Received: 26 June 2017 | Accepted: 4 July 2017

DOI: 10.1111/chd.12518

SPECIAL SECTION ON CORONARY ANOMALIES

WILEY Congenital Heart Disease

Anatomic types of anomalous aortic origin of a coronary artery: A pictorial summary

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Abstract

Anomalous aortic origin of a coronary artery is a congenital abnormality of the origin or course of a coronary artery that arises from the aorta. As commonly seen in congenital heart disease, a wide spectrum of anatomic variability is seen and hence, it is important for clinicians to document the precise anatomy and course of the anomalous vessel. This article describes coronary artery nomenclature using computerized tomography angiography and virtual angioscopy. These details are important for decision making, useful for surgical planning, and may have prognostic implications.

KEYWORDS

anomalous coronary arteries, interarterial, intramural, intramyocardial course, sudden cardiac death

1 | CLASSIFICATION

The Coronary Anomalies Program at Texas Children's Hospital, created in December 2012, has prospectively followed patients with anomalous aortic origin of a coronary artery (AAOCA) and chosen computerized tomography angiography (CTA) with virtual angioscopy as the preferred mode of imaging. The detailed course of the anomalous vessel and precise description of the location, shape and spatial relationship of the coronary ostia are reported in all patients. We have developed a standardized nomenclature system (Figure 1) that helps to describe and classify the location and morphology of the anomalous vessel, thus aiding to decision making and precise definition of the optimal surgical approach, if needed.[1] Normally, a coronary artery arises below the sinotubular junction (Levels I or II, Figure 1B) and, if the vessel arises ≥ level III, it is considered high origin (Figure 2). A coronary artery arising above the sinotubular junction (level IV) is described as originating from the ascending aorta.

The anatomic variants of AAOCA seen in our Program were classified, based on the origin of the anomalous vessel, as follows:

1. Anomalous left CA: a) Right sinus, b) Noncoronary sinus, c) High origin left lateral aortic wall, d) High origin above the commissure, e) High origin right lateral aortic wall.
2. Anomalous right CA: a) Left sinus, b) High origin left lateral aortic wall, c) High origin above the commissure, d) High origin right lateral wall.
3. Single CA: a) Left sinus, b) Right sinus, c) High origin above the commissure (Figure 3).
4. Anomalous circumflex CA: a) Right sinus, b) Origin from right CA.

2 | COURSE OF THE ANOMALOUS CORONARY ARTERY

The course of the coronary artery can be classified as follows[2,3]:

1. Intramural course: The proximal CA travels within in the aortic wall before entering the mediastinum. The presence and length of the intramural course is assessed using cross sectional shape of the lumen and the pericoronary fat sign (Supporting Information Videos S1 and S2, Figure 4), first described at our institution.[4]
2. Interarterial course: The proximal CA travels between the aorta and the pulmonary artery (Figure 4).
3. Intramyocardial course: The coronary artery travels within the myocardium instead of having a normal epicardial course (Figure 5), and this includes intraconal/intraseptal course.

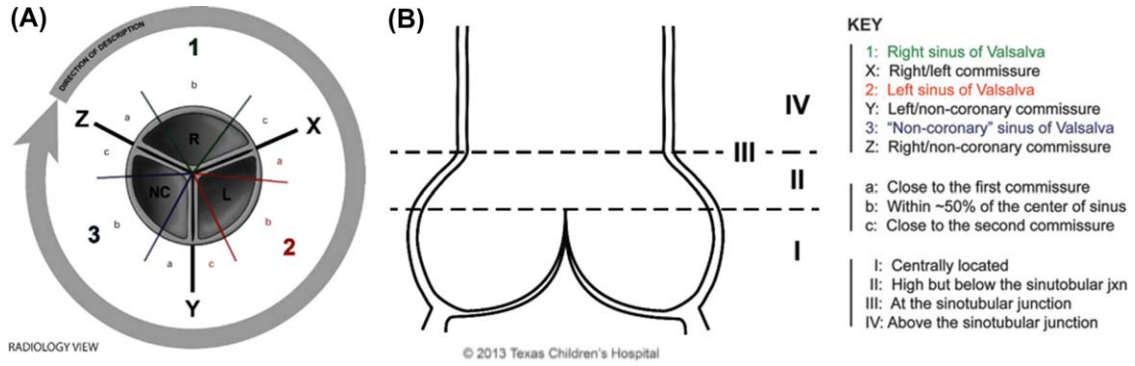

FIGURE 1 Standardized nomenclature map used to describe the origin of the coronary arteries by CT, echocardiography or surgical findings (Reprinted with permission from the 2013 ©Texas Children's Hospital).[1]

4. Prepulmonic course: The proximal CA travels anterior to the pulmonary valve.
5. Subpulmonic course: The proximal CA travels below the level of the pulmonary valve annulus.
6. Retroaortic course: The proximal CA travels posterior to the aorta.

3 | OSTIAL MORPHOLOGY OF THE ANOMALOUS CORONARY ARTERY

The shape and caliber of the ostia can be classified as: normal or round (antero-posterior (AP) diameter equals transverse diameter), oval (transverse diameter is 50%–90% of AP diameter), slit-like (transverse diameter is <50% of AP diameter) (Figure 6).

If the coronary arteries arise in close proximity to each other, the ostia are described using a four-level grading system as follows[5] (Figure 7):

1. Two separate non-confluent orifices arising from the same sinus.
2. Two confluent orifices located adjacent to each other.

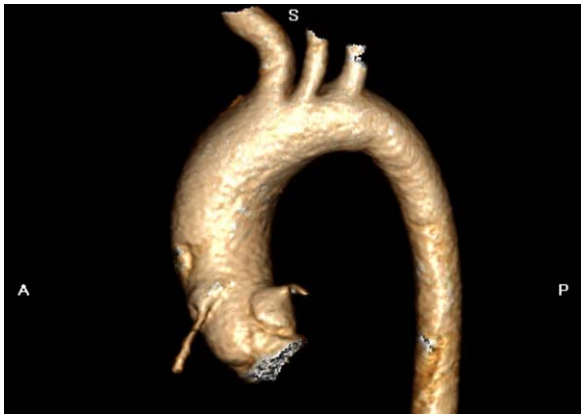

FIGURE 2 An 8-year-old female with a diagnosis of Ehlers Danlos syndrome and high origin of the right coronary artery from the right wall of the ascending aorta, just superior to the right sinus of Valsalva at the level of the sinotubular junction (1c, Level III)

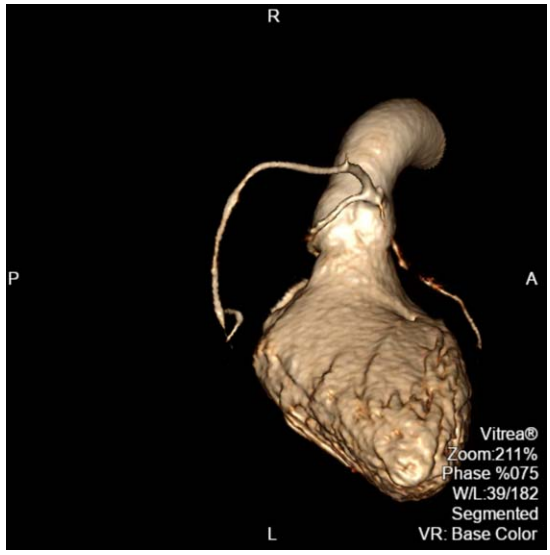

FIGURE 3 A 16-year-old male with single coronary trunk arising from the anterior facing right sinus with trifurcation into the right coronary artery, circumflex and left coronary artery. The left anterior descending demonstrated a long intramyocardial course of about 7 cm

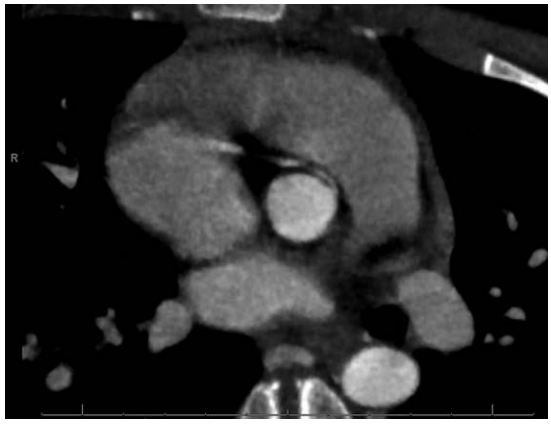

FIGURE 4 A 12-year-old male with anomalous right coronary artery from the left sinus (2a) with a 4.6 mm intramural and 14 mm interarterial course

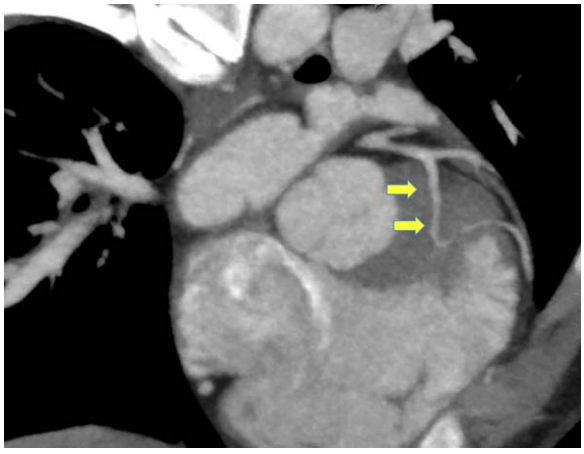

FIGURE 5 A 14-year-old male with a 4 cm intramyocardial course of the mid left anterior descending coronary artery coursing below the level of the pulmonary valve with an intraconal course

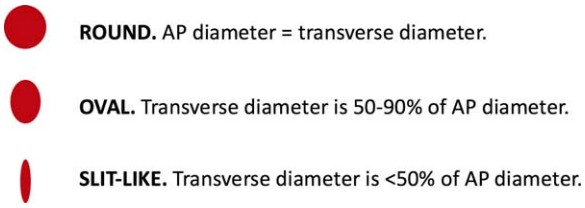

FIGURE 6 Schematic showing the classification of the shape of coronary ostium. (Reprinted with permission from the 2015 ©Texas Children's Hospital)

3. Single orifice with bifurcation in the aortic wall.
4. Single orifice with bifurcation outside the aortic wall.

4 | CLINICAL IMPLICATION

The postulated mechanisms of ischemia in patients with AAOCA and/or myocardial bridges include lateral compression, torsion or kinking of the intramural/interarterial coronary artery during exercise, systolic compression of the intramyocardial segment, or vasospasm of the anomalous coronary artery secondary to endothelial damage.[1,6,7] Coronary artery obstruction can lead to compromise in coronary flow, particularly during exercise, with resultant myocardial ischemia, ventricular tachycardia, or ventricular fibrillation. We believe appropriate description of the anatomic variant of the anomalous vessel, in addition to clinical presentation and myocardial perfusion studies, contribute to risk stratification and decision making in these patients.

5 | CONCLUSION

We describe the classification and anatomic considerations used in our Coronary Anomalies Program at Texas Children's Hospital based on CTA. We consider patients to be at high risk if they present with high risk anatomy (anomalous left CA with interarterial course, presence of intramurality, abnormal ostium), symptoms suggestive of ischemia, and/or evidence of myocardial perfusion abnormalities on stress nuclear

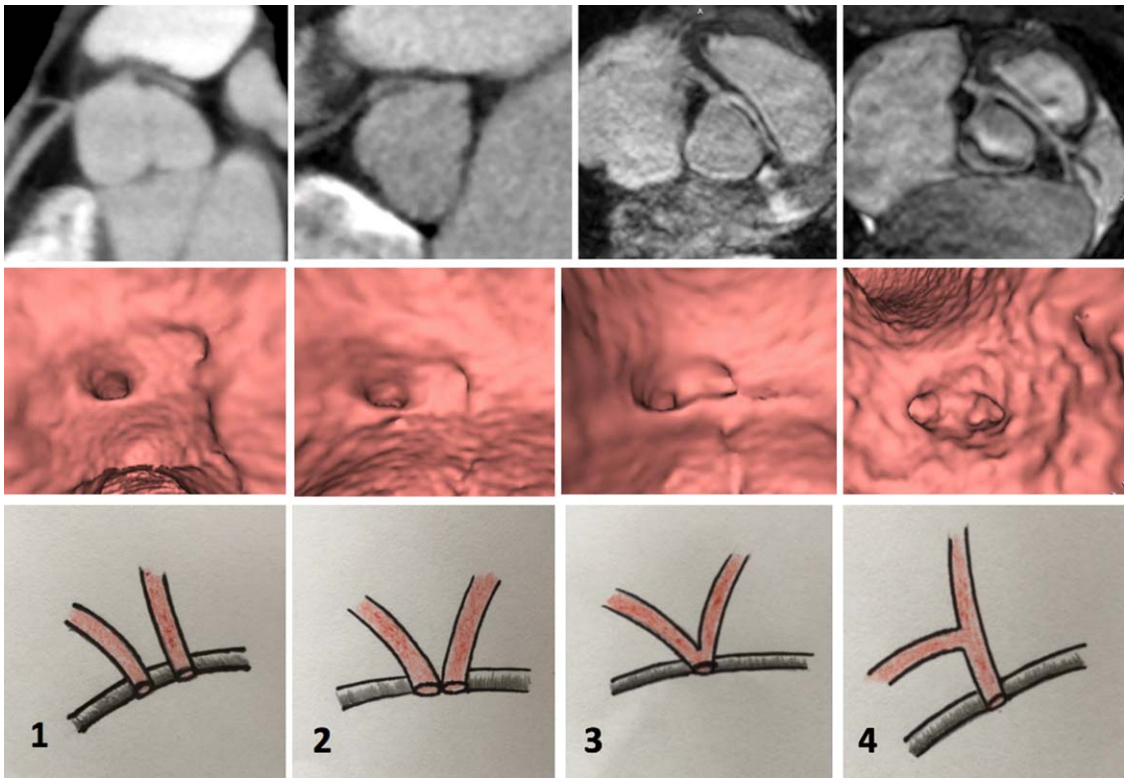

FIGURE 7 Grading of the coronary ostia using CT angiography and Virtual angioscopy

perfusion imaging and/or stress cardiac magnetic resonance imaging. In general, prepulmonic, subpulmonic, or retroaortic courses are considered benign. We propose these details are useful for decision making and surgical planning, and may have long term prognostic implications.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS

All the listed authors have made significant contribution in the preparation and review of the manuscript.

REFERENCES

[1] Mery CM, Lawrence SM, Krishnamurthy R, et al. Anomalous aortic origin of a coronary artery: toward a standardized approach. *Semin Thorac Cardiovasc Surg*. 2014;26:110–122.

[2] 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.

[3] Angelini P, Villason S, Chan AV, Diez JG. Normal and anomalous coronary arteries in humans. In: Angelini P, ed. Coronary Artery Anomalies. A Comprehensive Approach. Philadelphia, PA: Lippincott Williams & Wilkins; 1999:27–150.

[4] Krishnamurthy R, Masand P, Jadhav S, et al. Diagnostic accuracy of CT angiography (CTA) for critical pathologic features in anomalous aortic origin of the coronary arteries (AAOCA) in children: a comparative study with surgery in a single center. *J Am Coll Cardiol*. 2015;65:A1304.

[5] 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 Congenital Heart Surg*. 2014;5:22–30.

[6] Angelini P. Novel imaging of coronary artery anomalies to assess their prevalence, the causes of clinical symptoms, and the risk of sudden cardiac death. *Circulation: Cardiovasc Imaging*. 2014;7:747–754.

[7] Tarantini G, Migliore F, Cademartiri F, Fraccaro C, Iliceto S. Left anterior descending artery myocardial bridging: a clinical approach. *J Am Coll Cardiol*. 2016;68:2887–2899.

SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article.

VIDEO CLIP S1 CT angiography showing the pericoronary fat sign in a patient with anomalous left coronary artery. There is absence of pericoronary fat (which appears dark black) around the proximal left coronary artery.

VIDEO CLIP S2 CT angiography showing an intramural course based on the cross sectional shape of the lumen in a patient with anomalous left coronary artery. The proximal left coronary artery lumen is oval and changes to a round lumen after the intramural course.

How to cite this article: Agrawal H, Mery CM, Krishnamurthy R, Molossi S. Anatomic types of anomalous aortic origin of a coronary artery: A pictorial summary. *Congenital Heart Disease*. 2017;12:603–606. https://doi.org/10.1111/chd.12518