).

Taken together, in the 200 hearts, we found 203 arterial orifices in sinus 1 and 217 orifices in sinus 2. Take-off at or

TABLE 1. Patterns of Coronary Arterial Branching in 122 Hearts With Intact Ventricular Septation and 78 Hearts With One or More Ventricular Septal Defects

	Without VSD (%)	With VSD (%)
LAD, CX sinus 1; RCA sinus 2	86 (70.5)	53 (67.9)
LAD sinus 1; RCA, CX sinus 2	14 (11.4)	10 (12.8)
None sinus 1; LAD, RCA, CX sinus 2	8 (6.6)	7 (9.0)
LAD, RCA sinus 1; CX sinus 2	8 (6.6)	4 (5.1)
RCA sinus 1; LAD, CX sinus 2	3 (2.5)	1 (1.3)
LAD, RCA, CX sinus 1; none sinus 2	2 (1.6)	1 (1.3)
CX sinus 1; LAD, RCA sinus 2	1 (0.8)	2 (2.6)

Results are given in absolute numbers with percentages in parentheses. VSD indicates ventricular septal defect; LAD, left anterior descending artery; RCA, right coronary artery; and CX, circumflex artery.

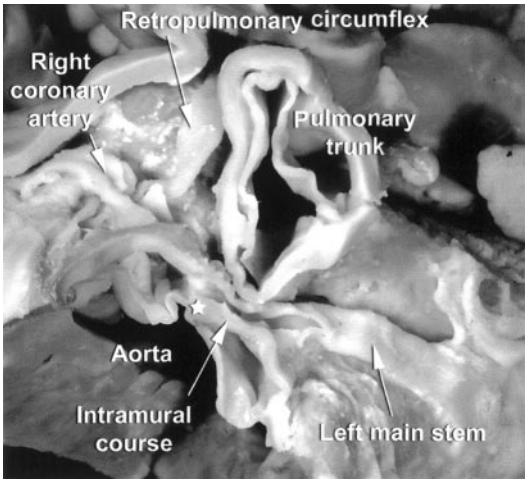

Figure 2. In this specimen, there is juxtacommissural origin (☆) of a main stem from the left-hand sinus (sinus 2). At the same time, there is high take-off of the artery.

above the sinutubular junction was observed relative to sinus 1 in 32 hearts (16%) and relative to sinus 2 in 32 hearts (again 16%). Of all 420 orifices, 10 were deemed juxtacommissural.

In 34 hearts, 2 arteries arose from the same sinus, but through separate orifices (Figure 3). The incidence of such dual sinusal origin was statistically significant in relation to the different patterns of coronary arterial branching ($P<0.001$, Table 2).

All the coronary arteries arose from the same sinus in 18 hearts, from sinus 2 in 15 hearts, and from sinus 1 in 3 hearts. In none of the hearts did the 3 arteries each have a separate orifice within the sinus. A solitary orifice was found in 7 hearts in which all arteries came from sinus 2 and 1 heart with origin of all arteries from sinus 1. Dual sinusal orifices were found in the other hearts in the absence of any orifices within the other sinuses. When all 3 arteries took their origin from sinus 2, the individual arteries themselves took markedly variable courses relative to the arterial pedicle (Figure 4). In one of the cases, a main stem initially took a retropulmonary

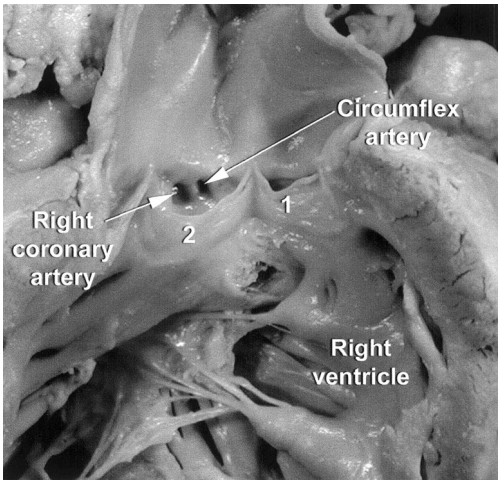

Figure 3. Dual origin of circumflex and right coronary arteries from the left-hand sinus (sinus 2). 1 and 2 indicate right-hand and left-hand sinuses, respectively.

TABLE 2. Origin of 2 Major Coronary Arteries From the Same Sinus Related to the Different Patterns of Branching

	Dual Origin (%)	Location of Dual Orifices
LAD, CX and acc RCA sinus 1; RCA sinus 2	0 of 139 (0)	Sinus 1
LAD sinus 1; RCA, CX sinus 2	8 of 24 (33)	Sinus 2
None sinus 1; LAD, CX and RCA sinus 2	7 of 15 (47)	Sinus 2
LAD, RCA sinus 1; CX sinus 2	9 of 12 (75)	Sinus 1
LAD, CX and RCA sinus 1; none sinus 2	1 of 3 (33)	Sinus 1
CX sinus 1; LAD and RCA sinus 2	2 of 3 (67)	Sinus 2
RCA in sinus 1; LAD, CX in sinus 2	3 of 4 (75)	Sinus 1

Results are given in absolute numbers with percentages in parentheses. LAD indicates left anterior descending artery; RCA, right coronary artery; CX, circumflex artery; and acc, accessory. A significant correlation is present between a certain coronary arterial branching pattern and the occurrence of dual origin, as determined by the Cochran-Mantel-Haenszel test based on table scores ($P<0.001$).

course, gave rise first to the circumflex artery, and then continued anteriorly to supply the anterior interventricular and right coronary arteries.

Accessory arteries arose within one of the sinuses in 9 hearts. In 6 hearts, accessory right coronary arteries supplied a considerable part of the right ventricle, all arising from sinus 1. In 2 hearts, accessory circumflex arteries arose from sinus 1 and supplied a small area within the left ventricular free wall. In the final case, the artery to the sinus node arose directly from sinus 1.

We found commissural mismatch (Figure 5) in 26 of the 200 hearts. In 7 hearts, the pulmonary valve had only two leaflets. Of these 7 hearts, there was mismatch relative to the aortic valve in 5.

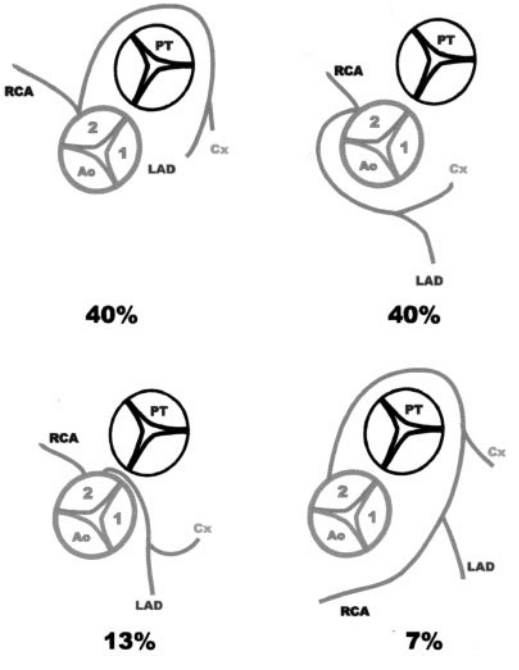

Figure 4. Schematic drawing of the variability in the 16 hearts in which all coronary arteries had their origin from the left-hand sinus (sinus 2). Seven of these hearts had single orifice, 9 hearts had double orifice.

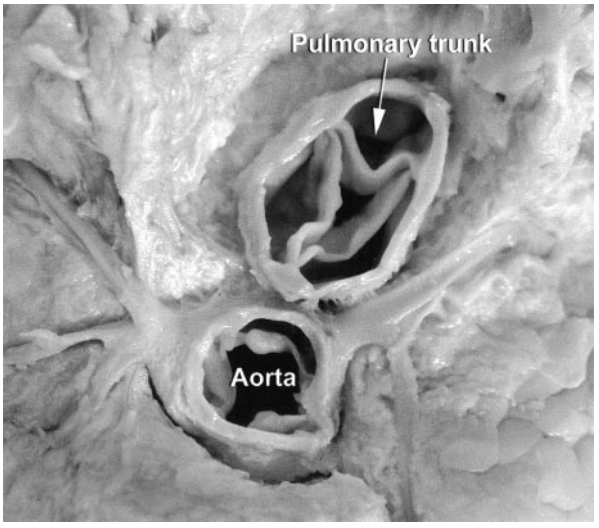

Figure 5. Commissural mismatch of the aortic and pulmonary valves.

The aorta was anterior and to the right relative to the pulmonary trunk in 148, directly anterior in 23, side by side in 22, anterior and to the left in 5, and posterior and to the right in 2 hearts. We found a statistically significant correlation between a certain position of the great arterial trunks and commissural mismatch ($P=0.036$, Table 3). Stronger correlation was found between a certain position of the great arterial trunks and an unusual pattern of coronary arterial branching ($P<0.001$, Table 3).

Discussion

The marked variability in the origin of the coronary arteries from the aortic sinuses has long been recognized in the setting of discordant ventriculoarterial connections, as has the varied relationships of the arterial trunks. It is the sinusal origin of the arteries that has most frequently been used as a means of classification.[5,8] Others previously had attempted to couple this information with the relationships of the arterial trunks, constructing in this way complicated alphanumeric classifications.[7] More recently it has been suggested that knowledge of the relationships of the arterial trunks themselves permits surgical inferences to be made concerning the arrangement of the coronary arteries.[13,14] Still others have emphasized the importance of the proximal course of the arteries relative to the arterial pedicles.[2] Overall experience, nonetheless, has shown that it is the so-called intramural origin that produces the greatest problems during surgical correction,[1] although some centers have shown the ability to neutralize this problem.[15]

TABLE 3. Unusual Patterns of Coronary Arterial Branching and Commissural Mismatch in the 5 Groups Defined According to the Relationship of the Aorta Relative to the Pulmonary Trunk

	Unusual Patterns (%)	Commissural Mismatch (%)
Anterior and to the right	37 of 148 (25)	19 of 148 (13)
Side by side, aorta to right	14 of 22 (64)	7 of 22 (32)
Directly anterior	5 of 23 (22)	None
Anterior and to the left	3 of 5 (60)	None
Posterior and to the right	2 of 2 (100)	None

Results are given in absolute numbers with percentages in parentheses. Description of position is always given from the stance of the aorta. There is a significant correlation between a certain position of the great arteries and the occurrence of an unusual coronary arterial branching pattern, as determined by the Cochran-Mantel-Haenszel test based on table scores ($P<0.001$). The correlation between a certain position of the great arteries and the existence of commissural mismatch, as determined by the Cochran-Mantel-Haenszel test based on table scores, is also significant ($P=0.036$).

Our study has confirmed that the relationships of the arterial trunks do correlate significantly with certain patterns of coronary arterial branching. We also found correlation with the feature of commissural mismatch. Origin of two or more arteries from one sinus, via one or two orifices, is also observed significantly more frequently in hearts with these unusual patterns of branching. Despite these correlations, however, we did not consider it feasible to attempt to present the huge amount of surgically relevant information in the form of an expanded alphanumeric classification. We believe it is much better to describe, rather than to try to classify, the surgically important aspects of the arrangements of the coronary arteries.

The arteries always take their origin from one or other of the aortic sinuses that are adjacent to the pulmonary trunk. As in the normal heart, even though we tend to speak of 3-vessel disease, it is most frequent for only 2 arteries to arise from the aortic root and almost always from separate aortic sinuses. Analysis and description in the hearts with discordant ventriculoarterial connections, however, are made easier by describing the origins of the right coronary, circumflex, and anterior interventricular branches, even though almost always 2 of these arteries arise from their respective sinus via a common stem. We are unaware of any well-documented example of origin of a coronary artery from the nonadjacent sinus in a heart with discordant ventriculoarterial connections. This means that only 8 basic patterns are possible for sinusal origin, excluding consideration of accessory orifices.[4,9] All but one of these patterns were represented in our material. The missing possibility, in which the right coronary and circumflex arteries arise from sinus 1 and the anterior interventricular artery originates from sinus 2, has similarly not been found in other large series.[4,5,15] Sinusal origin in itself, however, has not been shown to be of great surgical importance. Indeed, to the best of our knowledge, none of the features known to be surgically significant receive recognition in the alphanumeric classifications presently in vogue. This surely calls into question their surgical utility. We are well aware that to simplify communication and to establish collaborative databases, it will be necessary to provide some system of classification. This will require future international consensus, having agreed on the features of most surgical significance.

In our study, we tried to identify from anatomic findings the features that would likely have produced the greatest surgical challenge. In our judgment, these were intramural origin of a coronary artery, tangential origin and course, the relation and orientation of the orifice to the sinutubular junction, dual and single sinus origin, unusual relationship of

the arterial trunks to each other, and mismatch of the valvar commissures. Of these features, perhaps the best recognized at the present time is intramural origin. Even this feature creates problems when viewed from a morphological stance. Only a small proportion of our cases could be deemed intramural as initially described.[9] This anatomic pattern continues to be associated, in some series, with a high mortality.[1] We found another 8 cases with juxtaposition of an arterial origin to a commissure, although the coronary artery involved did not cross the commissure itself. Surgical correction might still have been difficult.

Strongly related to the intramural course itself were the features of tangential origin[10] and the origin relative to the sinutubular junction. High take-off also produces tangential attachment to the arterial wall, and this feature can lead to accidental injury when the aorta is opened. Of the 7 cases of tangential origin of the anterior interventricular artery from sinus 1, 5 also showed high take-off. These tangential origins might even be considered intramural, because the coronary artery was closely aligned to the aortic wall in its initial course. It is our belief that when such close attachment is encountered, echocardiography may overestimate the incidence of presumed intramural origin. Even the surgeon may have problems in differentiating a tangential from a true intramural course. Irrespective of definitions, when such close attachment to the aortic wall is present, surgical mobilization and creation of the button for reimplantation will certainly be more difficult.

Also of potential significance is the arterial origin relative to the middle, the left, or the right third of the space between the respective commissures. When two coronary arteries take origin from the same sinus, but through separate arterial orifices, the surgeon must decide whether there is enough tissue to permit their separation or whether they should be transferred together. The latter is usually the surgical maneuver. Such dual origin was much lower with the usual pattern of sinusal origin than in all the others. The problem of dual sinusal origin, therefore, should always be suspected with the less frequently encountered patterns of branching, themselves occurring more frequently with unusual arterial relationships. Of even greater concern than dual sinusal origin is when all 3 major coronary arteries arise from the same aortic sinus, albeit that we never found each artery with its own sinusal orifice.

The other feature of note in our study was commissural mismatch, found in one eighth of our material. This feature also, to date, has received relatively little surgical attention. It is to be expected when either of the arterial valves is bicuspid. But, even among the 184 hearts where both aortic and pulmonary valves were trifoliate, we observed considerable mismatch. The mismatch was encountered with only two patterns of relationship between the arterial trunks, namely when the aorta was anterior and to the right and when the arterial trunks were side by side. Surgical experience indicates that commissural mismatch complicates the transfer of the coronary arterial buttons during the arterial switch procedure. It is certainly a feature deserving of description and classification when large databases are to be established.

We did demonstrate a correlation between the patterns of relationship of the great arteries and unusual patterns of sinusal origin. The potential significance of such relationships of the great arterial trunks has been emphasized by Chiu et al.[14] If study of greater numbers confirms the correlations between certain relationships of the arterial trunks and unusual patterns of coronary arterial branching, then it may prove possible to identify preoperatively those patients at increased risk during the arterial switch procedure. At present, nonetheless, the correlation is insufficiently robust to permit the drawing of anything more than inferences. In this respect, nonetheless, analyzing the coronary arteries as arising from sinuses 1 and 2, as seen from the viewpoint of the nonadjacent sinus, permits the different relationships between the arterial trunks to be ruled out as an independent variable.

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