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Coronary arterial anomalies and variations

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ABSTRACT

A clear-cut and clinically-useful definition of a coronary anomaly is frequently not easy to obtain; many variables are to be taken into account. Because no unanimous agreement exist on the terms and classifications to be used, comparisons between various centers and between different series, are thus difficult to perform. The finding of an coronary arterial anomaly eventually raises lots of further questions due to the fact that it is difficult to make a parallel between an anatomical modification and its alleged clinical consequences, especially regarding the most severe ones such as dysrhythmias, myocardial ischemia and sudden death. The present review is intended to represent an aid to the clinician and aims to bridge the inherent gaps between the points of view of the specialists who directly approach this topic: angiographist, cardiologist, cardiac surgeon, pathologist. Thus, useful definitions, classifications, data of clinical relevance and treatment guidelines, are given - especially under the light of the new theories and updated concepts.

Key words: congenital heart disease, coronary anomalies, coronary circulation, myocardial ischemia, sudden death

INTRODUCTION

The coronary system consisting of two coronary arteries appears to be a relatively recent evolutionary acquisition [1]: fish and amphibia have only one coronary artery and only 60% of avian species have two coronaries. The human coronary arteries show a predominantly subepicardial course and frequent intramural course (the so-called myocardial bridging) – a disposition between that found in some species with a completely intramyocardial course (rat, guinea pig, hamster) and respectively, of other species, with entirely subepicaridal arteries (horse, cow, pig).

The right and left coronary arteries normally originate from the homonymous aortic sinuses adjacent to the pulmonary trunk (the facing sinuses). For a clearer definition and classification of coronary arterial disposition especially in complex congenital cardiopathies, a useful system was developed [2] (Figure 1). This scheme allows a universal and precise characterization of any type of coronary variation or anomaly, no matter the relative position of the aorta and pulmonary trunk to each other or related to the remainder cardiac structures

The right coronary artery usually courses as a single trunk; the left main coronary artery generates the left anterior descending and the circumflex branches: thus, three coronary arterial trunks eventually result. Taking into account the variability of coronary arterial origin and proximal course, the variable length of the left main trunk as well as the various diagnostic and therapeutic implications, attention has been focused on the three essential, elementary coronary trunks: left anterior descending (LAD), circumflex (Cx), and right coronary artery (RCA) (Table 1). ❑

***The definition of a coronary artery should be made without taking into account its origin and proximal course but focusing on its intermediate and distal segments and/or its dependent microvascular bed.*[3] ❑**

The coronary circulation can be divided into extramural and intramural portions.

The extramural component comprises the three essential trunks and their primary divisions. They run in the subepicardial layer showing a tortuous course (a feature already present in the newborn), due to the fixation to the myocardium by means of the penetrating intramural branches and to the direction of the vessels which predominantly coincides with that of the heart movements. A major characteristic of the extramural vessels unique to the coronary arteries is their subintimal fibro-muscular-elastic thickening, already developing during the first months of life.

The intramural vessels penetrate deep into the myocardium. They originate approximately at right angle and follow a more or less perpendicular course to the plane of the ventricles. The intramural branches course through the whole thickness of the ventricular wall giving off successive ramifications which will eventually form a fine vascular network present ***at all levels***[4] (rather than only at subendocardial or subepicardial levels, as previously thought).

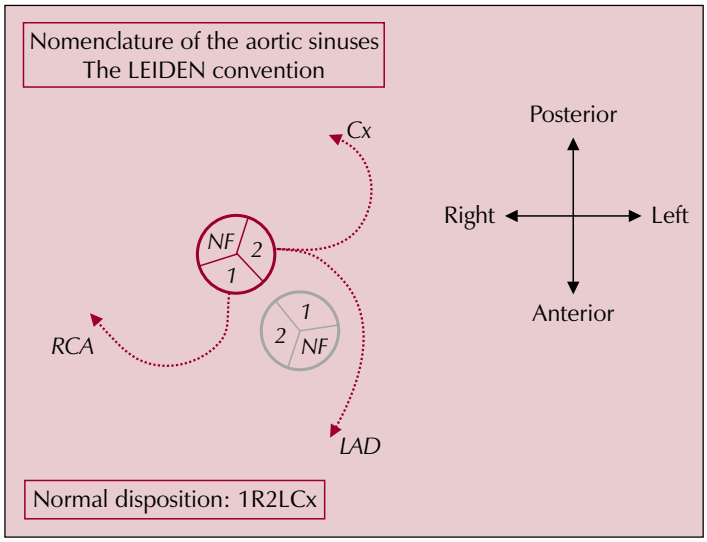

FIGURE 1. Nomenclature of the aortic sinuses ("The Leiden Convention")[2]

A schematic representation of the aortic and pulmonary roots as viewed from "above". The pulmonary root is depicted in blue color while the aortic root is in red. The origins and the approximate initial tract of the coronary arteries is shown with dotted lines. Three coronary trunks eventually result: the left anterior descending (LAD) , the circumflex (Cx) and the right coronary (RCA) respectively. The adjacent aortic and pulmonary sinuses (the facing sinuses) are numerated as "sinus 1" and "sinus 2" while the non-adjacent (non-facing sinuses) are indicated as "NF". For a hypothetical observer located within the aortic NF sinus, the right-handed sinus is the aortic sinus 1 (the classical right coronary sinus) and the left-handed, the aortic sinus 2 (the classical left coronary sinus). In an analogous way, the pulmonary facing sinuses a can also be defined. The normal disposition is thus: 1R2LCx.

CORONARY ARTERY	MINIMALLY REQUIRED FEATURES
Left anterior descending (LAD)	Location: the anterior interventricular sulcus Subepicardial position (but not infrequently intramyocardial) Provides septal branches and follows the direction of the septum. Accompanied by a conspicuous venous branch (greater cardiac vein)[A]
Circumflex (Cx)	Location: the left side of the coronary sulcus Subepicardial position Provides at least one marginal branch[B]
Right coronary artery (RCA)	Location: the right side of the coronary sulcus Subepicardial position Provides at least the right ("acute") marginal branch[C]

TABLE 1. The elementary coronary arteries (Adapted from Ref.[3])

[A]These represent the essential features of the LAD, no matter what origin it might have (e.g. not always from the aortic sinus 2, as it occurs in TGA). Its proximal course can also be very different (pre-infundibular, between the aorta and pulmonary trunk or intraseptal). The LAD can be split or doubled; it may give off few diagonal branches. It may also show an important and longer recurrent tract at level of the posterior interventricular sulcus. Even in complex cardiac malformations, such as in criss-cross hearts with a horizontal interventricular septum , the LAD always follows the septum.

[B]In some instances, the Cx might show a more "atrial" or respectively, a more "ventricular" course (i.e. not lying exactly at the level of the coronary sulcus) - a disposition of surgical relevance

[C]Variations in caliber and length of the RCA should be interpreted taking into account the conus artery (which might have a separate origin or which at times can be of considerable dimension: "the third coronary artery"). The conus artery may also originate at level of the aortic sinus 1 (e.g. TGA) The RCA might practically end (or become insignificant) after giving off the right (acute) marginal branch. The origin and course of the sinus node artery are very important for the surgeon (e.g. the atrial switch operation: Mustard or Senning).

Abundant anastomoses are present between the branches of the same coronary trunk (intra- or homo-coronary anastomoses) or between branches of different coronary trunks (inter- or hetero-coronary anastomoses). Intracoronary anastomoses are shorter (1-2 cm) and somewhat more slender (diameter of 20-250 µm) as compared with the intercoronary anastomoses (2-3 cm and respectively 20-350 µm)[4] ❑

The intramural coronary vessels together with the cardiac interstitial connective tissue, form the intimate skeleton of the heart. This "skeleton" must be looked upon as a dynamic structure that also offers an explanation for the non-constant behavior of the diastolic ventricular distensibility. ❑

THE DEFINITION OF "A CORONARY ANOMALY"

Like many other tissues and organs in the body, the coronary arterial system can show variable features that can be regarded either as "normal" or "abnormal". A clear distinction is often difficult to make as many of the coronary arterial variations are at the innocuous end of the broad clinical spectrum of possible consequences; in other cases, a direct causal relationship between a coronary anomaly and an unusual event as sudden death, is difficult to prove.

Even more, variations regarding for example the number of ostia, location, size, the proximal course, may actually mean nothing and this aspect can be certified by using the various diagnostic methods that prove the adequacy of myocardial perfusion.

An important detail is represented by ***the regional distribution of a coronary artery***, its actual denomination and origin being of less importance.

The surgeon or the hemodynamist must have clear in mind which branch vascularizes a given territory and how many of such vessels must be approached; their actual origin or denomination is of secondary importance. For example, a diagonal branch can represent in some cases the main source of blood to the anterior left ventricular wall and to the mitral anterior papillary muscle group [5], while in others, it may just be a slender and less important branch.

Probably, unless a major anomaly exists (vide infra), the subepicardial course of a coronary artery is of no particular significance or it may represent an alteration of the normal process of coronary genesis, in which case however, a clear-cut demarcation between "normal" and "abnormal" is not as obvious as it might be expected.

In defining an abnormality or variation, some important features of the coronary arteries should be taken into account (**Table 2**). Following the criteria presented in the table, "abnormality" can be looked upon as ***a quantitative variation*** (e.g. number of ostia) or by demonstrating ***very particular features***: very small ostium or trunk, acute angle of origin, obstructive membrane, lack of the proximal part of one coronary. With some other features, ***empirical criteria*** could be applied: *"what is observed in less than 1% of the population or what is more that 2 standard deviations of a Gaussian distribution curve"* [3]. Strict definitions should thus be issued by certified groups of experts on larger statistics - a task still difficult to accomplish.

LEVEL	VARIABLES
1.Ostium	Number of ostia
	Location
	Size
	Angle of origination
	Shape (e.g. slit-like; membrane)
2. Size	Small size Presence of a diaphragm
3. Proximal course	Especially intramural tract[A] Consider angle of origin[B]
4. Mid-course	Intraseptal tract or looping[A]
5. Intramyocardial ramifications	Regional distribution[A]
6. Termination	Regional distribution[A]

TABLE 2. The variable features of the coronary arteries[3]

[A]Possible surgical relevance
[B]Intravascular ultrasound examination valuable

It is also difficult and quite unnatural to call as "anomaly", a variation observed only at the level of the larger conduits (subepicardial vessels and their main branches) while ignoring their effect or associated lesions at microcirculatory level. Cases with coronary stenoses or occlusions and normal myocardial scintigram or in otherwise normal people [6] are encountered; on the other hand, cases with normal coronary arteries and altered myocardial perfusion (syndrome X) are also present. There is no relationship between the number of stenoses, degree of stenosis and the severity of ischemic heart disease and no correlation with the location, size and severity of an infarct [7]. The picture is thus complex and incompletely resolved yet.

Even when taking into account the three elementary coronary trunks, a pertinent comparison between different studies can not be made and the often-used classification of coronary lesions in *"one-, two- or three-vessel disease"* has little significance if no mention is made regarding the distal territory of each trunk, the coronary typology and the relative balance between the trunks.

The clinical consequences do represent another valid criteria for defining a coronary anomaly. There are anomalies involving obligatory ischemia , such as the origin of the left coronary artery from the pulmonary trunk [8]. At the other end of the spectrum there are anomalies not associated with ischemia. In between these categories, there is an ill-defined group of anomalies involving exceptional ischemia, that allow a normal life and even athletic activity [3].

*Many **questions** arise at this point, and these should be carefully evaluated by every physician when approaching a patient with a coronary anomaly or variation:*

- Following the various statistical evaluations and extrapolations, it results that millions of people should be the bearers of a coronary anomaly (0.2-1.2% of the general population)[9] but most of these, are either asymptomatic or undiagnosed
- Is there a causal relation between a rare event as sudden death and an otherwise uncommon condition such as a coronary anomaly, and can this be demonstrated[10] and if, in how many of the patients? Are preventive measures and therapies justified?
- In what measure these anatomical variations can influence on the pathological process; and do they have a predominant or auxiliary role in the disease process?
- In order to produce unfavorable clinical conditions, are additional factors such as spasm, compression, hypertrophy, dysrhythmias or clotting disorders, required?
- Are these anomalies and their effect, evolving with time in the same patient? Is there any gradual development of symptoms during the natural history of a given anomaly and can a threshold be established? What would be, for example the significance of an atherosclerotic lesion developing on an anomalous coronary branch?
- What are the differences in the management/follow-up of such patients?
- Are the specific coronary variations or anomalies heralded or characterized by any particular clinical sign or symptom?
- Is the coronary anomaly part of a more complex cardiac malformation?
- What should the physician do when recognizing a coronary anomaly? (besides the better-known ALCAPA or coronary fistulas)
- What are the exact clinical consequences of a coronary anomaly and is there a precise timing of application of therapeutic measures? ❑

The physician should concentrate on the possible relationship between anomaly and symptomatology, alteration of the diagnostic tests *and should think first at other possible causes for a given clinical picture.*

Probably, in this respect, the classification of coronary anomalies in "major" and "minor" makes more sense[11], though, some of the so called "minor anomalies" are not "minor" at all, as for example the abnormal origin of the left coronary from the right aortic sinus (1LCxR) which is associated with sudden death or myocardial infarction.

Some of the coronary arterial anomalies are acquired (aneurysms, fistulas, some forms of single coronary artery) and consequently, the term „congenital" should be applied only in selected cases. ❑

Coronary anomalies of clinical and surgical relevance
anomalous pulmonary origins of the coronaries(APOC);
anomalous aortic origins of the coronaries (AAOC);
congenital atresia of the left main (CALM)
coronary aterio-venous fistulas (CAVF);
coronary bridging (myocardial bridging);
coronary aneurysms (CAn);
coronary stenosis.

CLASSIFICATION OF CORONARY ARTERIAL ANOMALIES

Due to the different definition criteria, a universally accepted classification is difficult to be elaborated. A synthetic classification is given in Table 3. ❑

TABLE 3. Synthetic classification of the coronary anomalies[9]

ANOMALOUS PULMONARY ORIGIN[A] OF THE CORONARYARTERIES			
APOC[C]	"Major anomalies" [B]		
	ALCAPA	severe	Origin form Pulmonary sinus: 1, 2 or NF
	ARCAPA	severe, rare	
	ACxPA	severe, rare	
	ARCLCPA	severe, rare	
ANOMALOUS AORTIC ORIGIN[A] OF THE CORONARIES			
AAOC	"Minor anomalies"		
	LMCA from sinus 1 [D]	1/3 of all coronary anomalies	
	RCA from sinus 2 [D]		
	LAD from sinus 1		
	LAD from RCA		
	Cx from sinus 1		
	Cx from RCA		
	Single coronary artery		
	Inverted coronary arteries		
	Other		
CORONARY ARTERIO-VENOUS FISTULAS			
CAVF	"Major anomalies" [B]		
	RCA to RV	congenital / acquired single / multiple associated with: TOF ASD, VSD, PDA Pulm. atresia + intact septum	Angiographic classification: Type A = proximal (proximal dilated, distal normal) Type B = distal (entire length dilated)
	LAD to RA		
	RCA, LAD to LV		
	Cx to PA		
	Diag to CS		
	OM to SVC		
	Single coronary to LA		
INTRAMYOCARDIAL COURSE (MYOCARDIAL BRIDGING)			
Bridging	Cx	Symptomatic or asymptomatic Innocuous or may require surgery [B]	Stenosis at stress test: Group I <50% Group II 50-75% Group III > 75%
	LAD		
	RCA		
	Multiple		
	Other atypical / rare		
CORONARY ANEURYSMS (CAn)[B]			
CAn	Cx and LAD	Type I (diffuse, 2-3 vessels)	88% in males Congenital (types I-IV) Acquired: - atherosclerotic; - Kawasaki, Marfan, Ehlers-Danlos, Takayasu - other systemic diseases, polyarteritis, scleroderma - infectious (incl. syphilis) - traumatic Aneurysm +/- stenosis
Ø > 1.5 x diameter of adjacent normal coronary artery	Cx and RCA		
	LAD and RCA		
	Cx, LAD and RCA		
	Cx and LAD	Type II (diffuse in 1 vessel + localized in other)	
	Cx and RCA		
	LAD and RCA		
	Cx, LAD and RCA		
	Cx	Type III (diffuse in 1 vessel)	
	LAD		
	RCA		
	Cx	Type IV (localized in 1 vessel)	
	LAD		
	RCA		

[A] The formerly used phrase "*anomalous origin*" should be abandoned and replaced with "***anomalous connection***" which reflects better the actual embryological development: the peritruncal vessels eventually connect to the aorta (as an ingrowth and not as an outgrowth from the aorta); [B] Surgical relevance; [C] Origin at the level of a commissure complicates transfer or tunnel repair. High take-off distal to the sinutubular pulmonary junction can be fatal in case of pulmonary banding; [D]Associated with cardiac symptoms and sudden death

PATHOPHYSIOLOGIC CONSEQUENCES AND CLINICAL AND SURGICAL IMPLICATIONS

All of these depend on the type of coronary anomaly and on "the demonstrability" of such. In case of abnormal origin from the pulmonary trunk, or of coronary fistulas, the pathophysiology is more evident and the therapeutic measures follow an algorithmic approach. In other cases, there may not be such a direct causal relationship and indications and timing may differ.

A. Abnormal origin from the pulmonary artery (APOC)

Abnormal origin from the pulmonary trunk or artery may cause: myocardial ischemia (or infarction), mitral insufficiency, congestive heart failure and death in early infancy. The main pathophysiological mechanism is represented by the impoverishement of left ventricular myocardial blood flow due to retrograde flow toward the pulmonary trunk through the intercoronary anastomoses (the surgical creation of a two-artery coronary system is thus mandatory).

The ALCAPA may serve as a paradigm: during the neonatal period high pulmonary vascular resistance and resultant pulmonary artery pressure ensure antegrade flow from the PA to the anomalous coronary artery; as this pressure decreases the flow eventually reverses with resultant left-to-right shunting (into the pulmonary trunk). In face of this coronary steal the myocardial perfusion becomes dependent on the RCA by means of intercoronary anastomoses[12]. The rapidity of this sequence divides patients in two categories: the infantile type and respectively the adult type [13].

The infantile type has few or no collaterals and myocardial ischemia ensues rapidly with all the signs of myocardial ischemic dysfunction present. Infants present with poor feeding probably due to angina, tachypnea, tachycardia and over heart failure. Such clinical findings are however difficult to distinguish from those of cardiomyopathy or endocardial fibroelastosis. Electrocardiographic signs of anterolateral infarction can be present, along with those of left ventricular hypertrophy. Myocardial enzymes can be elevated. Cardiomegaly and interstitial pulmonary edema are present on the chest X-ray. Prompt surgical decision is needed; otherwise premature deterioration [14] and death supervene [15].

The adult type accounts for 10-15% of the patients [16] survival is aided by the presence of large collaterals. Clinical presentation with fatigue, dyspnea, palpitations and effort angina can develop beyond age 20 but in some cases, the patients can still remain asymptomatic with a nonspecific cardiac murmur (apical pansystolic) as a consequence of mitral regurgitation (this latter sign can sometimes dominate the clinical picture). The ECG is abnormal, revealing signs of an old anterolateral infarction. Cardiomegaly may be present. Ejection fraction can be still within normal limits but anterolateral hypokinesia is evident.

Surgery envisages the elimination of the abnormal origin of the coronary and ideally, the restitution of a two-vessels system: reimplantation into the aorta, coronary artery transfer, tunnel operation, subclavian-left coronary artery anastomosis or coronary artery bypass grafting. Ligation of the proximal left coronary artery is accepted only as an interim measure nowadays.

Anomalous connection of RCA, Cx or LAD to the PT is rarely encountered. The anomaly appears to be less lethal although cases related to sudden death have been described.

B. Abnormal aortic origin (AAOC)

Abnormal aortic origin is usually benign except the origin of LMCA from sinus 1 and of the RCA from sinus 2, which can be associated with cardiac symptoms and sudden death[17-21]. The origin of LMCA from sinus 1 with course between the great arteries is associated with the greatest risk of sudden death, even up to 82%[9].

The intramural course or between the great arteries is alleged to produce compression of the abnormal coronary, though not always demonstrated[3,22]. Under these circumstances, intravascular ultrasound or myocardial perfusion scan might represent valuable diagnostic tools.

Stretching of the abnormal left coronary might represent the main pathophysiologic mechanism in systole; during diastole the artery can be compressed by the closely related intercoronary commissure[23].

Other associated lesions and mechanisms, besides compression or stretching of the anomalous coronary artery may supervene in cases with AAOC: single ostium located near a valvar commissure, slit-like aortic orifice, a very oblique origin and proximal tract.

Clinical or ECG features are not characteristic. Angiography can be diagnostic in patients with exertional angina or syncope. When stretching of the coronary artery is the main mechanism, concomitant injection in the anomalous coronary and the PT is of diagnostic value[23].

Surgical solution is represented by coronary artery bypass grafting, reconstruction and reimplantation of the origin of the anomalous coronary trunk, unroofing of the intramural tract or division and reimplantation.

C. The single coronary artery

Definition: only one ostium is present and the coronary artery originating from the single ostium vascularizes the entire heart[1]. It can present with no intrinsic abnormalities of the artery or with associated intrinsic modifications such as: aneurysm or anomalous communication with a cardiac chamber.

The single coronary artery can present under various forms[1,24]

- Type I: "true single coronary": one artery supplies the entire heart ;
- Type II: single artery divides in RCA and LCA (actually 2 coronaries with common aortic origin);
- Type III: other atypical patterns.

This anomaly (per se) is considered "minor". Its pathological significance is related to lesions or disease processes affecting its proximal course that might induce dramatic events. In addition, single coronary artery may be the single "anomaly" or it may be part of the larger picture of complex malformations of the heart (tetralogy of Fallot, DORV, persistent truncus arteriosus, pulmonary atresia with intact septum, TGA, etc.)

D. Congenital atresia of the left main coronary artery (CALM)

This pattern is different from the single coronary in that a single RCA feeds the entire heart but flow in the LAD and Cx is not centrifugal but centripetal (i.e. retrograde). There is no ostium of the left main coronary artery and the proximal LMCA ends blindly. All known anastomoses between the left and right system may be apparent. The Cx and LAD are in normal position. This anomaly offers an example in favor of the theory of ingrowth of the proximal coronary trunks toward the aorta.

There are few cases described in the literature. Clinical consequences depend on the superimposed lesions (e.g. atherosclerotic). An association was found with supravalvular aortic stenosis especially in William's syndrome.

E. Coronary arteriovenous fistulas (CAVF)

A coronary fistula is a direct communication between a coronary artery and the lumen of any of the cardiac chambers, the coronary sinus (or one of its tributary veins), the superior vena cava, the pulmonary artery or veins close to the heart (left heart fistulae are in fact arterio-arterial, arterio-cameral or arterio-systemic).

The picture is protean, depending on the number of fistulas, their origin, their drainage, association with other cardiac pathologies, etiology (congenital or acquired), localization at the level of the coronary artery (i.e. proximal or distal), the status of the myocardium. More than 90% of the fistulae open into the right heart chambers or their connecting vessels. However, the Qp/Qs is seldom larger than 1.8 and the arterial pressure pulse is seldom greatly widened.

Presentation is late in life (occasionally in childhood). Most patients with a continuous murmur, mild cardiomegaly or pulmonary plethora. The most common symptoms are effort dyspnea and fatigue; angina in uncommon, myocardial infarction is rare; some patients are asyptomatic. The apearance of heart failure is related to the duration of fistula(s) and not to the amount of shunting. The ECG may be normal or show signs or ventricular (right or left) overload. If the fistula is large enough, the diagnosis can be made two-dimensional and Doppler echography.

A special distinction must be made in case of pulmonary atresia with intact ventricular septum[25], with "***right ventricular-dependent circulation***". In cases without a connection between a proximal coronary artery and the aorta (or with severe luminal stenosis / occlusion), part or all of the coronary circulation is dependent upon perfusion from the RV. Any maneuver that might obliterate the RV cavity (e.g. thromboexclusion, tricuspid oversewing) or that

decompresses the RV (e.g. RVOT reconstruction), will exacerbate ischemia.

In cases with continuity between the aorta, coronary arteries and RV, a bidirectional flow in the coronaries might exist.

Many patients exhibit ischemia due to a diastolic steal phenomenon. Lowering the RV pressure (as with the use of prostaglandins or the creation of a systemic-to-pulmonary shunt) may worsen the steal phenomenon and exacerbate ischemia.

A distinction must be made between myocardial sinusoids and the ventriculo-coronary arterial connections[26]. The myocardial sinusoids connect first to a capillary bed which is itself continuous with the epicardial coronary arteries. The ventriculo-coronary connections represent direct communications.

Fistulas can be closed from within the cardiac chamber involved, or through the enlarged coronary itself. Ligature or coronary bypass are other techniques indicated. Aneurysmal dilatations of the coronary arteries should be also addressed.

F. Complete transposition of the great arteries (TGA)

Many anatomical variations and anomalies of the coronary arteries are described in the various forms of TGA and these will not be reviewed here[27]. The relative position of the aorta and pulmonary trunk and the coronary disposition, pose important problems in view of the surgical correction.

The "normal" coronary disposition in TGA is: 1LCx 2R (the disposition appears inverted as compared with the disposition in the normal heart). The most frequent anomalies encountered are: 1L 2RCx, 1Cx 2RL, 1R 2LCx, 2LCxR, 2RLCx, 2 CxRL 1RLCx.. These may pose special surgical problems or even contraindicate the switch at arterial level.

In cases with side-by-side transposition of the great vessels, two anomalies are associated with intramural course of the LAD and Cx (1LCx 2R) or of the RCA and LAD (1RL 2Cx). High take-off of the coronary arteries (distal to the sinutubular junction) was also described in TGA.

The origin and course of the sinus node artery is important in view of the atrial switch operations (Mustard or Senning).

G. Congenitally corrected transposition of the great arteries (CC-TGA)

The morphology of the coronary arteries "follows" that of the ventricles, beyond their origin and proximal course[28] in an otherwise complicated and variable spectrum of malformations in which the atrioventricular and ventriculoarterial discordance render the coronary disposition "anomalous". The interest regarding the particular coronary disposition is more than academic nowadays, especially after the advent of the double-switch corrective procedure[29]. The most common major coronary artery variation is the single coronary artery arising from the aortic sinus 1.

H. Tetralogy of Fallot (TOF)

In various series, coronary artery anomalies have been reported in patients with TOF ranging from 2-9%. Of particular importance are:

- those in which a vessel crosses the RVOT: conspicuous conal branch, origin of the LAD from the RCA or from the aortic sinus 1, origin of the LMCA from sinus 1, origin of the Cx from the RCA or aortic sinus 1, origin of the RCA from sinus 2, origin of the LAD from the NF sinus of the pulmonary trunk;
- those in which a coronary artery contributes to pulmonary blood flow (TOF with pulmonary atresia). In very rare cases, the coronary artery may be connected to the pulmonary system being the major or sole source of pulmonary flow[30].

Another detail of surgical relevance in cases of TOF is the clockwise rotation of the aortic sinuses, which may pose additional problems during the surgical act.

I. Myocardial bridges

The incidence at catheterization is 0.5-16% while in the general population, it is estimated at 5.4-85.7%[9].

Myocardial bridging[31] represents usually a benign condition with an excellent long-term survival[32]. On the other hand, its presence has been linked to myocardial ischemia, infarction[33-41], exercise-induced tachycardia[42], conduction disturbances[43,44] and sudden death.[45-48] A careful analysis of the lesion(s) is mandatory as the therapeutic approach is totally different from case to case.

Many forms of bridging have been described[31]; the term itself does not represent a unique entity but a wide spectrum of modifications, and thus, clinical manifestations and consequences are protean. A correlation was attempted between myocardial bridging and ECG, myocardial perfusion scanning and clinical symptomatology[9] but with limited practical significance.

J. Coronary aneurysms (CAn)

Coronary aneurysms are defined as dilatations of a coronary artery 1.5 times more than the adjacent normal coronaries. They can present as saccular or fusiform. Their incidence is 0.3-4.9% in the general population and are more common in males[9]. Besides congenital forms, many are acquired (for example syphilis, Kawasaki disease). The existence of concomitant atherosclerotic lesions may complicate the clinical picture and render the surgical solution more difficult.

Aneurysms can produce compression, thrombosis, embolization, rupture, fistulization. This depends on the location, number, etiology, form (i.e. fusiform, saccular). The therapeutic measures depend on the ethiology, manifestation, complications and the status of the remainder coronary system including the distal vessels. ❑

CONCLUSION

As in the case of other organs in the body, there is a wide spectrum of anatomical presentations regarding the coronary arteries. The transition from "*variation*" toward "*anomaly*" is gradual and precise limits are impossible to be set. Anomalies should however be seen in the context of the heart and of the cardiovascular system: probably pure or singular anomalies do not exist (when single, the term "variation" probably would be more suited).

The significance of an anomaly is difficult to demonstrate; it is difficult to have the whole picture of every single case, starting with the clinical signs and symptoms and ending with the pathologic examination. In this way, our critical approach might refute a lot of improbable connections between the clinical event and the coronary anomaly but in the mean time our thinking and theories should anticipate the facts and prevent complications.

Coronary anomalies represent a good example of the dilemma between "*doing too much*" and "*doing too little*". ❑

Abbreviations

AAOC	Anomalous aortic origin of the coronaries
ACxPA	Abnormal circumflex from the pulmonary artery
ALCAPA	Anomalous left coronary artery from the pulmonary artery
APOC	Anomalous pulmonary origin of the coronaries
ASD	Atrial septal defect
ARCAPA	Anomalous right coronary artery from the pulmonary artery
ARCLCPA	Anomalous origin of both coronaries from the pulmonary artery
CAn	Coronary artery aneurysm
CAVF	Coronary arterio-venous fistula
Cx	Circumflex branch of the left coronary artery
CS	Coronary sinus (the venous collector)
DORV	Double outlet right ventricle
LA	Left atrium
LAD	Left anterior descending (anterior interventricular branch) of the left coronary artery
LMCA	Left main coronary trunk (left coronary artery)
LV	Left ventricle
OM	Obtuse marginal branch (from the left coronary artery)
PA	Pulmonary artery
PDA	Patent ductus arteriosus
RA	Right atrium
RCA	Right coronary artery
RV	Right ventricle
RVOT	Right ventricular outflow tract
SVC	Superior vena cava
TOF	Tetralogy of Fallot

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