My approach to the diagnosis of congenital coronary anomalies by transthoracic echocardiography

Como eu Faço o Diagnóstico das Anomalias Congênitas das Artérias Coronárias pelo Ecocardiograma Transtorácico?

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“Your vision will become clear, only when you can look into your heart. Who looks outside: dreams, who looks inside: awakes” (Carl Jung)

Why do we need to assess the anatomy of the coronary arteries?

Until a few decades ago, coronary artery anatomy assessment by echocardiogram (ECHO) was considered a difficult secondary task even if analyzed by other cardiovascular imaging modalities, such as angiography by cardiac catheterization. Important changes in this approach in recent years can be explained by three important aspects:¹ The origin and initial course of coronary artery anomalies can be a substrate for ischemia and even sudden death in young adults;^{1,2} 2) Transthoracic echocardiography (TTE) is a non-invasive radiation-free low-cost imaging test in the frontline of cardiac assessments;³ 3) The anatomical and functional assessment of coronary arteries is part of a transthoracic test used to diagnose congenital heart diseases.^{3,4}

Which congenital coronary artery anomalies are relevant and should be identified?

The classification of congenital coronary artery anomalies can be extensively sub-categorized by their anatomical and

hemodynamic presentation. Historically, they were simply classified as major versus minor anomalies. Currently, a more comprehensive, broad, and detailed classification covers all anatomical variation possibilities.⁵

The first step is to know the normal pattern (Table 1) from a quantitative and qualitative point of view. Once the normal pattern is excluded, all findings should be considered for the definitive diagnosis and classification (Table 2).

The objective of this review is to focus on anomalies originating from the coronary arteries of the aorta but from a different Valsalva sinus (from opposite sinuses), which have challenged pediatric and adult cardiologists.

Table 1 – Coronary artery normal pattern in humans.

Characteristic	Variation
Number of ostia	
Location	Right and left (anterior)
Proximal orientation	45° at 90° in the aortic wall
Proximal trunk	Left only (anterior descending and circumflex arteries)
Proximal course	Straight from the ostium to the end
Mean course	Extramural (subepicardial)
Branches	Suitable for dependent myocardium
Essential territories	Right coronary (free wall of the right ventricle), anterior descending artery (anterior-septal wall), marginal obtuse (free wall of the left ventricle)
End	Capillary bed

Keywords

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Table 2 - Classification of congenital coronary artery anomalies.

Anomalias de Origem e trajeto
1. Absence of the left coronary artery trunk: separate origin from the AD and Cx
2. Ostial location anomaly within the Valsalva sinuses: <ol style="list-style-type: none"> High origin Low origin Commissural
3. Abnormal ostial location outside the Valsalva sinuses: <ol style="list-style-type: none"> Posterior aortic sinus (non-coronary) Ascending aorta Left ventricle Right ventricle Pulmonary artery: <ol style="list-style-type: none"> LC originating from the posterior sinus Cx originating from the posterior sinus AD originating from the posterior sinus RC originating from the posterior sinus Ectopic origin of the pulmonary artery (outside the posterior sinus of the pulmonary valve): <ol style="list-style-type: none"> Anterior sinus Pulmonary trunk Pulmonary branches Origins of the other arteries (aortic arch, innominate artery, right carotid, mammary, bronchial artery, subclavian)
4. Anomalous origin of different Valsalva sinus coronaries (includes single coronary origin): <ol style="list-style-type: none"> RC originating from the left Valsalva sinus with an anomalous course: <ol style="list-style-type: none"> Posterior course to the atrioventricular junction (retrocardiac) Posterior course to the aorta (retroaortic) Between the aorta and the pulmonary artery (intramural) Intraseptal Anterior to the pulmonary outflow tract AD originating from the right anterior sinus with an anomalous course: <ol style="list-style-type: none"> Between the aorta and the pulmonary artery (intramural) Intraseptal Anterior to the pulmonary outflow tract Posterior to the interventricular septum Cx originating from the right anterior sinus with anomalous course: <ol style="list-style-type: none"> Posterior course to the atrioventricular junction Posterior to the aorta (retroaortic) LC originating from the right anterior sinus with anomalous course: <ol style="list-style-type: none"> Posterior to the atrioventricular junction Posterior to the aorta (retroaortic) Between the aorta and the pulmonary artery Intraseptal Anterior to the pulmonary outflow tract Posterior to the interventricular septum
End anomalies:
1. Coronary fistulas (originating from the RC or LC) for: <ol style="list-style-type: none"> Right ventricle Right atrium Coronary sinus Superior vena cava Pulmonary artery Pulmonary vein Left atrium Left ventricle

How should equipment be adjusted to assess coronary anatomy?

Advanced equipment has favored the diagnosis of structures such as the coronary arteries. Some equipment comes with specific settings for this assessment that adjust automatically. However, some tips or basic rules can be very useful and improve diagnostic accuracy as follows:

Use transducers with high-frequency as often as possible, even in adult patients (pediatric transducers with a frequency > 8 MHZ); focus on the coronary arteries in the field near the sector.

Decrease compression (to enable more distinct identification of the coronary arteries).

Use the long axis/short axis parasternal window possibly one intercostal space upward: rotate it clockwise toward the left coronary artery and counterclockwise toward the right coronary artery, pointing toward the right shoulder. All movements must be very subtle.

Never use still imaging for diagnostic purposes: Due to the size of the arteries and the possibility of false diagnosis, it is important to evaluate them very slowly, usually frame by frame. A real-time assessment is not ideal as an important translation of the coronary arteries occurs during the cardiac cycle.

Color Doppler evaluation is mandatory (with speed adjustments).

What to assess for anomalies originating from the coronary arteries of the opposite sinuses?

Anomalies originating from opposite sinuses occur in around 1% of the general population.¹ One of the great challenges is that most patients remain asymptomatic; when they become symptomatic, most present with a more serious event. Right coronary anomalies originating from the left sinus are more frequent; however, left coronary anomalies originating from the right sinus have the greatest risk for ischemic events. Anatomical risk factors include: An interarterial and intramural course, slit ostium, and acute emergence angle tangent to the vessel wall. All of these possibilities increase the risk for ischemic events, ventricular arrhythmias, and sudden death. Exhausting physical exercise increases the chance of sudden death, making this entity the second cause of sudden death in young athletes after hypertrophic cardiomyopathy.²

Despite controversies and discussions about athlete screening protocols, the echocardiographer should be prepared to make this diagnosis as it is the method of choice for this population. TTE has excellent diagnostic accuracy at detecting the ostium and the proximal course of the coronary arteries.⁶ However, as it is operator-dependent, the expertise of this professional makes this cardiovascular imaging modality the key piece in an algorithm for diagnosing this congenital heart disease.

To assess diagnostic quality and establish guidelines and algorithms, a multicenter study reviewed the vulnerable points of echocardiographers in recognizing the most important anatomical markers.^{3,7}

Interarterial course of the coronary arteries

When emerging from the left sinus, the initial epicardial course of the normal left coronary artery is short (in adults < 4 cm) before dividing into circumflex (Cx) and anterior descending (AD) arteries. The RC originates from the right Valsalva sinus and runs behind the pulmonary artery, below the right atrial appendage, and along the right atrioventricular groove (Figure 1).

There are several alternative courses anomalous according to the origin of the coronary arteries (Figure 2). The anomaly originating from the LC of the right sinus and the RC originating from the left sinus allow an interarterial course (between the aorta and the pulmonary artery). In both, the coronary artery may originate from a single ostium or from separate ostia. (Figure 3).

The echocardiographic image in the short-axis parasternal plane, with the transducer pointed at the patient's left shoulder, delimits the initial course between the two vessels, with a space (echogenic) separating it from the arterial wall (Figure 4). Another very important detail is the ostium position in relation to the commissure. The ideal position would be equidistant (in the middle of the Valsalva sinus), as it would reduce the risk of ostial stenosis and, in cases of surgical treatment, would facilitate the surgical technique. It is important to highlight that the visualization of the coronary commissures and ostia with very subtle movement indirectly informs that there is no high coronary origin (above the sinotubular junction [STJ]).

Intramural course

The second anatomical marker, and perhaps the most important as a predictor of ischemic events, is the proximal RC or LC intramural course originating from the opposite sinus. Unfortunately, it is also the most diagnostically challenging. Histologically, it is defined as sharing of the tunica media by the coronary artery and the aorta (Figure 5). An ECHO diagnosis is made using the same two-dimensional image associated with color Doppler, which is

mandatory (Figures 6 and 7). The first characteristic is the angulation of the proximal portion of the coronary artery at an acute angle (< 30 degrees) and the diastolic flow in the proximal course located "inside" the wall of the aorta (Video 1). One of the most remarkable and specific characteristics is called diastolic "hang-up" (Videos 2 and 3), which means the final diastolic flow in this portion, practically when it would no longer have antegrade flow. In addition to being essential for diagnosis, color Doppler should be explored and, often, subtle changes in the direction of the ultrasound beam can enhance the flow tangent to the aortic wall. Always remember to adjust the speed of the color flow mapping. A pulsed Doppler pattern is also valuable despite technical, angulation, and translation of the heart difficulties. As for anatomical markers, the presence of separated ostia is related to a higher incidence of an intramural course.⁸ An origin very close to the commissure must also be reported, as it would be a non-compliant area contributing to flow restriction.

High origin of the coronary arteries

The coronary ostia follow a normal pattern, which is usually equidistant from the commissures below the STJ. A high origin is identified when the origin is above the STJ (Figure 8). This anomaly alone would not represent risk of ischemic events; however, the possibility of an ostial anomaly or compression, or even of an acute angle at the origin (< 45 degrees) can be associated with high output. Echocardiographically, the standard long-axis parasternal plane requires some modification, with an angle of around 30 degrees toward the patient's right shoulder (Video 4).

What is the role of multimodality cardiovascular imaging in congenital coronary artery anomalies?

In the current era, multimodality is already well established and used precisely to complement and

AD: anterior descending artery; AO: aorta; Cx: circumflex artery; LC: left coronary artery; M: mitral valve; RC: right coronary artery; T: tricuspid valve.

Figure 1 – Normal coronary artery anatomy showing the main branches and epicardial distribution.

AD: anterior descending artery; Cx: circumflex artery; LC: left coronary artery; LCC: left coronary cusp; M: mitral valve; RC: right coronary artery; RCC: right coronary cusp; NCC: non-coronary cusp; T: tricuspid valve.

Figure 2 – Possible coronary artery courses.

LC: Left coronary artery; RC: Right coronary artery.

Figure 3 – Main anomalies originating from the opposite sinuses.

AD: anterior descending artery; Cx: circumflex artery; LC: left coronary artery; RD: right coronary artery.

Figure 4 – Orientation of the ultrasound beam plane used to obtain the echocardiographic image of the coronary arteries. Short-axis echocardiographic image showing the initial course of the left coronary artery and the bifurcation.

Figure 5 – Representation of the interarterial and intramural courses of the left coronary artery originating from the right sinus.

Figure 6 – Orientation of the ultrasound beam plane used to obtain the echocardiographic image of the coronary arteries. Short-axis echocardiographic image showing the initial course of the right coronary artery originating from the left sinus in the “Color compare” mode. The color Doppler image showed the right coronary artery flow practically within the aortic wall: interarterial and intramural course.

Figure 7 – Orientation of the ultrasound beam plane used to obtain the echocardiographic image of the coronary arteries. Short-axis echocardiographic image showing the initial course of the left coronary artery originating from the left sinus in the “Color compare” mode.

Video 1 – Parasternal short axis view, in Color Doppler compare mode: Anomalous origin of the left coronary artery from the right sinus with interarterial and intramural course. There is an angle < 30 degrees of the coronary artery in relationship with aortic wall. There is a late color demonstration of the intramural and interarterial course of the coronary artery (“hang-up”).

Video 2 – Parasternal short axis view with Color Doppler: Anomalous origin of the left coronary artery from the right sinus with interarterial and intramural course. There is an angle < 30 degrees of the coronary artery in relationship with aortic wall, however, there is a shorter course than the case 1. There is a late color demonstration of the intramural and interarterial course of the coronary artery (“hang-up”).

Video 3 – Parasternal short axis view with Color Doppler: Anomalous origin of the right coronary artery from the left sinus with interarterial and intramural course. There is an angle < 30 degrees of the coronary artery in relationship with aortic wall. There is a late color demonstration of the intramural and interarterial course of the coronary artery (“hang-up”). Also, the ostio of the left coronary artery located very close, both close to the commissure between the right and left cusp of the aortic valve.

help establish an assessment and definition algorithm for possible therapeutic decision-making that may even be surgical.³

The role of the pediatric cardiologist and echocardiographer is consolidated within the circle of care even for adult patients, especially young athletes. National and international guidelines are unanimous in establishing the role of these professionals as a pillar for an appropriate diagnosis of these anomalies and adequate conduct, especially for young athletes whose future depends on cardiovascular system integrity.⁹

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Conflict of interest

The author declares no conflicts of interest.

Figure 8 – Possible ectopic locations of the coronary artery ostia (in red) versus the usual location (in blue).

Video 4 – Parasternal short axis view with Color Doppler: High-take off of the right coronary artery above of the sinotubular junction.

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