Can We Better Understand the Known Variations in Coronary Arterial Anatomy?

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Coronary arterial anatomy is remarkably diverse. Identification of surgical risk factors, however, requires description in a uniform fashion. Such description mandates that account be given of both aortic sinusal origin and variability in aortopulmonary relationships. Currently, however, it is rare to find all this information provided either in clinical reports or published reviews. In this review, therefore, we summarize why both these features are important, emphasizing the marriage of convenience between the aortic root position within the cardiac base and the arrangement of the epicardial coronary arteries. The inductive approach accounts for all potential variations.

What Determines the Anatomy of the Coronary Arteries?

The major coronary arteries occupy the atrioventricular and interventricular grooves [18]. Although there are 4 such grooves—right and left atrioventricular and anterior and inferior interventricular—we describe only 3 major coronary arteries. This is because the inferior interventricular artery, usually described incorrectly as the posterior descending artery [19], takes its origin from 1 or the other, or sometimes both, of the 2 arteries located within the atrioventricular grooves. These arteries, the right coronary artery (RCA) and the circumflex coronary artery (CXA), extend around the orifices of the tricuspid and mitral valves, respectively. The third major coronary artery is located in the anterior interventricular groove (anterior interventricular artery [AIVA]), also known as the left anterior descending (LAD) artery. The arteries take their proximal origin from the aortic valvar sinuses that are adjacent to the pulmonary trunk [18, 20, 21], with the right-sided adjacent aortic sinus giving rise to the RCA and the left-sided adjacent sinus to the CXA and AIVA, usually through a short common stem. This normal pattern can be described as dual sinus origin. Its essential feature is that irrespective of the relationship of the arterial roots to each other, coronary arteries enter the aortic root from each side. An alternative description therefore is bilateral root entry (Fig 1A). One well-recognized variation is seen when the CXA extends through the transverse pericardial sinus, taking its origin from the right-sided adjacent aortic sinus (Fig 1B). Another important variation is found when either the RCA
or the main stem of the left coronary artery runs between the subpulmonary infundibulum and the aortic root, taking origin from the opposite adjacent aortic sinus [1, 22]. This produces an interarterial variation, which can exist with or without an additional intramural variant (Fig 2) [23]. Other variations include those in which a single coronary artery takes its origin from either side of the aortic root, or all major coronary arteries arise from the same sinus in the absence of interarterial coursing. These patterns, with their variations [22, 24, 25], can collectively be described as unilateral root entry, thus distinguishing the arrangements from the more usual situations in which the coronary arteries take origin from each side of the aortic root, in other words bilateral root entry. As we will show, these differences are crucial in understanding the overall variations in coronary arterial anatomy.

Embryologic Development of the Coronary Arteries

In the past, concepts based on embryology have been said to serve as a hindrance rather than a help in analyzing normal and abnormal anatomic arrangements [26]. Nonetheless, advances in the understanding of the mechanisms of development require reevaluation of this opinion. Thus all of the normal patterns for coronary artery disposition, along with their variations, can now readily be understood on the basis of recent findings concerning their embryologic development [20, 27–30]. The major epicardial coronary arteries attain their aortic connections relatively late in development, subsequent to the process of aortopulmonary rotation [28, 30]. It used to be thought that the proximal parts of the coronary arteries budded out from the aortic sinuses. It is now accepted that the coronary arteries develop within the epicardial atrioventricular and interventricular grooves and that their proximal parts grow into the aortic valvar sinuses [27].
The existence of arteries coursing anterior or posterior to the arterial roots also supports the notion that a crown-like configuration of arterial primordia is to be found around the developing aortic and pulmonary roots [29]. Since the major coronary arteries hardly ever take origin from the nonadajacent aortic sinus, and when arising anomalously from the pulmonary trunk also frequently arise from adjacent sinuses [31], factors related to the separation of the arterial roots by fusion of the major outflow cushions almost certainly guide their proximal in-growth toward the valvar sinuses [20]. This notion is further supported by the marked variability in sinusal origin of the coronary arteries in the setting of a common arterial trunk [32, 33], which exists because of failure of septation of the developing outflow tract [34]. An exaggeration of this guiding pattern would also be sufficient to direct one or the other of the right and left coronary arteries beyond its intended site of origin, producing the intramural variant with single-sinus origin but bilateral root entry [35, 36]. We can expect therefor e that these intramural patterns will be seen as variants of the patterns already well recognized for dual sinus entry [11, 36]. It is also intuitive to suggest that part of the guiding mechanism depends on the proximity of the 2 adjacent aortic sinuses to the atrioventricular and anterior interventricular groove. Since the coronary arteries must perform the sinusal wall only subsequent to the establishment of aortic position, it is the final location of the aortic root relative to the cardiac base that is responsible for producing the marriage of convenience between the major coronary arteries and their sinuses of origin [11, 16, 17, 37, 38].

**Evidence From Previous Descriptions**

Review of publications describing the patterns of the coronary arteries in transposition, congenitally corrected transposition, and tetralogy of Fallot shows that the coronary arteries are not always arranged in their anticipated fashion and that this fact also holds for the otherwise normal heart [1, 2, 9]. In tetralogy of Fallot, eg, the rightward location of the aorta reduces the distance between sinus #1 and the AIVA, thus explaining why an AIVA is often found crossing the narrowed subpulmonic infundibulum (Fig 3c) [39]. Furthermore, the anomalous course in this setting is more frequent when the aortic root is rotated and located even more anteriorly [40]. Nonetheless, the greatest variation in coronary artery arrangement [8, 14] is found when transposed arterial trunks exist in the setting of concordant atrioventricular connections [41], in other words “regular” transposition (Fig 3). In this setting, the right-sided aortic valvar sinus, known as sinus #2, is the one that is closest to the right atrioventricular groove. An artery exiting from this sinus is able not only to enter the anterior groove but also to give a branch that passes directly into the left atrioventricular groove. It is not surprising therefore that the most frequent arterial relationship in transposition (when the aorta is positioned rightward and anterior) is associated with the origin of the AIVA and CXA from sinus #1 and the RCA arising from sinus #2 (Fig 3a). There are nonetheless multiple alternate patterns. Most existing classifications were devised in attempts to cater for description of these variations. No single classification has achieved universal acceptance. The classification of Yacoub and Radley-Smith [7, 42], eg, accounts for relatively few of the known variations. It also mixes the features used for classification, with 4 of the described patterns accounting for varying sinusal origin of the main coronary arteries, but 2 of them (types B and C) showing the same sinusal anatomy. This classification was subsequently expanded to include details of the course of the proximal coronary arteries relative to the arterial pedicle, a feature of obvious surgical significance [43–45]. In this regard, the feature of the second most common pattern is for the CXA to run a retropulmonary course, passing
through the transverse pericardial sinus to reach the left atrioventricular groove (Fig 3b). This retropulmonary passage of the CXA can also be found when both the RCA and AIVA arise from sinus #1 (Fig 3c). Furthermore, the latter patterns with retropulmonary CXA are known to be more frequent when the arterial trunks are rotated from the oblique orientation toward a more side-by-side arrangement [10, 42–44]. One of the oldest classifications, that proposed by Elliott and colleagues [46], had previously emphasized this effect of rotation of the aortic root. The clockwise rotation of the aortic root from the anticipated right anterior location takes the left-handed aortic sinus into a posterior position and closer to both the transverse pericardial sinus and the left atrioventricular groove (Fig 3b, 3c). This influence of the location of the aortic root relative to the cardiac base also offers an explanation for why, when the aorta arises from the right ventricle but in a posterior position (Fig 3d) (the arrangement known as posterior transposition), the coronary arteries arise in the fashion expected for the normal heart [11, 47]. It also explains why, when the aorta is anterior but leftward located in so-called regular transposition, the RCA has to cross in front of the pulmonary trunk to take origin, along with the AIVA, from sinus #2 (Fig 3e) [11, 48].

Taken together, these associations point to the significant influence of aortic position in determining sinusal origin of the coronary arteries [12, 37, 48–50]. There are nonetheless other subtle variations to be found in arterial anatomy, such as duplication or absence of the major epicardial branches. An attempt to incorporate all this information into one system [8] produced huge complexity (Fig 4). Another system, devised by Gittenberger-de Groot and colleagues [12], with its roots in the doctoral thesis of Quaegebeur [13], removed the necessity for describing the location of the aortic root. These studies showed that irrespective of arterial relationships, the 2 aortic sinuses adjacent to the pulmonary root could always be designated as being to the right or left of the observer standing in the nonadjacent sinus and codified as #1 and #2. Although the system is now well accepted by the surgical community [4, 13–15, 37, 51, 52], it fails to provide for each individual patient the crucial information concerning the location of the aortic root. It is only when we take note of the location of the aortic root in addition to sinusal origin that we can explain one of the conundrums that remains concerning the arrangement of the coronary arteries in the setting of transposed arterial trunks.

Fig 4. Numeric-alphabetic classification devised by Shaher and Puddu [8], combining information concerning sinusal origin and the epicardial course of the coronary arteries. All variants drawn in a fixed aortic position rightward and anteriorly have been modified to show the more typical locations of the aortic root for the various patterns identified [11]. Type 9, the arrangement seen in the normal heart, is a mirror image of type 1. Types 3, 5, and 7 illustrate variants of single coronary artery, whereas types 2, 4, 6, and 8 elucidate various types of unusual circumflex coronary artery.
The “Missing Option” in Patients With Transposition

When one of us (RHA) was working with the archive of congenitally malformed hearts housed at Children’s Hospital of Pittsburgh, a selection was made at random of 100 hearts with concordant atrioventricular and discordant ventriculoarterial connections. At the time, RHA was convinced that the system proposed by Quaegebeur [13] was the optimal means of describing coronary arterial anatomy, so as discussed earlier, he neglected to note the specific relationship of the transposed aortic root relative to the ventricular base when describing the sinusal origins. It remains impressive, nonetheless, that 7 of the 8 possibilities predicted on the basis of 3 coronary arteries arising from only 2 aortic sinuses [15, 41] were present within the small group of hearts selected from the archive (Fig 5, first 2 rows). The question was not posed, however, as to why one of the patterns had not been encountered. Our subsequent review of the salient published literature providing details on more than 6,000 patients [3, 4, 7–9, 11, 12, 35, 42–44, 46, 48–50, 53, 54] now shows that this option was not only missing in the 100 hearts selected at random from Pittsburgh but also that it has never been described in the setting of “regular” transposition. The missing option exists when the RCA and the CXA are supposed to arise from sinus #1 and the AIVA is supposed to arise from sinus #2 (Fig 5, third row middle). We are now able, based on marriage of convenience, to explain the lack of this option. As already discussed, usually the aorta is anterior and rightward in patients with typical transposition. In this situation, sinus #2 is located rightward and posterior relative to the heart itself. If the AIVA was taking origin from this sinus, as would be necessary to produce the missing option, this artery would need to cross the epicardial territory of either the RCA or the CXA to reach the anterior interventricular groove (Fig 5, gray figure in third row left). This is unlikely to happen. The only situation that would favor the missing option on the basis of marriage of convenience is when the aortic root is positioned posterior and leftward rela-

Fig 5. Eight theoretical possibilities for sinusal origin of the coronary artery patterns on the basis of 3 major coronary arteries taking origin from the 2 adjacent aortic valvar sinuses. The numbers show the frequency of the various patterns discovered within a cohort of 100 hearts selected from the archive of congenitally malformed hearts held at Children’s Hospital of Pittsburgh. The 8 potential patterns in three rows include the 2 variations with origin of a solitary coronary artery, which is shown in the middle and right panels, middle row. These 2 patterns then have multiple variations in terms of epicardial branching, which are grouped together here on the basis of identical sinusal origin. The remaining 6 possibilities are those with bilateral root entry. (Left-hand panel, bottom row) The missing option is impossible when the aorta is anterior and rightward. (Right hand panel, bottom row) The missing option is possible if the aorta is positioned posteriorly and leftward. This aortic position, however, has never been encountered in patients with “regular” transposition. (A = anterior ventricular artery; L = circumflex coronary artery; PT = pulmonary trunk; R = right coronary artery.)
tive to the pulmonary trunk so that both the RCA and the CXA could arise from sinus #1, whereas the AIVA would arise from sinus #2. This aortic position has yet to be described in regular transposition but does exist when congenitally corrected transposition coexists with a left posterior aortic root (Fig 6A) [16] or when the arterial trunks arise concordantly and in parallel fashion from the ventricular mass, but with the aorta posteriorly located relative to the pulmonary trunk (Fig 6B). In this instance, the retroaortic CXA arose from the RCA, which itself took origin from sinus #2. (C) This pattern is a more frequent finding in the otherwise normal heart, as shown in Figure 1B. In this setting, an almost directly posterior aortic position produces greater adjacency between the left atrioventricular groove and sinus #1.

How Should We Describe the Variations in Coronary Artery Pattern?

The sophistication in diagnostic techniques is now sufficiently great for nearly all the crucial information relating to coronary artery patterns to be provided preoperatively [22, 56]. This information should now include analysis of the precise relationship of the aorta to the pulmonary trunk, as well as the location of the facing commissure and sinuses [8, 57]; the sinusal origin of the 3 major coronary arteries; information relating to ectopic location, including eccentric origin and high or low takeoff [1, 21, 35, 36, 57]; intramural course of their orifices; their epicardial course relative to the arterial pedicles; and salient information concerning the individual branching of the arteries themselves, such as dual origin of anterior interventricular arteries. Ideally, descriptions should also include details of the origin of the artery to
the sinus node [50, 58], which may need to be redi-
rected if it originates directly from an aortic valvar
sinus (Fig 4), albeit current resolution of computed
tomography still makes the identification of this detail
difficult. The diagnostician will hopefully, nonetheless,
seek to provide as much information as possible for
each patient being discussed for corrective surgery.
When we compare the accounts given by different
groups, all this information may prove to be of clinical
significance. It is the information regarding the loca-
tion of the aortic root, however, that currently is most
often ignored [4, 14, 15, 51, 52].

Patterns With Origins From Two Aortic Sinuses and
With Bilateral Entry to the Aortic Root
Sinusal origin and aortic position are closely related in
terms of the development of the given patterns to the
arrangements seen in those patients in whom 2 aortic
sinuses give rise to 3 major coronary arteries, these
sinuses almost without exception being adjacent to the
pulmonary trunk. It was recognition of this association
that underscored the system proposed by our group
working in Taiwan, which uses the concept of “2 × 3” to
produce a unifying system for the various patterns seen
in transposition [11, 54, 59] and other congenitally mal-
formed hearts [16, 17, 40]. We now recognize the need to
construct 2 circles to provide full understanding of the
potential patterns (Fig 7). The first circle accounts for the
marriage of convenience between the rotated aortic root
and the proximal orifices of the coronary arteries located
in 3 anatomic grooves in the setting of right-hand ven-
tricular topology, whereas the second circle accounts for
left-hand topology [16, 59]. Both circles are proposed
because in the presence of left-handed topology, the
morphologically right and left atrioventricular grooves
are on the opposite side of the heart relative to the facing
coronary sinus origins (Fig 6A). In the setting of left-
hand topology, the CXA, occupying the right-sided atrio-
ventricular groove, is closest to sinus #2, as in the usual
variant of congenitally corrected transposition, and
therefore usually arises from this sinus, along with the
AIVA (Fig 7B, illustrated at the 3 o’clock position). Ap-
preciation of the orientation of the aortic root within our
2 suggested circles can therefore provide a rational ex-
planation for all the known anomalous patterns of the
major coronary arteries in the setting of dual sinus origin
with bilateral root entry.

Patterns in Which the Coronary Arteries Arise From a
Single Aortic Sinus but With Bilateral Entry to the
Aortic Root
Appreciation of the orientation of the aortic root rela-
tive to the cardiac base inside the circle in Figure 8A
also provides a rational explanation for the anomalous

Fig 7. The 6 potential options for bilateral root entry illustrated relative to most likely locations of the aortic root. (A) Options for right-hand (RH) ventricular topology. (B) Left-hand (LH) topology. The patterns reported most frequently for transposition (TGA), tetralogy of Fallot (TF), congenitally corrected transposition (CCT), and concordant ventriculoarterial connection with parallel arterial trunks (Conc VA conn with PAT) and otherwise normal heart each occupy a different portion of the horizontal circle representing the location of the aortic root within the cardiac base. The gap between panels (d) and (e) in Figure 3 is now filled by the missing option in Figure 6. In patients with left-hand ventricular topology, the left-sided atrioventricular groove contains the morphologic right coronary artery (RCA), whereas the right-sided groove contains the morphologic circumflex coronary artery (CXA). (A = anterior inter-
ventricular artery; L = circumflex coronary artery; R = right coronary artery.)
coronary artery patterns observed when both arteries take their origin from the same sinus but with the arteries entering from opposite sides of the aortic root, one or the other then taking an interarterial course (Fig 8A, outside the circle). The pattern illustrated between the 10 and 11 o’clock positions outside the circle in Figure 8A is the interarterial and intramural variant shown in Figure 2, known now to be a harbinger of sudden cardiac death.

Patterns With Unilateral Entry to the Aortic Root
Our review of the salient literature indicates that aortic position varies not only for the different patterns seen when coronary arteries enter through both sides of the aortic root but also when all the coronary arteries enter the aortic root from the same side (Fig 8B), these being the variants for single coronary arteries. The specific types discovered in each disease entity labeled in the center have occupied different portions of the circle, suggesting that these abnormal patterns are also influenced by the location of the aortic root. (AVG = atrioventricular groove; CCT = congenitally corrected transposition; IVG = interventricular groove; TF = tetralogy of Fallot; TGA = transposition.)

Surgical Implications
It could be argued that the full clinical significance of the multiple variations we have reviewed has yet to be established. It might also be thought that intraventricular anatomy also influences coronary artery arrangements. We are unaware of any convincing evidence supporting this latter notion. Indeed, evidence supporting such a suggestion could not be assembled unless a unified concept is constructed to provide the basis for understanding all the variations. Our hope is that our review might provide the impetus for agreement on such a concept. Nonetheless, in the current surgical climate, information concerning the origins and course of the coronary arteries, and their orientation to the aortic root, is of most significance to those performing the arterial switch procedure, in which optimal redirection of all the coronary arteries in situ remains the cornerstone of surgical success [61, 62]. All known patterns are amenable to transfer [3, 53], although the late recurrence of coronary artery obstruction, which has been reported to be as high as 3% to 7.8% [63, 64], may well be related to variations in coronary arterial anatomy [50, 53]. We have already proposed diagnostic techniques for the recognition of potentially dangerous coronary artery patterns and surgical techniques for their correction [36, 57, 62]. In particular, when redirecting the origin of coronary arteries adjacent to the facing commissure, or an artery passing intramurally across this commissure, we consider it advantageous to create a single arterial button by means of a superiorly based trapdoor. In addition to the short axis rotation, which reflects the juxtaomissural origin of the coronary artery [36], it is also frequently necessary to be aware of additional rotation in the long
axis should the abnormal coronary artery also show high takeoff [50, 57], this being a frequent additional finding in the setting of intramural origin.

Conclusions

We suggest that in the future when charting details of surgical correction of coronary artery anomalies or describing the operative procedure for the arterial switch, information should be provided not only of sinusual origin but also of the location of the aortic root relative to the cardiac base. Emphasis should also be given to whether the coronary arteries enter the aortic root from both sides or whether there is unilateral entry. Only when all this information is widely available will we be able to judge whether it is truly possible to provide a universally acceptable classification for coronary arterial anatomy. Irrespective of whether we achieve a suitable classification, the provision of information concerning aortic position as well as sinusual origin will make it easier to assess the remaining surgical risk factors for procedures such as the arterial switch.

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References


