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STATE-OF-THE-ART REVIEW

Proposed Standardized Nomenclature for Anomalous Aortic Origin of a Coronary Artery

An International Multidisciplinary Initiative

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ABSTRACT

Increasing use of advanced noninvasive imaging has provided great anatomic and morphologic details to characterize congenital coronary artery anomalies. Anomalous aortic origin of a coronary artery (AAOCA) is a subtype that may lead to myocardial ischemia with variable clinical manifestations across the lifespan. Management decision making in AAOCA is hindered by anatomic variants of differing physiologic consequences. Several systems of nomenclature have been used that lack uniformity in potentially high-risk morphologic features. The ICAAC (International Coronary Artery Anomalies Collaborative), a group of experts in coronary artery anomalies, proposes a standardized nomenclature to describe anatomy and morphologic features of AAOCA using the Delphi methodology. This system is agnostic of risk and intended to unify the descriptors of AAOCA within the scientific community. A common language will allow definition of high-risk morphologic features inducing myocardial ischemia while fostering collaborative efforts to improve risk stratification in AAOCA across ages, ultimately guiding evidence-based management decision making. (JACC Cardiovasc Imaging. 2026;■:■-■) © 2026 by the American College of Cardiology Foundation.

Congenital coronary artery anomalies comprise a wide variety of conditions, including anomalies of origin, course, and termination, with some carrying a potential risk of hemodynamic consequences. Anomalous aortic origin of a coronary artery (AAOCA) is a subtype of congenital coronary abnormalities, occurring in 0.2% to 0.7% of the population.[1] It is the second leading cause of sudden cardiac death in young athletes according to autopsy studies.[2,3] Management is complicated by anatomic variants with differing physiologic consequences, ranging from none (incidental finding) to sudden and life-threatening ischemic or arrhythmic events.

Several systems of nomenclature have been proposed and applied to the description of coronary

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

AAOCA = anomalous aortic origin of a coronary artery

ACAOS = anomalous coronary artery from the opposite sinus

CSA = cross-sectional area

CTA = computed tomography angiography

HO = high origin

IM = intramural

LS = left sinus

NS = nonfacing sinus

RS = right sinus

RVOT = right ventricular outflow tract

ST = single trunk

anomalies. Details regarding ostial location and geometry and proximal course are essential elements, as these have implications for treatment and management. Accurate descriptive classification that is accepted by imaging experts, cardiologists, surgeons, and pathologists alike is essential, not just for care of individual patients but also for addressing knowledge gaps in risk stratification and for developing evidence-based recommendations and guidelines for management.

In 1983, Gittenberger-de Groot et al[4] proposed an alphanumeric coronary coding system to describe coronary anatomy in the setting of transposition of the great arteries (the "Leiden Convention").[4] Recently, this was modified to be applicable in cases of normally related great arteries and cases with bicuspid aortic valve.[5] The Leiden Convention is intuitive from the surgical perspective (cranial view of the aortic root) but not from the imaging perspective, which traditionally uses a caudal view of cross-sectional anatomy. Further modifications in 2022 (the "imaging Leiden Convention") incorporated an imaging perspective (caudal rather than cranial perspective), with additional descriptors regarding coronary course.[6] In 1989, Angelini[7] had differentiated what should be considered coronary artery anomalies vs normal variants and classified coronary anomalies as anomalies of origin and course and as anomalies of intrinsic coronary arterial anatomy. His subsequent treatises on coronary anomalies[8,9] emphasized the potential importance of "wrong-sinus origin," postulating that dynamic narrowing of the associated proximal intramural (IM) segment was a key element of ischemic pathophysiology. In 2000, a new system of coronary artery anomaly nomenclature was proposed as part of the IPCCC (International Pediatric and Congenital Cardiac Code)/International Congenital Heart Surgery Nomenclature and Database Project of the Society of Thoracic Surgeons Congenital Heart Surgery Database and the European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Database.[10] This was updated a decade later.[11] In 2021, the IPCCC nomenclature and codes used in these international clinical registries were incorporated as the pediatric and congenital cardiac component of the International Classification of Diseases-11th Revision (ICD-11).[12] A 2016 consensus document from the European Society of Cardiology Working Group on Development, Anatomy and Pathology included a modified classification scheme of coronary artery anomalies based on developmental considerations.[13]

Despite the existence of nomenclature systems for congenital coronary anomalies, there is no standardized way to describe the relevant specific anatomic features in AAOCA that may have implications for risk stratification, clinical decision making, and outcomes, and may advance our knowledge about these anomalies. In addition, the lack of a standardized terminology to describe key reporting elements in AAOCA has led to multiple labels for the same variants, causing confusion in the published reports and seriously hindering the generation of clinical consensus recommendations. In December 2022, an international group of pediatric and adult cardiologists, cardiothoracic surgeons, morphologists, and radiologists with interest and expertise in coronary artery anomalies was convened to form the ICAAC (International Coronary Artery Anomalies Collaborative). This group comprises individuals who have extensive published work in this area and are at the forefront in the evaluation and management of these patients. The first aim of the ICAAC was to propose a new standardized nomenclature system for AAOCA, building on the concepts and strengths of Angelini's classification, the IPCCC, and the concepts introduced by the Leiden Convention, with the goal of creating a system that can be easily understood and applied by all relevant stakeholders. The intent of the proposed nomenclature system is to be inclusive, intuitive, precise, and hierarchical, describing a given patient's coronary anatomy with increasingly specific levels of coding based on the reliability of source information, and without inferring risk. Through a series of multidisciplinary meetings and using a modified Delphi methodology, the ICAAC designed a specific nomenclature system for description of AAOCA, which is described in this document.

METHODOLOGY

This research did not involve human participants or identifiable data; therefore, Institutional Review Board approval was not required.

CREATION OF THE ICAAC. The ICAAC was created as a consortium of internationally renowned medical experts in cardiology, cardiac surgery, imaging, and morphology, with expertise in AAOCA and other coronary anomalies. The goal of the ICAAC was set to develop recommendations for standardized nomenclature, uniform imaging techniques, standardized reporting templates, prediction rules for high-risk

AAOCA, and tailored diagnostic and management algorithms. The first project of the ICAAC, described in this document, was to present a standardized nomenclature for accurately describing the anatomy and morphologic features of patients with AAOCA. Additional work by the ICAAC will focus on imaging techniques, functional assessment, risk stratification, management decision making, and surgical approaches in AAOCA.

The consensus-building process occurred between December 1, 2022, and December 1, 2023. Ten global adult and pediatric experts (7 from the United States, 2 from Italy, and 1 from Switzerland) were invited to participate in the ICAAC. Panelists were selected on the basis of clinical and research expertise related to AAOCA and other coronary anomalies.

PRELIMINARY MEETINGS AND DISCUSSIONS. Preliminary meetings set the stage for this cooperative endeavor, with experts exchanging insights on anomalous coronary arteries, with an initial focus on AAOCA. Monthly virtual meetings facilitated a robust exchange of ideas, showcasing the collective knowledge of the ICAAC. The diverse backgrounds fostered a holistic approach, encompassing both clinical and research-oriented viewpoints. This synthesis of expertise laid the groundwork for a comprehensive understanding of anatomy, diagnostic modalities, and therapeutic interventions for anomalous coronary artery origins. The initial dialogues underscored the need for a standardized anatomic classification system, deemed paramount for effective communication among specialists. The ICAAC also deliberated on research initiatives, including multicenter studies and data-sharing platforms, to accelerate accumulation of knowledge and evidence-based decision making. Risk stratification was not contemplated in this initial work and will be part of future iterations of the work compiled by this collaborative.

MODIFIED DELPHI METHODOLOGY TO CONSENSUS. The RAND-modified Delphi technique[14] has been used for >5 decades to develop indicators that evaluate the appropriateness and quality of a set of metrics. Several modifications of this system[15,16] have been used in developing consensus statements in health care. The advantage of these modifications, which allow careful selection of items/statements at the beginning of the process based on published reports and synthesized reviews and content from experts in the field of interest, provides a solid base for arriving at a consensus usually in 3 rounds of mailings/meetings. These mailings/meetings are designed to reduce bias related to group interactions by allowing anonymous commenting and voting while providing controlled feedback to arrive at consensus.[17-20]

For the AAOCA nomenclature project, expert consensus statements were developed on the basis of the available published reports and expert opinion, and using the modified Delphi methodology, each statement was reviewed and revised through a combination of e-mail and virtual face-to-face discussions. The final revised versions of all statements developed through several virtual meetings were anonymously voted on individually via a cloud-based system, with each expert providing comments detailing the reason for agreement or disagreement on each statement. For validation, >80% voting and >80% agreement was required. The final validated statements (100% agreement achieved at the 5th round) were used to establish a hierarchical system for anatomic description of AAOCA, which will be described in greater detail in subsequent sections. The Supplemental Methodology provides all approved statements and their rationale that were used to finalize the AAOCA nomenclature. Additional minor modifications were made during manuscript review with peer experts.

PROPOSED NOMENCLATURE

The proposed coronary artery anomaly nomenclature system is designed to be a strictly anatomic description and risk agnostic. The nomenclature is constructed in a simple, flexible, and modular manner and can be tailored to be as general or detailed as desired. Therefore, the proposed nomenclature consists of a core descriptor that defines the general type of coronary anomaly, along with additional descriptors that provide specific anatomic and morphologic details (Figure 1).

CORE DESCRIPTOR. The core descriptor includes the involved coronary vessel(s), the type of anomaly (AAOCA), the general location of aortic origin of the anomalous coronary, and the course of the vessel. The core descriptor includes 4 terms (Table 1): 1) the anomalous vessel(s); 2) the term “AAOCA” to indicate the type of coronary anomaly being described; 3) the sinus or location of origin of the anomalous coronary; and 4) the term describing the proximal course of the anomalous vessel. For example, an anomalous right (R) coronary artery arising from the left sinus (LS) of Valsalva and with an IM course would be described as R-AAOCA/LS-IM. The nomenclature is designed to allow the use of individual terms, with “AAOCA”

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FIGURE 1 **Proposed Nomenclature for AAOCA**

Core Descriptor

L	- AAOCA /	RS	-	IM
R		LS		IA
LAD		NS		IS
LCX		HO		PP
ST		LR, LN, RN		RA

Additional Descriptors

1. Ostium characteristics
   a. Location (topography map)
   b. Relationship
   c. Morphology
2. Details of proximal course
3. Coronary dominance
4. Additional findings and comments

The nomenclature includes a core descriptor that summarizes the most important features of the coronary artery anomaly and additional descriptors, which include further details regarding the anatomy. AAOCA = anomalous aortic origin of a coronary artery; HO = high origin; IA = interarterial; IM = intramural; IS = intraseptal; L = left; LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery; LN = left-nonfacing commissure; LR = left/right commissure; LS = left sinus; NS = nonfacing sinus; PP = prepulmonic; R = right; RA = retroaortic; RN = right-nonfacing commissure; RS = right sinus; ST = single trunk.

being the single term required consistently. Notations such as AAOCA, R-AAOCA, AAOCA-IM, and R-AAOCA/LS would all be acceptable, depending on the anatomic information available at any given point in time.

Anomalous vessel. The published reports have used different notations to indicate the anomalous vessel. Examples of these notations include anomalous origin of the left coronary artery (ALCA) or anomalous origin of the right coronary artery (ARCA),[1] and anomalous aortic origin of the left coronary artery (AAOLCA) or anomalous origin of the right coronary artery (AAORCA).[21] Even though this nomenclature is easy to use when referring to anomalies of the left main coronary artery or the right coronary artery, it becomes more difficult when referring to more complex anomalies (eg, anomalous left anterior descending or circumflex coronary arteries, or when several major coronary branches are involved). We therefore recommend stating the anomalous vessel(s) as a prefix before the term "AAOCA":

- R: right coronary artery
- L: left main coronary artery
- LAD: left anterior descending coronary artery
- LCX: left circumflex coronary artery
- ST: single trunk

Terms such as R-AAOCA, L-AAOCA, LAD-AAOCA, and LCX-AAOCA would all be appropriate. If several coronary arteries are anomalous, each prefix can be listed, separated by a comma if arising from the same sinus, for example, right and left coronary arteries from the nonfacing sinus (NS): L,R-AAOCA/NS. If the details of each of the anomalous coronary arteries differ, the corresponding full core descriptor for each vessel can be listed, separated by a semicolon, for example, R-AAOCA/LS; L-AAOCA/NS.

In certain cases, a single coronary artery trunk may arise from the aorta and give rise to all major coronary artery branches. The term "single coronary"[22,23] does not accurately reflect this entity, because anatomically there are 3 separate major coronary arteries (right coronary artery, left anterior descending coronary artery, and circumflex coronary artery) although they all arise from the same trunk.[9] The descriptor "left AAOCA" for a single trunk (ST) from the right sinus (RS) is not accurate, because it assumes that only the left coronary artery is anomalous. However, from embryological and physiological standpoints, the origin of both coronary arteries is anomalous, and the flow dynamics cannot be assumed to be normal in this setting. As such, we recommend the use of the term "ST" AAOCA to define these anomalies, for example, ST-AAOCA/LS or ST-AAOCA/RS.[24]

Type of anomaly. There are 2 different terms that have been mainly used to describe coronary artery/arteries arising anomalously from the aorta: AAOCA[1] and anomalous coronary artery from the opposite sinus (ACAOS).[25] ACAOS describes coronary anomalies in which the abnormal vessel originates from the incorrect and opposite sinus (ie, right coronary artery from the LS and left coronary artery from the RS). This term has been used because of the perception that these types of coronary anomalies carry an increased risk over other types of AAOCA. The term, however, does not allow the description of other types of AAOCA, which may also be associated with increased risk,[26] and may also suggest that all types of AAOCA from the opposite sinus are associated with such a risk. We therefore discourage the use of the term ACAOS and propose the use of the more general term, AAOCA, to describe coronary artery anomalies in which the anomalous coronary artery arises from the aorta but from an abnormal site.

In addition, several authors use the term "cusp" to identify the sinus of origin.[27,28] This term relates to the actual leaflet of the aortic valve and is therefore inappropriate to use.

TABLE 1 Proposed Nomenclature for AAOCA

	Examples
Core descriptor	
1. Anomalous vessel	
R: right coronary artery	R-AAOCA
L: left main coronary artery	L-AAOCA
LAD: left anterior descending coronary artery	LAD-AAOCA
LCX: left circumflex coronary artery	LCX-AAOCA
ST: single trunk (all major coronary arteries)	ST-AAOCA
2. Sinus of origin	
LS: left sinus	R-AAOCA/LS
RS: right sinus	L-AAOCA/RS; ST-AAOCA/RS
NS: nonfacing sinus	L-AAOCA/NS
HO (RS or LS or NS): high origin (≥5 mm or >20%[a] above the sinutubular junction in children where the distance is <5 mm)	R-AAOCA/HO (RS)
LR, LN, RN: left-right, left-nonfacing, right-nonfacing commissures (directly above the commissure but <5 mm or 20%[a] above the sinutubular junction)	R-AAOCA/LR L-AAOCA/LN R-AAOCA/RN
In the setting of bicuspid aortic valve:	
• If 3 sinuses present, use regular description	
• If 2 sinuses present, the respective sinuses should be noted according to the horizontal or vertical raphe	
LS: left sinus	R-AAOCA/LS
RS: right sinus	L-AAOCA/RS
AS: anterior sinus	R-AAOCA/AS
PS: posterior sinus	R-AAOCA/PS
3. Course (state all that apply)	
Interarterial (IA)	AAOCA-IA; L-AAOCA/RS-IA
Intramural (IM)	AAOCA-IM; R-AAOCA/LS-IA,IM; L-AAOCA/NS-IM
Intraseptal (IS)	AAOCA-IS; L-AAOCA/RS-IS
Prepulmonic (PP)	LAD-AAOCA/RS-PP
Retroaortic (RA)	L-AAOCA/NS-RA
Additional descriptors	
1. Ostium characteristics	
a. Location (use a topography map to state circumferential and craniocaudal locations) i. Circumferential location • 1: right sinus • 2: left sinus • 3: nonfacing sinus • a, b, c (location within the sinus) ii. Height location: • Level I: within the sinus and up to the level of the commissures (where the leaflets meet) • Level II: between the level of the commissure and the sinutubular junction (where the sinuses give rise to the tubular ascending aorta) • Level III: from the level of the sinutubular junction up to <5 mm above the sinutubular junction or <20%[a] of the depth of the sinus above the sinutubular junction (within 120% of the depth of the sinus), whichever dimension is smaller • Level IV: ≥5 mm or ≥20%[a] of the depth of the sinus above the sinutubular junction (whichever dimension is smaller)	2a-III, 1b-II, 2c-IV, 3a-I-JC
b. Relationship • Type 1: separate ostia • Type 2: separate but adjacent ostia • Type 3: single ostium with bifurcation inside the aortic wall • Type 4: single ostium and single coronary trunk	Type 1, Type 2, Type 3, Type 4
c. Morphology • Round (major axis = minor axis) • Oval (minor axis is 50%-90% of the major axis) • Slit-like (minor axis is <50% of the major axis) • Pinhole (vertical/horizontal dimensions are equal but smaller than the coronary vessel)	Round, oval, slit-like
2. Details of proximal course	
a. Presence and length of the IM segment	IM (6 mm)
b. Ellipticity (major axis/minor axis)	Ellipticity: 2.1
c. CSA narrowing (%CSA) (percent narrowing based on the CSA of the most narrow segment with respect to the largest segment); include the minimal and distal reference CSA used for the calculation	%CSA narrowing: 55% (minimal CSA 4.02 mm^2, distal reference CSA 9 mm^2)
d. Effective lumen diameter narrowing (%) (percent narrowing of the vessel as calculated on the basis of the effective diameter); specify the details of the effective minimal and distal reference diameters used for the calculation	Effective diameter narrowing: 33% (effective minimal diameter 2.26 mm, effective distal reference diameter 3.38 mm)
e. Acute angle of take-off	Acute angle of take-off: Yes

Continued on the next page

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TABLE 1 Continued

	Examples
3. Dominance	
RD: right dominance	RD
LD: left dominance	LD
CD: codominance	CD
4. Additional findings and comments	
Specify any additional findings such as MB, coronary fistulae, atherosclerotic changes, course through a commissural pillar, intracavitary/intracameral course, distance from the respective sinus in HO, description of "pulmonary-facing" or "RVOT-facing" IA course (if appropriate), other	MB-LAD (2 cm) IA course: pulmonary-facing

[a]The distance from the aortic valve annulus to the sinutubular junction.

AAOCA = anomalous aortic origin of a coronary artery; CSA = cross-sectional area; HO = high origin; JC = juxtacommissural; L = left; LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery; LN = left-nonfacing commissure; LR = left/right commissure; MB = myocardial bridge; NS = non-facing sinus; R = right; RN = right-nonfacing commissure; RVOT = right ventricular outflow tract; ST = single trunk.

Sinus of origin. We suggest that the sinus of origin or location of the ostium of the anomalous coronary artery be stated as a suffix following the term "AAOCA" and separated by a forward slash. Most coronary artery ostia are located within the corresponding sinus of Valsalva or slightly above the corresponding sinus.[24,29] Several studies have shown that normal coronary arteries can arise up to 5 mm above the corresponding sinus of the adult-sized aorta or up to 20% above the distance from the aortic valve annulus to the sinutubular junction in the case of children.[29-32] We therefore suggest stating the particular sinus of origin for coronary arteries that arise from within that sinus or up to 5 mm or 20% of the depth of the sinus above the sinutubular junction, whichever dimension is smaller. We suggest the use of the most proximal (caudal) edge of the ostium to make such a determination. If the anomalous vessel originates from the ascending aorta above this threshold, we suggest the use of high origin (HO) instead of a particular sinus of origin.[30] However, the distance above a specific sinus can be added in "additional comments" and the specific sinus may also be used in parentheses. The use of a topography map (described later) will also help define the specific radial location of the ostium with respect to the sinuses. The sinus of origin or HO can therefore be defined as follows:

- RS: right sinus
- LS: left sinus
- NS: nonfacing or "noncoronary" sinus
- HO (RS or LS or NS): high origin

The term "juxtacommissural" has been variably used in the published reports to denote coronary artery origins adjacent to one of the commissures of the aortic valve.[33,34] The term, however, is not specific enough and can be used variably for origins a few millimeters away from the commissure, above the commissure, or below the commissure. We suggest that vessels that originate close to the commissures still be described as arising from the corresponding sinus of origin, even if they are a few millimeters on one side or the other of the commissure. In the rare instance of an ostium originating directly above or below the commissure (except in cases of HO, as defined earlier), the specific commissure can be used as the location of origin instead. For these rare instances, the following terms can be used for the location of origin:

- LR: left-right juxtacommissural origin
- LN: left-nonfacing juxtacommissural origin
- RN: right-nonfacing juxtacommissural origin

The term juxtacommissural may still be added as an additional descriptor when describing ostial location (see later).

The description of AAOCA in the setting of a bicuspid aortic valve should take into consideration the specific sinus and valve morphology. If 3 aortic sinuses are present with commissural fusion between leaflets, one may use the same nomenclature as for a normal tricuspid aortic valve. In the setting of an aortic root with only 2 sinuses, we suggest dividing the root into the LS and RS, or into an anterior sinus and a posterior sinus, thus specifying the orientation of the commissures in the description of the anatomy.

General proximal course. There are many different courses that each of the 3 major coronary arteries can take proximally as they exit from the aortic root.[1,9] The proximal course of the anomalous coronary is an important feature that helps classify AAOCA and has relevance in diagnostic work-up, risk stratification, counseling, and management. The general proximal course is therefore incorporated in the core description as a suffix following a hyphen.

The different potential proximal courses of anomalous coronaries include the following:

- IM: intramural—The proximal segment travels within the aortic wall for a variable length before exiting the vessel wall and should be used solely in this context of describing the segment of the anomalous coronary artery sharing its wall with the aortic wall before exiting and becoming mediastinal. It is imperative to reserve the term "intramural" solely to describe the course of the anomalous vessel within the aortic wall, which is fundamentally distinct from an intramyocardial course.
- IA: interarterial—The proximal course travels between the aorta and the pulmonary artery.
- IS: intraseptal—The proximal vessel, usually the left main coronary artery or anterior descending coronary artery, enters the interventricular septum below the level of the pulmonary valve, then exits after a distance, and becomes mediastinal lateral to the right ventricular outflow tract (RVOT). There may be contiguous longer intramyocardial segments that extend beyond the conal septum. It is essential not to use the term intraseptal course interchangeably with IM course, as "IM" should be solely used to describe the course of the anomalous vessel within the aortic wall and not within any portion of the ventricular myocardium.
- PP: prepulmonic—The proximal vessel crosses anterior to the RVOT before reaching its destination.
- RA: retroaortic—The vessel travels posterior to the aorta before reaching its destination.

Different proximal course types can coexist. For example, it is not unusual for an anomalous coronary to have both IM and interarterial courses. In such instances, all applicable courses can be stated and separated by commas (eg, L-AAOCA/RS-IM,IA to describe an anomalous left coronary artery from the RS with both IM and interarterial courses), as some coronary anomalies can have an IM course but not an interarterial course (eg, L-AAOCA/NS-IM to describe an anomalous left coronary artery from the NS with an isolated IM course).

ADDITIONAL DESCRIPTORS. Additional descriptors provide important details about the anatomy of the anomalous coronary (Table 1). Whatever details are available can be either spelled out after the core nomenclature or abbreviated as a "one-liner" within brackets after the core nomenclature, with each item separated by commas.

In general, every attempt should be made to cite the additional descriptors, which should include the following items: *Ostium* characteristics, details of the *Course* of the anomalous coronary, coronary *Dominance*, and any *Additional* findings (the acronym OCD-A can be used to remember these details).

Ostium characteristics. Several important items should be included when describing the characteristics of the ostium of the coronary arteries. These features include *Location*, *Relationship*, and *Morphology* (the acronym LoReM can be used to remember these features).

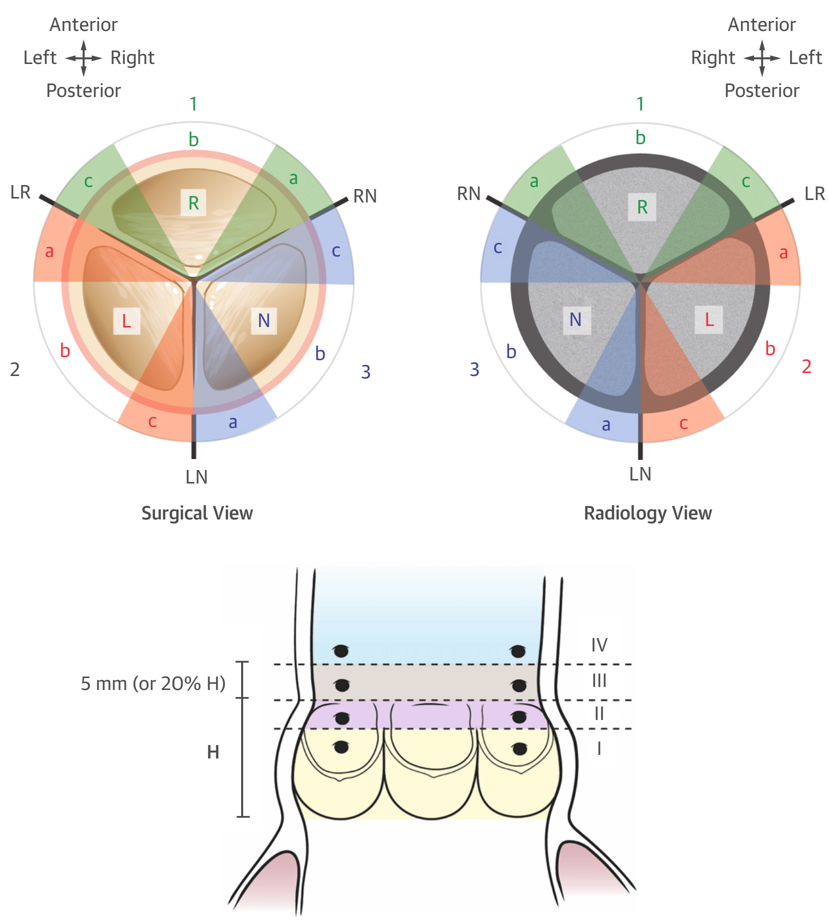
Ostial location. Even though the general location of the ostium (sinus of origin) is already specified in the core descriptor, the specific location of the ostium is critical to fully characterize the anatomy. A standardized method to describe this location, shared by imagers, clinicians, and surgeons, is important to facilitate a common understanding.

We suggest the use of a topography map that includes details regarding the circumferential and craniocaudal locations of the coronary ostium (Figure 2). This topography map has been modified from a previously described map.[35] For the description of the circumferential location, each sinus is given a number (the RS is 1, the LS is 2, and the NS is 3). This nomenclature is consistent with the previously described Leiden Convention of coronary arteries used to characterize the anatomy in transposition of the great arteries[4] and has been used in some recent publications to describe overall coronary anatomy.[5,6] Each sinus is then divided into 3 segments (a, b, and c), with the middle segment (b) spanning the middle 50% of the sinus, and each of the lateral segments (a, c) spanning the respective 25% of the sinus closest to the corresponding commissure. Each commissure is labeled as LR, LN, and RN, as described earlier.

The specific topography in the setting of a bicuspid aortic valve depends on the specific anatomy. If 3 aortic sinuses are present, one should use the regular topography map dividing the root into sinuses 1, 2, and 3. If only 2 aortic sinuses are present, one should use only sinuses 1 and 2. Sinus 1 should be defined as the most anterior sinus and sinus 2 as the most posterior sinus. If the sinuses are completely directed in a leftward/rightward direction with directly anterior and posterior commissures, the right-sided sinus should be defined as sinus 1 and the left-sided sinus as sinus 2. Additional detail may be included in the "Additional findings and comments" section (see later).

The craniocaudal location of the ostium is defined using roman numerals to determine 1 of 4 levels:

FIGURE 2 Topography Map Used to Define the Location of Origin of the Coronary Arteries



The map includes 2 components: a circumferential location reference and a height reference. Both descriptors should be listed, separated by a hyphen. Used with permission from Carlos Mery at The University of Texas at Austin and Ascension (Dell Children's), licensed under Creative Commons Attribution-NonCommercial-ShareAlike 4.0 International (CC BY-NC-SA 4.0). H = height from the depth of the sinus; N = nonfacing; other abbreviations as in Figure 1.

- Level I: Within the sinus and up to the level of the commissures (where the leaflets meet).
- Level II: Between the level of the commissures and the sinutubular junction (where the sinuses give rise to the tubular ascending aorta). The length of this segment is variable from patient to patient and usually corresponds to the area where a thickening of the aortic wall extends from the level of the actual commissures (where the aortic valve leaflets meet) up to the sinutubular junction. This thickening of tissue has been called the "pillar" and is believed to play a potential role in the pathophysiology of AAOCA.[35]
- Level III: From the level of the sinutubular junction up to <5 mm above the sinutubular junction (for adult-sized aortas) or <20% of the depth of the sinus above the sinutubular junction (within 120% of the depth of the sinus), whichever dimension is smaller, as measured to the proximal (caudal) edge of the ostium. This is consistent with previous published reports,[30] which show that most normal coronary arteries in children arise from within the sinus and up to 20% above the sinutubular junction.[29,31,32]
- Level IV: ≥5 mm (for adult-sized aortas) or ≥20% of the depth of the sinus above the sinutubular junction, whichever dimension is smaller. This level would be designated as HO in the core descriptor, as outlined in the "Sinus of origin" section (see earlier).

The circumferential and craniocaudal locations of the coronary ostium are combined (separated by a hyphen) to define the exact location of the ostium. For example, the location of the ostium of an anomalous right coronary artery arising from the LS of Valsalva, just to the left of and above the intercoronary commissure but below the sinutubular junction, would be described as "2a-II." The term "juxtacommissural" (JC) may be added as an additional descriptor for coronary ostia that arise within 1 mm of a commissure on any direction.

Ostial relationship. When there is >1 coronary ostium arising from the same sinus, as is often the case in AAOCA, describing the spatial relationship between ostia becomes relevant. We suggest using a simple nomenclature, originally described by the Congenital Heart Surgeons' Society as part of their data collection forms for the AAOCA Registry,[36] which includes 4 types (**Figure 3**):

- Type 1: completely separate coronary ostia
- Type 2: separate but adjacent coronary ostia
- Type 3: single ostium bifurcating into separate vessels within the aortic wall
- Type 4: single coronary trunk bifurcating into separate vessels in the mediastinum (outside the aortic wall)

Both types 3 and 4 correspond to coronary arteries with a single coronary ostium, but only type 4 corresponds to an ST. As such, coronary anomalies that involve type 3 patterns should be described in the core nomenclature as anomalies of the corresponding vessel (eg, L-AAOCA) whereas type 4 patterns should instead be considered "ST" anomalies (ST-AAOCA), as described earlier.

Ostial morphology. The anatomic morphology of the ostium may play a significant role in the hemodynamic consequences of AAOCA.[37,38] As such, describing its anatomy becomes critical. We suggest the use of a simple system based on the relationship between the major and minor axes of the ostium to define its morphology. This nomenclature, which is slightly modified from a previously described system,[39] uses major and minor axes instead of height (or superior or inferior diameter) and width (or anteroposterior or lateral diameter) because some coronary ostia may be oriented differently and the major axis may not always correspond to the superior or inferior diameter. Acknowledging the difficulty of accurately measuring ostial dimensions, 4 suggested descriptors of the ostial morphology include the following (**Figure 4**):

- Round: minor axis is estimated to be approximately equal to the major axis
- Oval: minor axis is approximately 50% to 90% of the major axis
- Slit-like: minor axis is approximately <50% of the major axis
- Hypoplastic: vertical/horizontal dimensions are equal but smaller than the distal coronary vessel

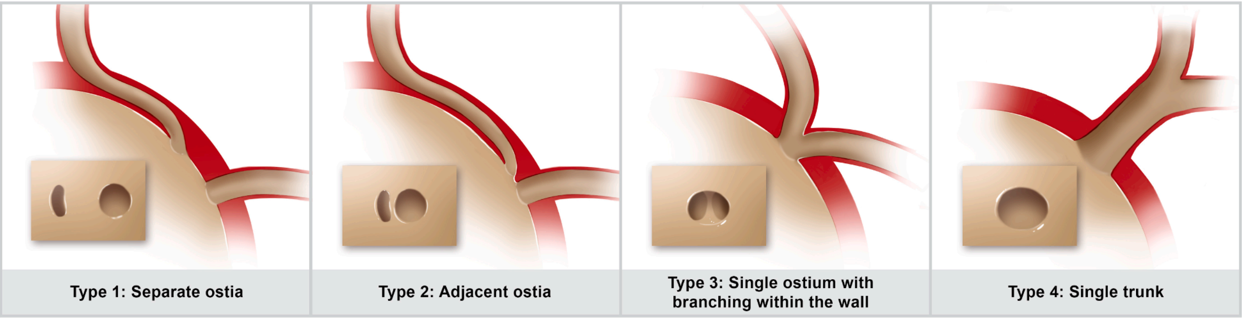
In cases where there is a single coronary ostium (types 3 and 4, as described earlier), a single ostial morphology should be described.

The previously used term "stenotic ostium"[40] does not provide enough granularity and has an implication of risk, to which this nomenclature system remains agnostic. In general, we suggest the use of more descriptive and specific terms to define narrowing of the ostium or proximal course as advanced imaging allows. We encourage the use of specific dimensions as additional descriptors of the ostium, if feasible.

Details of proximal course. The generalities of the proximal course are included as part of the core descriptor. Additional details, however, become important in describing an anomalous coronary. The 5 additional anatomic details that we encourage including, if available, are: 1) the presence and length of intramurality; 2) the maximum ellipticity of the proximal course; 3) the percentage of cross-sectional area (CSA) narrowing; 4) the effective lumen diameter narrowing; and 5) the presence or absence of an acute angle of takeoff.

Presence and length of intramurality. The presence and length of intramurality are considered a potentially significant factor in the pathophysiology and decision making in AAOCA.[38,41] Intramurality can be defined with certain accuracy using cross-sectional imaging or during direct surgical inspection.[40] On computed tomography angiography (CTA), 2 different signs can be used: 1) the presence of an oval shape of the proximal coronary course (related to the ellipticity, described later); and 2) a difference in pericoronary tissue density between the IM segment and the mediastinal segment of the proximal coronary (the "pericoronary fat sign").[40] Although good correlation between radiologic accuracy of IM segment determination identified at surgery has been demonstrated,[40] surgery remains the gold standard to precisely define the IM length of the anomalous vessel. Intramurality is included as part of the main descriptor (eg, L-AAOCA/RS-IM to define an anomalous left coronary artery from the RS with an IM course). In addition, the presence (yes/no) and length (in millimeters) of intramurality should be described as part of the additional descriptors.

FIGURE 3 Relationship Between the Different Coronary Ostia Arising From the Same Sinus



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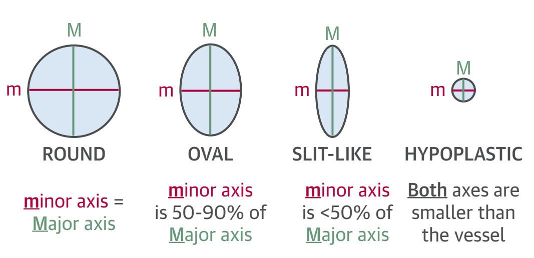
Ellipticity of the proximal course. Ellipticity or "ellipticity index" has been used to describe the lateral deformation of the proximal course of the anomalous coronary, especially in the setting of an IM segment.[37] Ellipticity is calculated using cross-sectional imaging by dividing the major axis of the proximal vessel by the minor axis of the vessel at the same level. This should be calculated at the location where the vessel is most deformed (most oval). The minor and major axes of the vessel should be measured at this level and used to calculate the ellipticity. A normal round vessel would have an ellipticity of 1. An oval proximal segment measuring 3 × 1.5 mm would have an ellipticity of 2. The larger the ellipticity, the more oval the vessel. We encourage describing not only the ellipticity but the actual dimensions of each axis used to calculate the ellipticity.

FIGURE 4 Morphology of the Coronary Ostium as Defined by the Relationship Between its Minor and Major Axes



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CSA narrowing (%CSA narrowing). Measurements of the degree of maximal narrowing of the proximal segment may be useful in helping with risk stratification and for future initiatives to determine the significance of narrowing in life-threatening events in AAOCA. This area of narrowing may be related to an IM segment or to a proximal non-IM segment. Consistent with current standards,[42] we suggest reporting 2 different measurements: CSA narrowing (%CSA narrowing) and effective lumen diameter narrowing.

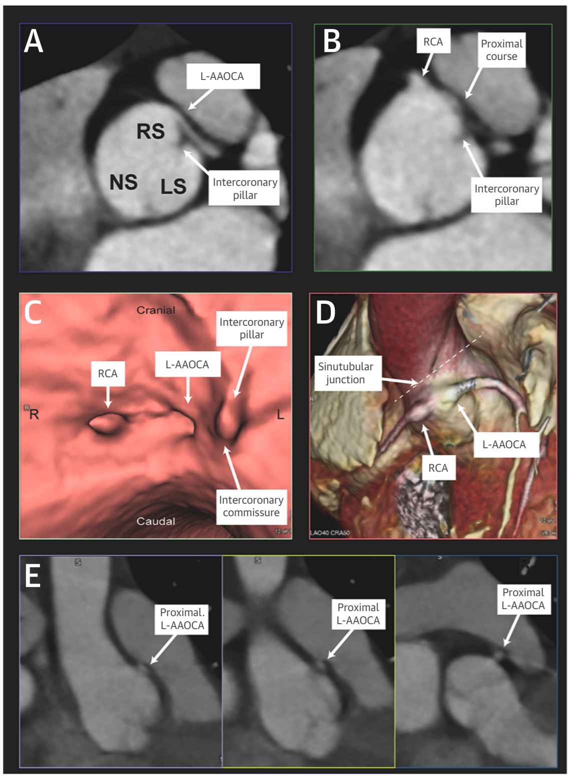
To calculate the CSA narrowing (%CSA narrowing), one should measure the area in square millimeters of the vessel at its narrowest segment (minimal CSA)[42] and the CSA of the vessel at a normal segment of the proximal coronary artery (distal reference CSA).[9,25] The CSA narrowing (%CSA narrowing) is calculated using the following formula:

$$\%\text{CSA narrowing} = \left(1 - \frac{\text{Minimal CSA (mm}^2)}{\text{Distal reference CSA (mm}^2)}\right)$$

As an example, if the calculated CSA by CTA of the narrowest proximal segment of an anomalous coronary is 2.3 mm^2 (which would correspond to the area of a 2 × 1.5-mm oval-shaped vessel) and the CSA of its distal round mediastinal segment is 7.1 mm^2 (which would correspond to the area of a 3-mm round vessel), the %CSA narrowing would be 68% (1 – [2.3/7.1]). Reporting the actual segmental measurements (CSA) is encouraged.

Effective lumen diameter narrowing (%). Percent lumen diameter narrowing is a measurement commonly used to describe atherosclerotic narrowing of coronary arteries.[42] To be consistent with current standards and taking into account the ellipticity of many of the anomalous vessels, we

FIGURE 5 Illustrative Example of a Structured Reporting Template for a Patient With IA L-AAOCA



1. Type of anomalous coronary: L-AAOCA
2. Sinus of origin: RS
3. Proximal course: IM,IA
4. Ostial location: 1b-II
   - Sinus of origin: 1 (right sinus of Valsalva)
   - Circumferential location: b (within ~50% of center of the sinus)
   - Craniocaudal location: II (below the sinutubular junction)
5. Ostial relationship: Type 1 (separate coronary ostia)
6. Ostial morphology: Slit-like
7. Intramurality: Yes, length: 8.5 mm
8. Ellipticity of proximal course: 2.3 mm (major axis: 3 mm × minor axis: 1.3 mm)
9. % Cross-sectional area (CSA) narrowing: 70% (minimal CSA 2.1 mm^2, distal reference CSA 7 mm^2)
10. Effective lumen diameter narrowing: 45% (effective minimal diameter 1.63 mm, effective distal reference diameter 2.99 mm)
11. Acute angle of take-off: Yes
12. Coronary dominance: Right
13. Additional findings and comments:
    - Course behind intercoronary pillar: Yes
    - Dynamic change across cardiac cycle: Yes
      - Ellipticity in diastole: 2
      - Ellipticity in systole: 2.3
    - Myocardial bridge: No
    - Coronary fistulae: No
    - Coronary atherosclerotic lesions: No
    - Interarterial segment: Pulmonary-facing
    - Other comments: None

(Left) Images from computed tomography angiography. (A) Anomalous origin of the left coronary artery from the RS. The proximal course travels behind the intercoronary pillar. (B) Narrowing of the proximal IM course,[40] traveling behind the intercoronary pillar. The RCA arises normally from the RS. (C) Virtual angioscopy view showing separate ostia for the anomalous left and normal right coronary arteries. The anomalous vessel arises from a craniocaudal plane between the actual intercoronary commissure of the aortic valve and the end of the intercoronary pillar (ie, level II). (D) 3-dimensional volume rendering of the aortic root showing the origin of the anomalous left and normal right coronary arteries from the RS of Valsalva. The dashed line indicates the sinutubular junction. (E) Cross-sectional cuts of the proximal course of the left coronary artery as it travels into the mediastinum. The vessel starts as a very elliptical vessel in its IM segment (left). The ellipticity of the vessel progressively decreases (middle) and eventually becomes completely extramural and round (right). L-AAOCA = anomalous aortic origin of the left coronary artery; RCA = right coronary artery; other abbreviations as in Figure 1.

recommend describing the *effective* lumen diameter narrowing of the proximal segment with respect to the distal (reference) coronary artery segment. Describing this parameter from cross-sectional imaging requires converting the minimal and reference CSA of the vessel to *effective* diameters (ie, the diameter of a circle with the same CSA as the measured lumen) and then calculating the percent lumen diameter narrowing. The effective lumen diameter can be derived from the CSA using the following formula:

$$\text{Effective lumen diameter (mm)} = 2 \times \sqrt{\frac{\text{CSA (mm}^2)}{\pi}}$$

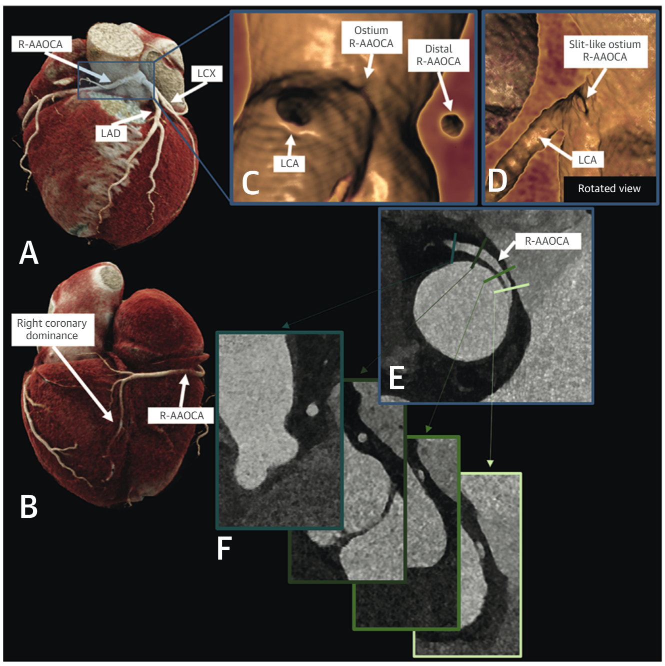
For example, if the minimal CSA of the vessel is 2.3 mm^2, the effective minimal diameter would be 1.71 mm. Similarly, if the distal reference CSA is 7.1 mm^2, the effective distal reference diameter would be 3 mm. The effective diameter narrowing (in percentage) can then be calculated using the following formula:

$$\text{Effective lumen diameter narrowing (\%)} = \left(1 - \frac{\text{Minimal effective diameter}}{\text{Distal reference effective diameter}}\right)$$

For the abovementioned example, the effective lumen diameter narrowing of the vessel would be 43% (1 − [1.71/3]).

Acute angle of takeoff. An “acute angle of takeoff” has been used to describe an angle of the anomalous coronary artery tangential to the aortic wall of <45°.[1] The angle of origin of the anomalous

FIGURE 6 Illustrative Example of a Structured Reporting Template for a Patient With IA R-AAOCA



1. Type of anomalous coronary: R-AAOCA
2. Sinus of origin: LS
3. Proximal course: IM, IA
4. Ostial location: 2a-II
   - Sinus of origin: 2 (left sinus of Valsalva)
   - Circumferential location: a (within ~25% of the sinus closer to the commissure)
   - Craniocaudal location: II (just below the sinutubular junction)
5. Ostial relationship: Type 2 (adjacent coronary ostia)
6. Ostial morphology: Slit-like
7. Intramurality: Yes, length: 8 mm
8. Ellipticity of proximal course: 2
   (major axis: 3.2 mm × minor axis: 1.6 mm)
9. % Cross-sectional area (CSA): 55% (minimal CSA 4.02 mm^2, distal reference CSA 9 mm^2)
10. Effective lumen diameter narrowing: 33% (effective minimal diameter 2.26 mm, effective distal reference diameter 3.38 mm)
11. Acute angle of take-off: Yes
12. Coronary dominance: Right
13. Additional findings and comments:
   - Course behind intercoronary pillar: Yes
   - Dynamic change across cardiac cycle: Not assessed
   - Myocardial bridge: No
   - Coronary fistulae: No
   - Coronary atherosclerotic lesions: No
   - Interarterial course: RVOT-facing
   - Other comments: None

(A, B) 3-dimensional volume-rendered computed tomography angiography with an R-AAOCA from the LS and right coronary dominance. (C, D) Virtual angioscopy views with the slit-like ostium of the right coronary artery and mediastinal (distal) normal coronary vessel shape. (E) Multiplanar reconstruction with an axial view of the aorta showing the acute angle of takeoff of the R-AAOCA. (F) Progression of the vessel with the proximal oval-shaped IM course[40] with proximal narrowing (light green) and the normal round-shaped mediastinal extramural distal vessel (dark green). LCA = left main coronary artery; R-AAOCA = anomalous aortic origin of the right coronary artery; RVOT = right ventricular outflow tract; other abbreviations as in Figure 1.

coronary may be reported as an additional descriptor.

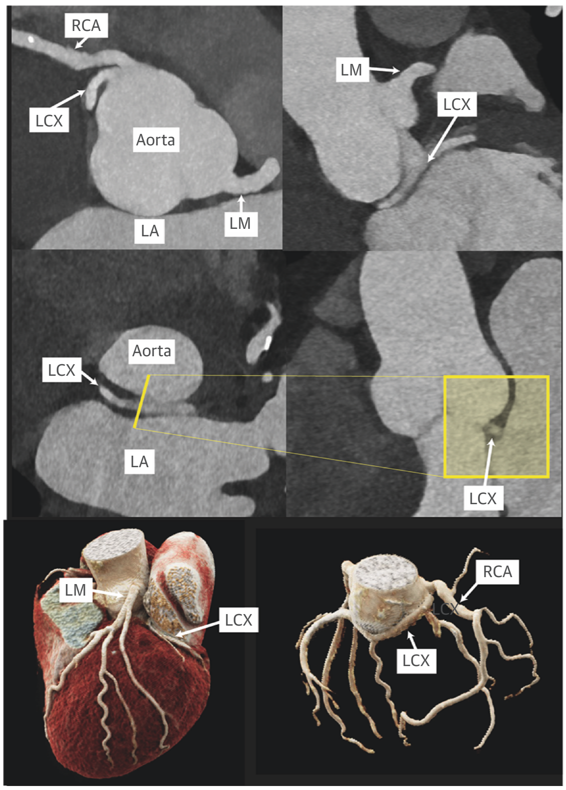
Coronary dominance. Even though it is unclear whether coronary dominance plays a role in the pathophysiology of AAOCA, we encourage reporting whether the coronary artery system is right dominant, left dominant, or codominant. Coronary dominance is defined on the basis of the major coronary artery supplying the posterior interventricular branch (posterior descending artery) and posterolateral branches.[43] In a right-dominant system, the posterior interventricular branch is a terminal branch of the right coronary artery, whereas in a left-dominant system, the vessel is a terminal branch of the left circumflex coronary artery. Approximately 84% to 88% of people have a right-dominant system, 8% have a left-dominant system, and 3% to 8% have a codominant system.[43,44]

Additional findings and comments. AAOCA may coexist with other coronary artery pathology. Description of these findings, if present, is encouraged. Potential additional findings can include proximal vessel crossing behind an aortic valve pillar, dynamic narrowing of the coronary ostium across the cardiac cycle, presence of myocardial bridges, coronary artery fistulae, coronary atherosclerotic lesion, and interarterial segment description, among others. The pillar refers to a thickening of the aortic wall that extends cranially from each of the commissures up to the sinutubular junction and is believed to potentially play a role in coronary compression in AAOCA.[35]

This section is also an opportunity for clinicians to add to the description whatever elements they think are relevant, even if not included as part of the formal nomenclature.

Functionally significant myocardial bridges have been reported in patients with AAOCA.[45-47] In the presence of a myocardial bridge, we suggest reporting the vessel involved, the location of the bridge, the length of the bridge, and the maximum depth of the coronary artery from the epicardial surface.

FIGURE 7 Illustrative Example of a Structured Reporting Template for a Patient With LCX-AAOCA



1. Type of anomalous coronary: LCX-AAOCA
2. Sinus of origin: RS
3. Proximal course: RA
4. Ostial location: 1b-I
   - Sinus of origin: 1 (right sinus of Valsalva)
   - Circumferential location: b (within ~50% of center of the sinus)
   - Craniocaudal location: I (within the sinus)
5. Ostial relationship: Type 2 (adjacent coronary ostia)
6. Ostial morphology: Round
7. Intramurality: No
8. Ellipticity of proximal course: 1 (major axis: 3.6 mm x minor axis: 3.6 mm)
9. % Cross-sectional area (CSA) narrowing: no narrowing (minimal CSA 10 mm^2, distal reference CSA 10 mm^2)
10. Effective lumen diameter narrowing: no narrowing (effective minimal diameter 3.6 mm, effective distal reference diameter 3.6 mm)
11. Acute angle of take-off: No
12. Coronary dominance: Co-dominance
13. Additional findings and comments:
    - Course behind intercoronary pillar: No
    - Dynamic change across cardiac cycle: Not assessed
    - Myocardial bridge: No
    - Coronary fistulae: No
    - Coronary atherosclerotic lesions: Yes, calcifications without stenosis in multiple vessels
    - Interaterial segment: RVOT-facing
    - Other comments: None

The patient is a 52-year-old asymptomatic patient with hypertensive cardiomyopathy, normal left ventricular ejection fraction, left ventricular hypertrophy, and cardiovascular risk factors including dyslipidemia, positive family history for coronary artery disease, and statin intolerance. Coronary computed tomography angiography was performed to rule out obstructive coronary artery disease. Images demonstrate nonstenotic calcifications in the coronary arteries, with an Agatston calcium score of 126.6, corresponding to the 75th to 90th percentile for age and sex. Coronary artery anatomy revealed an anomalous origin of the LCX arising from the RS with an RA course: LCX-AAOCA/RS-RA. Multiplanar reformats (left) and 3-dimensional volume-rendered images (right) demonstrate the RA course of the LCX (red arrows). The ostium appears round (ie, not slit-like), and there was no evidence of an intramural course. The vessel followed an RA course. LA = left atrium; LCX-AAOCA = anomalous aortic origin of the left circumflex coronary artery; LM = left main; other abbreviations as in Figures 1, 5, and 6.

For patients with coronary artery fistulae, we suggest reporting the overall characteristics of the fistulae, such as the vessel of origin, the location and size of the fistulae, and the termination chamber(s).

Coronary atherosclerotic lesions should be described using accepted methods.[42] The description of acquired coronary atherosclerotic lesions is beyond the scope of this nomenclature system.

A recent study has suggested the use of the terms “high” and “low” to describe whether the interarterial segment courses between the aorta and the pulmonary artery or between the aorta and the RVOT, respectively.[48] Even though describing this morphology may be important, we find the use of these terms potentially confusing in the setting of vessels with anomalous origins. As such, we recommend the interarterial course be described as “pulmonary-facing” if it courses between the aorta and the pulmonary artery and “RVOT-facing” if it courses between the aorta and the RVOT.

PROPOSED TEMPLATE FOR IMAGING REPORTING

The work-up of patients with AAOCA generally includes a series of different imaging studies. A standardized approach to reporting the anatomic features of AAOCA from these imaging studies is necessary. Krishnamurthy et al[40] have previously created a structured reporting template for AAOCA, which can be used for CTA or cardiac magnetic resonance and, with minimal modification, for echocardiography, catheter angiography, surgery, and pathology. We

CENTRAL ILLUSTRATION **Reference Guide to Describe the Nomenclature of Anomalous Aortic Origin of a Coronary Artery and Associated Morphologic Descriptors**

Standardized Reporting Template for AAOCA for Imaging and Surgery

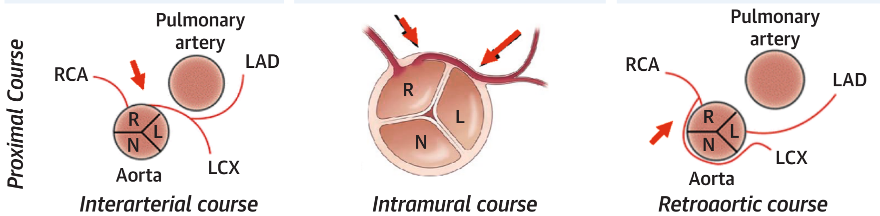
Core Descriptors

Anomalous Vessel–AAOCA/Sinus of origin–Course

1. Type of Anomaly
- R-AAOCA
- L-AAOCA
- LAD-AAOCA
- LCX-AAOCA
- ST

2. Sinus of Origin
- RS (right sinus)
- LS (left sinus)
- NS (nonfacing sinus)
- HO (high origin)

3. Proximal Course
- IA (Interarterial)
- IM (Intramural)
- IS (Intraseptal)
- PP (Prepulmonic)
- RA (Retroaortic)



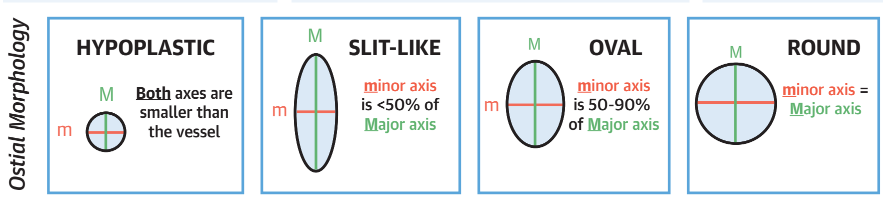
Ostial Characteristics

4. Ostial Location
- a, b, c, LR, LN, RN (radial location)
- I, II, III, IV (vertical location)

See back page

5. Ostial Relationship
- 1 (separate coronary ostia)
- 2 (adjacent coronary ostia)
- 3 (single coronary ostium branching within the wall)
- 4 (single coronary ostium branching outside the wall)

6. Ostial Morphology
- Round
- Oval
- Slit-like
- Hypoplastic

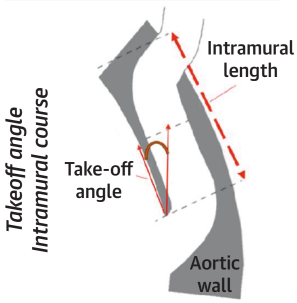


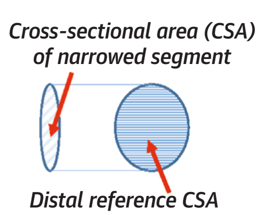
Proximal Course

7. Length of Intramural Course (mm)
8. Ellipticity
9. Percentage CSA narrowing

10. Effective Lumen Diameter Narrowing

10. Acute angle Takeoff (°)





12. Coronary dominance
- Right
- Left
- Codominant

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Continued on the next page

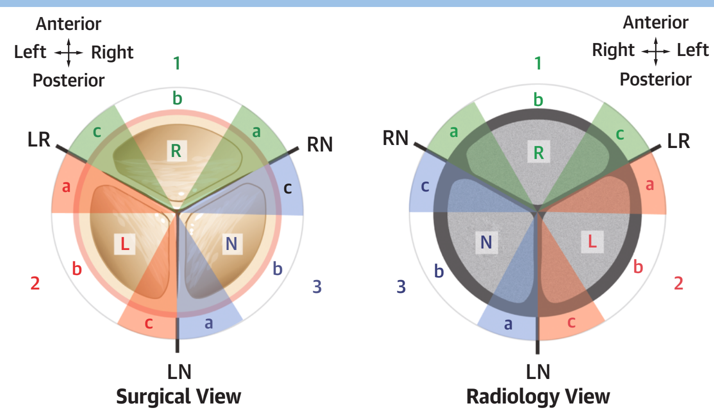
CENTRAL ILLUSTRATION Continued

Additional Findings and Comments

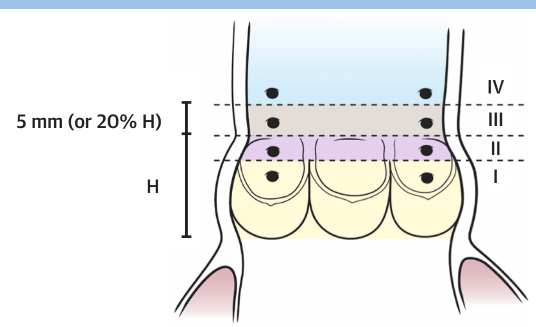
13. Additional Findings and Comments:
- Course behind the intercoronary pillar
- Dynamic narrowing across cardiac cycle
- Presence of myocardial bridge
- Coronary fistulae
- Coronary atherosclerotic lesion
- Interarterial segment description
- Other comments

Detailed Description of Ostial Anatomy in AAOCA

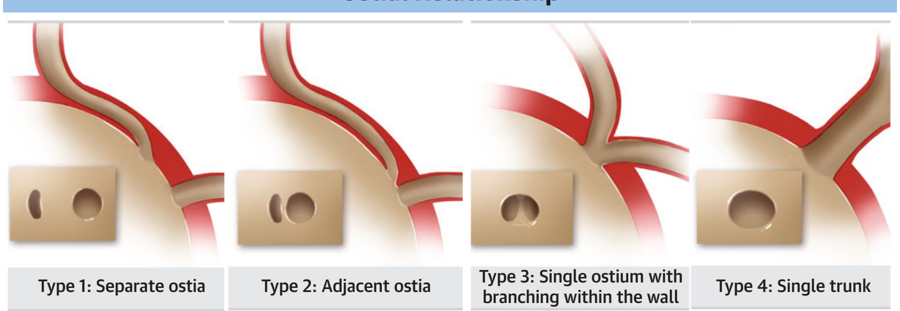
Sinus of Origin and Ostial Radial Location



Ostial Vertical Location



Ostial Relationship



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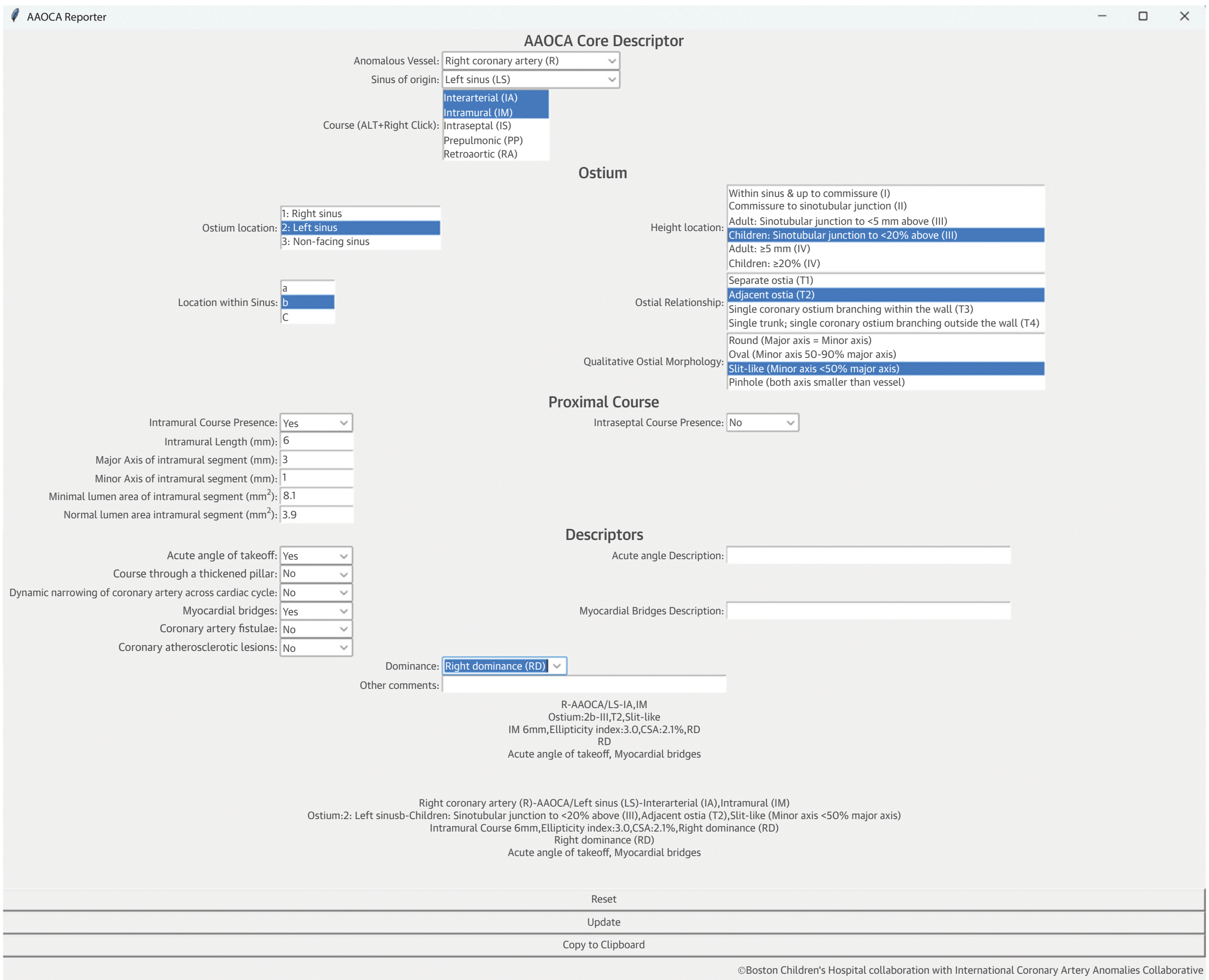
Mery CM, et al. JACC Cardiovasc Imaging. 2026;■(■):■-■.

AAOCA = anomalous aortic origin of a coronary artery; CSA = cross-sectional area; H = height from the depth of the sinus; HO = high origin; IA = interarterial; IM = intramural; IS = intraseptal; L = left; L-AAOCA = anomalous aortic origin of the left coronary artery; LAD = left anterior descending coronary artery; LAD-AAOCA = anomalous aortic origin of the left anterior descending coronary artery; LCX = left circumflex coronary artery; LCX-AAOCA = anomalous aortic origin of the left circumflex coronary artery; LN = left-nonfacing commissure; LR = left/right commissure; LS = left sinus; M = major; m = minor; N = nonfacing; NS = nonfacing sinus; PP = prepulmonic; R = right; R-AAOCA = anomalous aortic origin of the right coronary artery; RA = retroaortic; RCA = right coronary artery; RN = right-nonfacing commissure; RS = right sinus; ST = single trunk.

FIGURE 8 **Example of a Report Crafted on the Online Reporting Tool for Automatic Generation of Report on AAOCA According to the Proposed Nomenclature**



Abbreviation as in Figure 1.

propose a modification of this template that contains coded numerical variables described in this consensus document for the type of AAOCA and includes structured coded information regarding the anomalous coronary, ostial characteristics, proximal course, and dominance. Examples of the use of this proposed template can be found in Figure 5 (L-AAOCA) and Figure 6 (R-AAOCA). Figure 7 depicts an example of LCX-AAOCA. In addition, a quick reference guide has been created that can be digitized or printed for imagers to review upon compiling a report, or nonimagers to refer to if desired, which contains relevant illustrations and nomenclature (Central Illustration).

DEVELOPMENT OF AN ONLINE REPORTING TOOL

To facilitate the deployment of the nomenclature system, a Python application (Python Software Foundation) using Tkinter and Python Imaging Library (Pillow/PIL) interfaces was developed to create summary reports for anomalous coronary artery descriptions. The application uses drop-down menus, multiselect list boxes, text boxes, result mapping, ifelif conditions, and image function definitions to facilitate proper selection of criteria and development of the AAOCA report. The developed Python code was then converted to code for R to enable embedding in a cloud-based website, with a click-through disclaimer before access to the reporter. The end user can input the required information in the various subcomponents of the online reporting tool and will be provided with a composite report that can then be saved as a text document or cut and pasted into the medical record. No identifiable information will be input into the online reporter, and the online reporter can be reset at the end of each session to create a new blank template. This tool will allow automatic generation of the report according to the proposed nomenclature. An example of such a reporting tool is shown in **Figure 8**.[49]

STUDY LIMITATIONS. The proposed AAOCA nomenclature has limitations that need to be acknowledged. The area of expertise of the members of the ICAAC may have introduced bias to the consensus reached using the Delphi methodology. There is a wide variability in the published reports regarding the terminology and definitions used to describe the different features of AAOCA. In general, we tried to use the most commonly used terms and definitions as long as they can be defined in a specific way and remain flexible to allow description of all potential anomalies. We also avoided the use of any terms that carried a connotation of risk. As a new nomenclature system, it may take some time for it to be adopted and widely used. The development of 2 additional tools—the quick reference guide for review of morphologic descriptors (**Central Illustration**) and the online reporting tool for automatic generation of the report according to the proposed nomenclature (**Figure 8**)—should allow ease of adoption over time.

We used consensus to reach an agreement on which terms to use and how to define each of those terms. Several terms that are currently in use were not included as part of the nomenclature system, such as "stenotic ostium," as they are difficult to quantify and define in a standardized way and infer risk, which this document is agnostic of. Similarly, several anatomic features, such as length of interarteriality and length of coronary artery hypoplasia, were not included in the nomenclature system because of the difficulty in defining them using current imaging techniques, the lack of consensus on how to define them, and the subjective nature of describing them.

We also acknowledge that several descriptors may require the use of advanced imaging, such as CTA, to completely define them. The nomenclature was purposefully created in a modular fashion to allow description of whatever items can be described by the available methodology. As such, terms such as L-AAOCA/RS are appropriate on their own without additional descriptors.

Despite the stated limitations, the proposed nomenclature system represents a starting point to create a common and standardized language to be used by clinicians and researchers working with patients with AAOCA. We believe the current proposal should not be viewed as a static document but rather as an evolving system, with the understanding that the principles used herein will evolve as new data and imaging techniques become available in the future. Future efforts of the ICAAC will include specific ways to define the terms used in this nomenclature system for different imaging modalities, surgical inspection, and pathologic examination. Standardizing the methodology for the assessment and description of these anatomy and morphologic features will contribute to future efforts to correlate them with the risk of ischemic events. Furthermore, this could be the impetus for organizations with an established multicenter registry of AAOCA to adopt the proposed standardized nomenclature for use in structured reporting, thus contributing to its validation. It is also conceivable that over time a harmonized, overarching international multicenter registry would be compiled using the proposed nomenclature. It is our hope that the ICAAC will expand its structure and possibly foster this initiative. Finally, efforts should be made to adopt a more detailed description of AAOCA using this nomenclature as part of future iterations of ICD-11 codes and the IPCCC. As mentioned earlier, this proposed nomenclature is not meant to be static, but rather to evolve to reflect the most up-to-date understanding of these congenital coronary anomalies, and working with these organizations is crucial to the process.

CONCLUSIONS

There are many anatomic nuances in AAOCA that are likely associated with increased clinical risk, but these high-risk anatomic characteristics are still incompletely understood. Over the past few decades, many different terms have been used to describe the anatomy and morphologic features in AAOCA with no standardized definitions or usage. The creation of a

HIGHLIGHTS

- There has been no standardized nomenclature to describe the relevant anatomic features of AAOCA, the second most common cause of sudden cardiac death in young athletes.
- The ICAAC, a group of international experts in coronary artery anomalies, developed a proposed standardized nomenclature to describe anatomy and morphologic features of AAOCA using a modified Delphi methodology.
- The described nomenclature system is agnostic of risk and intended to unify the descriptors of AAOCA within the clinical and scientific community.

standardized, simple, hierarchical, modular, and comprehensive nomenclature that is agnostic to risk is paramount to allow clinicians to speak a common language to describe the anatomy of AAOCA. In so doing, clinicians will accurately capture the anatomic subtleties that will allow future initiatives to determine which anatomic characteristics are associated with increased risk. Moreover, likely physiologic factors influence the anatomic substrate leading to a sudden cardiac event and, ultimately, risk stratification in patients with AAOCA will likely include the combination of both. Accuracy and uniform determination of the anatomic/morphologic substrate will enable this ultimate approach to lead evidence-based management decision making. Lastly, it is yet to be determined how this nomenclature will influence existing clinical guidelines addressing anomalous coronary arteries.

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REFERENCES

1. Cheezum MK, Liberthson RR, Shah NR, et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of Valsalva. *J Am Coll Cardiol*. 2017;69:1592-1608.

2. Finocchiaro G, Behr ER, Tanzarella G, et al. Anomalous coronary artery origin and sudden cardiac death: clinical and pathological insights from a national pathology registry. *JACC Clin Electrophysiol*. 2019;5:516-522.

3. Maron BJ, Haas TS, Ahluwalia A, Murphy CJ, Garberich RF. Demographics and epidemiology of sudden deaths in young competitive athletes: from the United States National Registry. *Am J Med*. 2016;129:1170-1177.

4. Gittenberger-de Groot AC, Sauer U, Oppenheimer-Dekker A, Quaegebeur J. Coronary arterial anatomy in transposition of the great arteries: a morphologic study. *Ped Cardiol*. 1983;4:15-24.

5. Gittenberger-de Groot AC, Koenraadt WMC, Bartelings MM, et al. Coding of coronary arterial origin and branching in congenital heart disease: the modified Leiden Convention. *J Thorac Cardiovasc Surg*. 2018;156:2260-2269.

6. Koppel CJ, Vliegen HW, Bökenkamp R, et al. The Leiden Convention coronary coding system: translation from the surgical to the universal view. *Eur Heart J Cardiovasc Imaging*. 2022;23:412-422.

7. Angelini P. Normal and anomalous coronary arteries: definitions and classification. *Am Heart J*. 1989;117:418-434.

8. Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. *Circulation*. 2002;105:2449-2454.

9. Angelini P. Coronary artery anomalies: an entity in search of an identity. *Circulation*. 2007;115:1296-1305.

10. Dodge-Khatami A, Mavroudis C, Backer CL. Congenital Heart Surgery Nomenclature and Database Project: anomalies of the coronary arteries. *Ann Thorac Surg*. 2000;69:S270-S297.

11. Jacobs ML, Mavroudis C. Anomalies of the coronary arteries: nomenclature and classification. *Cardiol Young*. 2010;20(suppl 3):15-19.

12. Jacobs JP, Franklin RCG, Beland MJ, et al. Nomenclature for pediatric and congenital cardiac care: unification of clinical and administrative nomenclature—the 2021 International Paediatric and Congenital Cardiac Code (IPCCC) and the Eleventh Revision of the International Classification of Diseases (ICD-11). *World J Pediatr Congenit Heart Surg*. 2021;12:E1-E18.

13. Perez-Pomares JM, de la Pompa JL, Franco D, et al. Congenital coronary artery anomalies: a bridge from embryology to anatomy and pathophysiology—a position statement of the Development, Anatomy, and Pathology ESC Working Group. *Cardiovasc Res*. 2016;109:204-216.

14. Brook RH. The RAND/UCLA appropriateness methodology. In: McCormack KA, Moore SR, Seigal RA, eds. *Clinical Practice Guidelines Development: Methodology Perspectives*. Agency for Health Care Policy and Research; 1994:59-70.

15. Dalkey N, Helmer O. An experimental application of the Delphi method to the use of experts. *Manage Sci*. 1963;9:458-467.

16. Dalkey NC. The Delphi method: an experimental application of group opinion. In: Dalkey NC, Rourke DL, Lewis R, Snyder D, eds. *Studies in the Quality of Life*. Lexington Books; 1972.

17. Gurvitz M, Marelli A, Mangione-Smith R, Jenkins K. Building quality indicators to improve care for adults with congenital heart disease. *J Am Coll Cardiol*. 2013;62:2244-2253.

18. Nathan M, Newburger JW, Bell M, et al. Development of the Residual Lesion Score for congenital heart surgery: the RAND Delphi methodology. *Cardiol Young*. 2022;33:977-990.

19. Miller JR, Stephens EH, Goldstone AB, et al. The American Association for Thoracic Surgery (AATS) 2022 expert consensus document: management of infants and neonates with tetralogy of Fallot. *J Thorac Cardiovasc Surg*. 2023;165:221-250.

20. Brothers JA, Frommelt MA, Jaquiss RDB, Myerburg RJ, Fraser CD, Tweddell JS. Expert consensus guidelines: anomalous aortic origin of a coronary artery. *J Thorac Cardiovasc Surg*. 2017;153:1440-1457.

21. Poynter JA, Williams WG, McIntyre S, Brothers JA, Jacobs ML, Congenital Heart Surgeons Society AAOCA Working Group. Anomalous aortic origin of a coronary artery: a report from the Congenital Heart Surgeons Society Registry. *World J Pediatr Congenit Heart Surg*. 2014;5:22-30.

22. Mainwaring RD, Reddy VM, Reinhartz O, Petrossian E, Punn R, Hanley FL. Surgical repair of anomalous aortic origin of a coronary artery. *Eur J Cardiothorac Surg*. 2014;46:20-26.

23. Brothers JA, Whitehead KK, Keller MS, et al. Cardiac MRI and CT: differentiation of normal ostium and intraseptal course from slitlike ostium and interarterial course in anomalous left coronary artery in children. *AJR Am J Roentgenol*. 2015;204:W104-W109.

24. Angelini P, Villason S, Chan AV, Diez JA. Normal and anomalous coronary arteries in humans. In: Angelini P, ed. *Coronary Artery Anomalies: A Comprehensive Approach*. Lippincott Williams & Wilkins; 1999:27-79.

25. Angelini P, Uribe C. Anatomic spectrum of left coronary artery anomalies and associated mechanisms of coronary insufficiency. *Catheter Cardiovasc Interv*. 2018;92:313-321.

26. Doan TT, Zea-Vera R, Agrawal H, et al. Myocardial ischemia in children with anomalous aortic origin of a coronary artery with intraseptal course. *Circ Cardiovasc Interv*. 2020;13:e008375.

27. Rauf H, Zhang X, Hokanson JS. A survey on the management of anomalous aortic origins of the coronary arteries. *Pediatr Cardiol*. 2024;45:1542-1549.

28. Medepalli LC, Medepalli VM, Scully TA. The complementary nature of multimodal imaging in the management of an anomalous aortic origin of the right coronary artery from the left coronary cusp. *J Nucl Cardiol*. 2021;28:2399-2402.

29. Muriago M, Sheppard MN, Ho SY, Anderson RH. Location of the coronary arterial orifices in the normal heart. *Clin Anat*. 1997;10:297-302.

30. Loukas M, Andall RG, Khan AZ, et al. The clinical anatomy of high take-off coronary arteries. *Clin Anat*. 2016;29:408-419.

31. Cantinotti M, Marchese P, Franchi E, et al. Screening of coronary artery origin by echocardiography: definition of normal (and abnormal) take-off by standard echocardiographic views in a healthy pediatric population. *Healthcare (Basel)*. 2022;10:1890.

32. Knight J, Kurtcuoglu V, Muffly K, et al. Ex vivo and in vivo coronary ostial locations in humans. *Surg Radiol Anat*. 2009;31:597-604.

33. Massoudy P, Baltalarli A, de Leval MR, et al. Anatomic variability in coronary arterial distribution with regard to the arterial switch procedure. *Circulation*. 2002;106:1980-1984.

34. Doan TT, Wilkes JK, Reaves O, Neal DL, et al. Clinical presentation and medium-term outcomes of children with anomalous aortic origin of the left coronary artery: high-risk features beyond interarterial course. *Circ Cardiovasc Interv*. 2023;16:e012635.

35. Mery CM, De León LE, Molossi S, et al. Outcomes of surgical intervention for anomalous aortic origin of a coronary artery: a large contemporary prospective cohort study. *J Thorac Cardiovasc Surg*. 2018;155:305-319.e4.

36. Brothers JA, Gaynor JW, Jacobs JP, et al. The registry of anomalous aortic origin of the coronary artery of the Congenital Heart Surgeons' Society. *Cardiol Young*. 2010;20(suppl 3):50-58.

37. Harris MA, Whitehead KK, Shin DC, Keller MS, Weinberg PM, Fogel MA. Identifying abnormal ostial morphology in anomalous aortic origin of a coronary artery. *Ann Thorac Surg*. 2015;100:174-179.

38. Jegatheeswaran A, Devlin PJ, McCrindle BW, et al. Features associated with myocardial ischemia in anomalous aortic origin of a coronary artery: a Congenital Heart Surgeons' Society study. *J Thorac Cardiovasc Surg*. 2019;158:822-834.e3.

39. Agrawal H, Mery CM, Krishnamurthy R, Molossi S. Anatomic types of anomalous aortic origin of a coronary artery: a pictorial summary. *Congenit Heart Dis*. 2017;12:603-606.

40. Krishnamurthy R, Masand PM, Jadhav SP, et al. Accuracy of computed tomography angiography and structured reporting of high-risk morphology in anomalous aortic origin of coronary artery: comparison with surgery. *Pediatr Radiol*. 2021;51:1299-1310.

41. Molossi S, Agrawal H, Mery CM, et al. Outcomes in anomalous aortic origin of a coronary artery following a prospective standardized approach. *Circ Cardiovasc Interv*. 2020;13:e008445.

42. Mézquita AJV, Biavati F, Falk V, et al. Clinical quantitative coronary artery stenosis and coronary atherosclerosis imaging: a consensus statement from the Quantitative Cardiovascular Imaging Study Group. *Nat Rev Cardiol*. 2023;20:696-714.

43. Goldberg A, Southern DA, Galbraith PD, et al. Coronary dominance and prognosis of patients with acute coronary syndrome. *Am Heart J*. 2007;154:1116-1122.

44. Bazzocchi G, Romagnoli A, Sperandio M, Simonetti G. Evaluation with 64-slice CT of the prevalence of coronary artery variants and congenital anomalies: a retrospective study of 3,236 patients. *Radiol Med*. 2011;116:675-689.

45. Agrawal H, Molossi S, Alam M, et al. Anomalous coronary arteries and myocardial bridges: risk stratification in children using novel cardiac catheterization techniques. *Pediatr Cardiol*. 2017;38:624-630.

46. Mainwaring RD, Murphy DJ, Rogers IS, et al. Surgical repair of 115 patients with anomalous aortic origin of a coronary artery from a single institution. *World J Pediatr Congenit Heart Surg*. 2016;7:353-359.

47. Doan TT, Bonilla-Ramirez C, Eilers L, et al. Myocardial bridges in a pediatric population: outcomes following a standardized approach. *J Thorac Cardiovasc Surg*. 2024;168:1203-1212.

48. Lee HJ, Hong YJ, Kim HY, et al. Anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course: subtypes and clinical importance. *Radiology*. 2012;262:101-108.

49. AAOCA reporter. Accessed March 18, 2026. https://deduce.shinyapps.io/aaoca_reporter/

KEY WORDS AAOCA, anomalous aortic origin of a coronary artery, anomalous coronary, coronary artery anomalies, nomenclature

APPENDIX For Supplemental Methodology, please see the online version of this paper.