JACC: CARDIOVASCULAR INTERVENTIONS
VOL. 14, NO. 9, 2021

PUBLISHED BY ELSEVIER

STATE-OF-THE-ART REVIEW

Catheter Selection and Angiographic Views for Anomalous Coronary Arteries

A Practical Guide

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ABSTRACT

Although congenital coronary artery anomalies are relatively rare, they are the second most common cause of sudden cardiac death among young athletes. When encountered in the cardiac catherization laboratory, they are often challenging to selectively engage, requiring multiple catheters, plus increased contrast volume and radiation exposure. In the setting of acute coronary syndromes, it is not infrequent that percutaneous intervention is delayed because of the inability to engage an anomalous coronary artery. The aim of this review is to provide a comprehensive and concise overview of coronary artery anomalies, with particular attention to diagnostic and guide catheter selection for each type of anomaly and recommendations on how to recognize the vessel course angiographically.
(J Am Coll Cardiol Intv 2021;14:995–1008) © 2021 by the American College of Cardiology Foundation.

Congenital coronary artery anomalies are relatively rare, encountered in 0.2% to 1.2% of patients undergoing percutaneous coronary intervention (PCI), and are the second most common cause of sudden cardiac death among young athletes (1). The unusual ostial location and course of an anomalous coronary artery can pose considerable technical challenges in the catheterization laboratory, often requiring large iodinated contrast volumes and high radiation exposure, and may delay intervention in acute coronary syndromes (ACS). If the anomalous origin is not recognized, the operator may wrongly diagnose the anomalous vessel as being occluded. This review provides a comprehensive overview of coronary artery anomalies and summarizes typical patterns of anomalous coronary artery anatomy, with particular attention to diagnostic and guide catheter selection for each type of anomaly. The selection of an appropriate catheter is critical to enhancing rapid and high-quality angiography, properly assessing lesion characteristics, and facilitating successful PCI if needed. In the setting of previously undocumented or unknown coronary artery anomalies, it is usually more efficient to first obtain a high-quality deep aortic root angiogram, which will often provide a broad view, allowing rapid identification of at least the origins of the coronary arteries, rather than "hunt" for them with a long series of

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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.

Manuscript received October 1, 2020; revised manuscript received December 28, 2020, accepted January 19, 2021.

ISSN 1936-8798/$36.00
https://doi.org/10.1016/j.jcin.2021.01.054

ABBREVIATIONS AND ACRONYMS

ACS = acute coronary syndrome(s)

CT = computed tomographic

FFR = fractional flow reserve

IVUS = intravascular ultrasound

LAD = left anterior descending coronary artery

LCx = left circumflex coronary artery

PCI = percutaneous coronary intervention

RCA = right coronary artery

TAVR = transcatheter aortic valve replacement

standard-shaped selective diagnostic coronary catheters. In this setting, it is also wise to obtain the initial aortogram in the straight anterior-posterior and straight lateral projections, which provide the operator a solid reference point from which to begin the identification process and assist in selecting the most likely catheter shape to engage selective coronary artery anomalies. After this, one can simply fine-tune the catheter to obtain the best engagement and additional compound views. Biplane angiographic systems provide an obvious advantage over single-plane laboratories in these situations. With biplane imaging, a single injection of contrast medium allows 2 cineangiography runs to be recorded simultaneously. Both femoral and radial access routes are available; in cases with subclavian loop or tortuosity, femoral access is preferred. If there is a pre-existing high degree of suspicion for a coronary artery anomaly in the non-ACS setting, computed tomographic (CT) angiography can provide rapid and accurate information regarding the origins and course of the coronary arteries, which will reduce the need for some preliminary angiograms and selective coronary catheter uncertainty, which can arise during a cardiac catheterization.

ABSENT LEFT TRUNK

Absence of the left main coronary artery, the most common anomaly, is found in 1% to 2% of the general population. In this anomaly, the left anterior descending coronary artery (LAD) and left circumflex coronary artery (LCx) arise directly from the aortic root as separate vessels. As a general rule, a half-size smaller catheter should be used to engage the LAD

HIGHLIGHTS

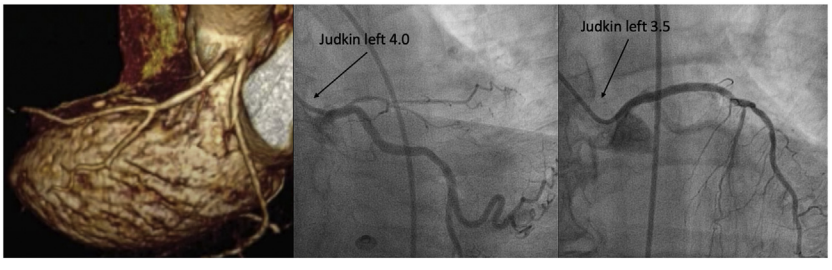
- Anomalous aortic origin of coronary artery is associated with sudden cardiac death.
- Coronary artery anomalies are challenging to selectively engage.
- The authors summarize the best angiographic views for anomalous vessel course.
- The authors review catheter selection for selective engagement for each anomaly type.

ostium compared with the LCx ostium. Then, to move the selective catheter from the LCx to the LAD, a counterclockwise maneuver is performed, which moves the catheter tip anteriorly within the left sinus. For example, in a normal aortic root size, a Judkins left 3.5 should be used for the LAD and a Judkins left 4.0 for the LCx (Figure 1).

ANOMALOUS RIGHT CORONARY ARTERY FROM A VARIETY OF LOCATIONS WITHIN THE SINUSES OF VALSALVA

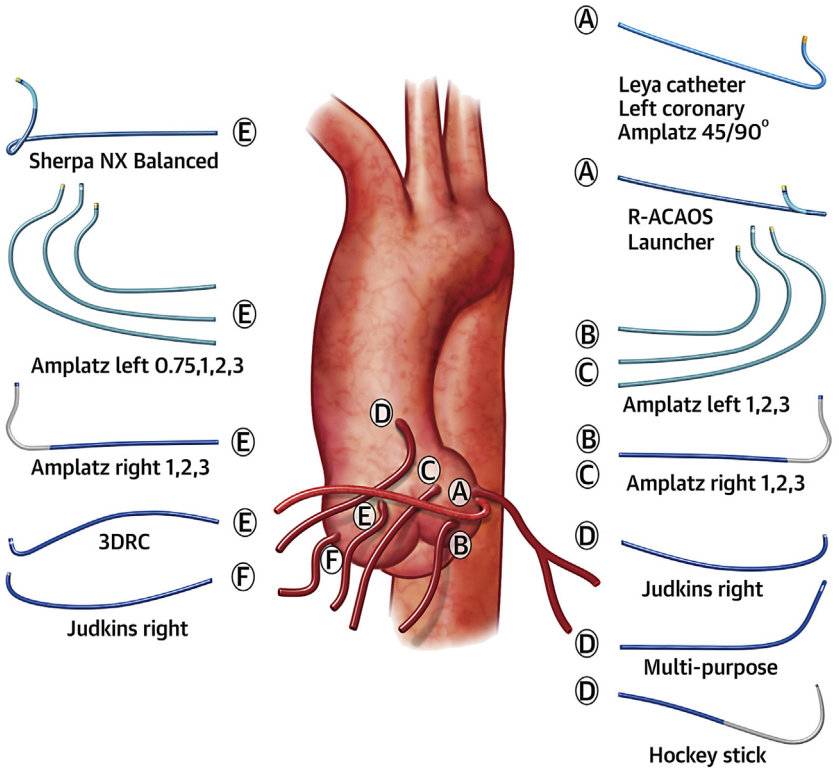
An anomalous right coronary artery (RCA) can arise from a variety of locations within the sinuses of Valsalva. The Central Illustration summarizes these different locations: A) RCA ostium adjacent to left main coronary artery ostium; B) RCA ostium inferior to left main coronary artery ostium; C) RCA ostium toward the commissure of the right and left cusps; D) ostium above the sinotubular plane; E) ostium from right cusp superior and toward the left cusp; and F) usual origin.

FIGURE 1 Separate Ostia of Left Anterior Descending Coronary Artery and Left Circumflex Coronary Artery



Computed tomographic angiographic imaging of absent left trunk variant with separate ostia of left anterior descending coronary artery (LAD) and left circumflex coronary artery (LCx); angiographic image with Judkins left 3.5 engagement for LAD and Judkins left 4.0 for LCx.

CENTRAL ILLUSTRATION **Representative Diagram of Anomalous Origin of Right Coronary Artery From Left Sinus of Valsalva**



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(A) Origin adjusted to the ostium of the left main coronary artery; **(B)** inferior to left main coronary artery ostium; **(C)** toward the commissure of the right and left cusps; **(D)** origin from the aorta above the sinotubular plane; **(E)** origin from right cusp superior and toward the left cusp; **(F)** usual origin.

For location A, the origin adjacent to the ostium of the left main coronary artery is the most common and is a technical challenge to engage. One of the 2 best catheters with special 3-dimensional shapes for selective engagement is the Leya catheter left coronary Amplatz 45° or 90° (Cordis Cardinal Health, Dublin, Ohio) in 7-F size only. **Figure 2 and** Videos 1 and 2 demonstrate angiography using an Amplatz 90° catheter with the corresponding CT image with intravascular ultrasound (IVUS) pull back showing severe lateral compressions of the proximal anomalous RCA that increase during systole.

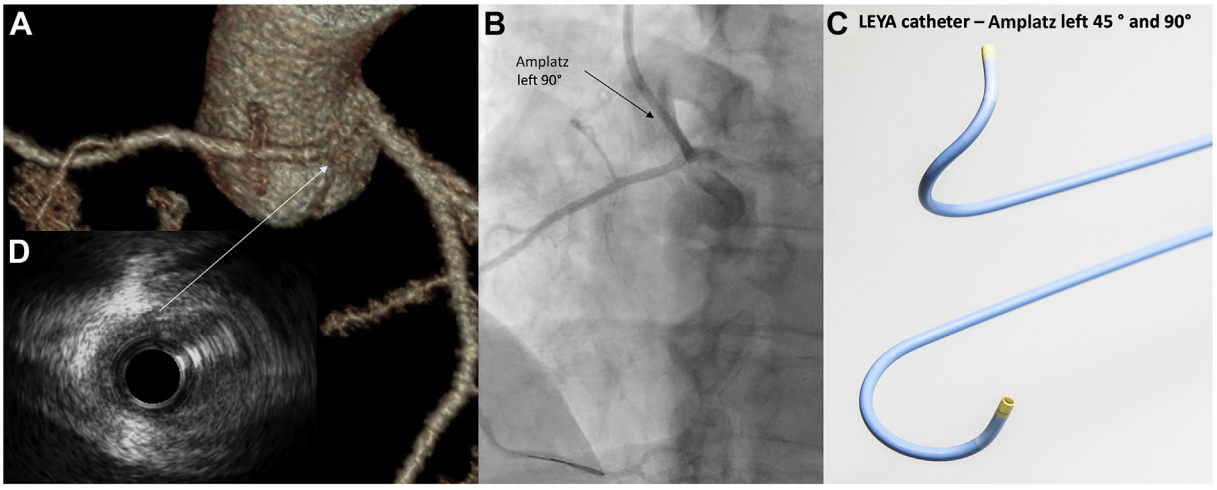
The other option for location A is an R-ACAOS Launcher coronary guide catheter (Medtronic, Minneapolis, Minnesota) in 6-F size (**Figure 3 and** Video 3). Because of the common presence of a proximal intramural course of the RCA in this setting, extra care should be taken to mitigate the heightened risk for dissection by the catheter tip.

Locations B and C usually are easier to engage with Amplatz left 1,2,3 or Amplatz right 1,2,3 on the basis of the aortic root dimensions. Location D is similar to saphenous vein graft engagement with Judkins 4 right catheter, hockey stick, or multipurpose or Amplatz left or right. If selective engagement cannot be achieved, the operator can point a guide catheter nonselectively toward the ostium, pass a coronary guidewire through the ostium and down the coronary artery, and then advance a guide extension catheter (**Figure 4**).

ANOMALOUS RCA FROM RIGHT CUSP: SUPERIOR AND TOWARD THE LEFT CUSP

Location E, an anomalous RCA from the right cusp, can be superior or toward the left cusp and might be difficult to achieve selective engagement and support if intervention is needed. Options include 3DRC and

FIGURE 2 Anomalous Right Coronary Artery From the Left Cusp Engaged With Leya Catheter



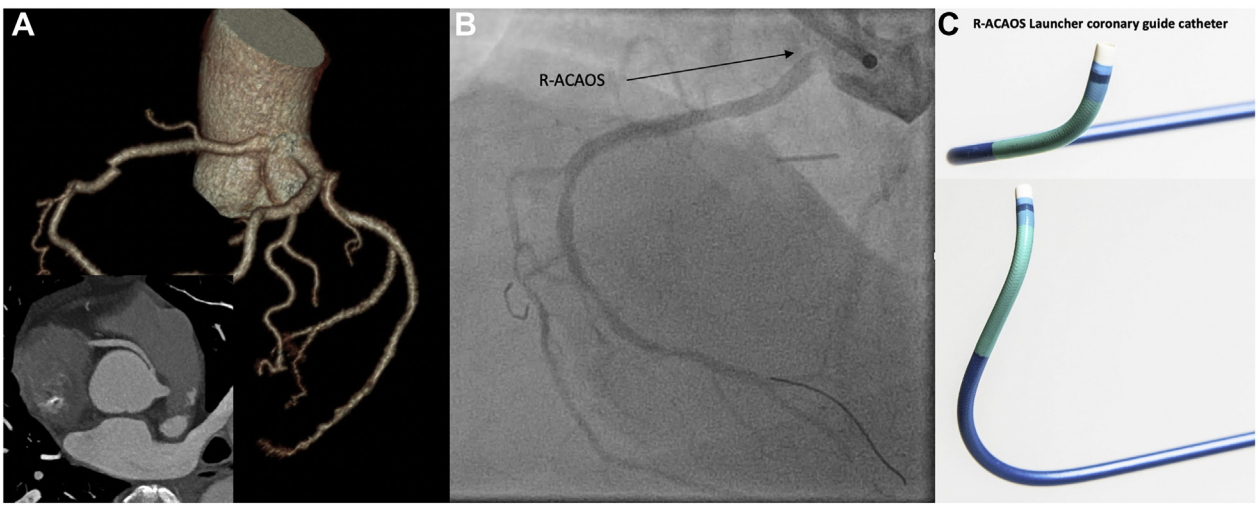
(A) Computed tomographic angiographic imaging of the intramural course of the right coronary artery (RCA) from the left sinus. **(B)** Anomalous RCA adjusted to the left main coronary artery engaged with Leya catheter: Amplatz left 90°. **(C)** Leya catheter: Amplatz left 45° and 90°. **(D)** Intravascular ultrasound pull back shows severe lateral compression of the proximal anomalous RCA, which increases during systole.

Amplatz left or right (Figure 5), but if support is needed, then the Sherpa NX Balanced (Medtronic) is a better option (Figure 6 and Video 4).

ANOMALOUS LCx FROM THE RIGHT CUSP

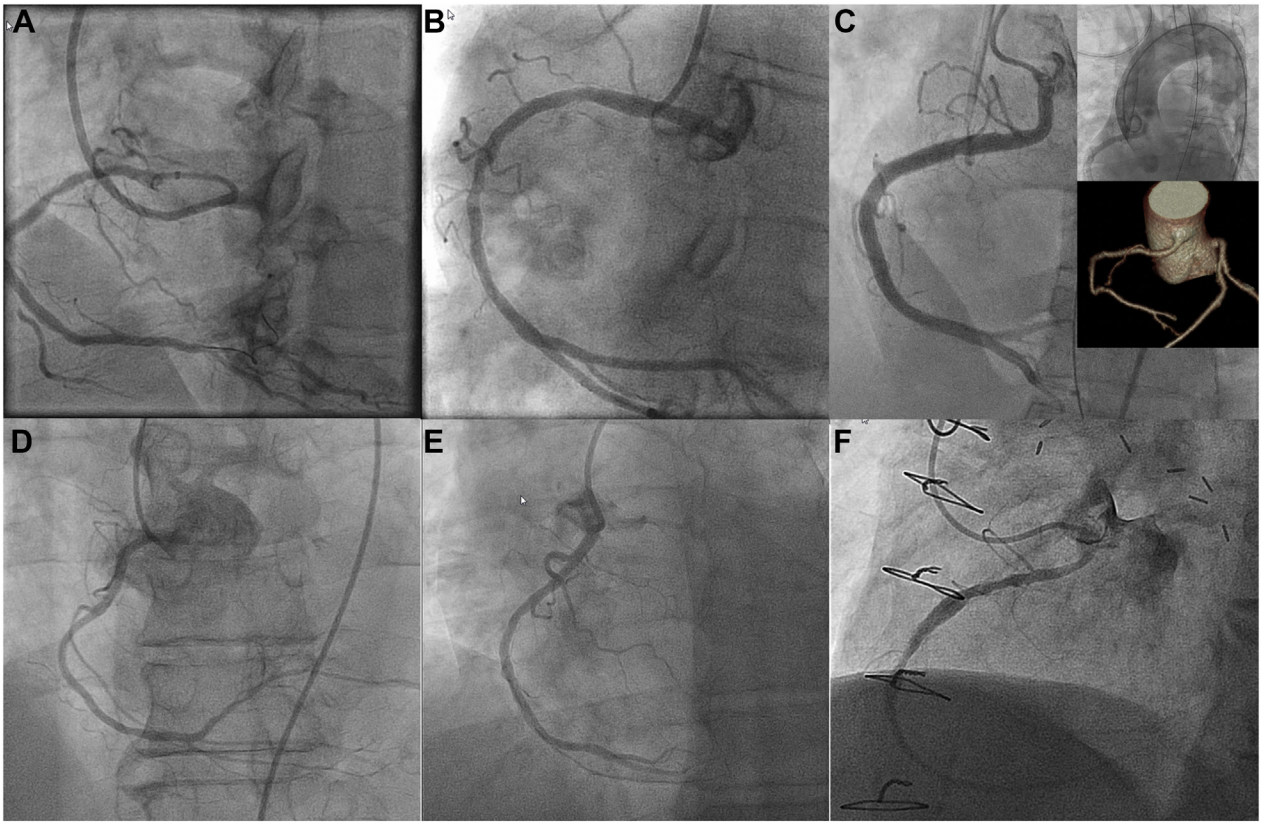
Anomalous LCx from the right coronary sinus with retroaortic course is the most common benign anomaly of coronary arterial origin or course in adults, with a prevalence of approximately 0.3% when assessed on coronary angiography. This variant is not hemodynamically significant but may complicate aortic valve replacement surgery, which risks iatrogenic occlusion of anomalous LCx by inadvertent misplacement of a suture (2). In more recent years, with transcatheter aortic valve replacement (TAVR),

FIGURE 3 Anomalous Right Coronary Artery From the Left Cusp Engaged With R-ACAOS Catheter



(A) Computed tomographic angiographic imaging of the intramural course of the right coronary artery (RCA) from the left sinus. **(B)** Anomalous RCA adjusted to the left main coronary artery engaged with R-ACAOS catheter. **(C)** R-ACAOS catheter.

FIGURE 4 **Anomalous Right Coronary Artery in Various Locations Engaged With Different Catheters**



Anomalous right coronary artery not adjusted to the left main coronary artery, engaged with **(A)** Amplatz left 2 nonselective assists with GuideLiner for selective engagement. **(B)** Amplatz right 1. **(C)** Origin from aorta above the sinotubular plane engaged with Amplatz left 1. **(D)** Amplatz left 0.75. **(E)** 3DRC. **(F)** Amplatz left 3.

there is risk for compression of the anomalous artery by a prosthetic valve (**Figure 7**). In some cases, balloon aortic valvuloplasty has been performed prior to TAVR to assess the potential risk for coronary artery compression by the TAVR valve (3).

An anomalous LCx originating from the right cusp near the RCA ostium or toward the left cusp is best engaged with an Amplatz left or right catheter (**Figure 8 and** Video 5) or with common catheters used to engage the RCA (Judkins 4 right, hockey stick). If the origin is close to the RCA ostium, then selective engagement of the LCx can be challenging, and the anomalous LCx can be missed if the RCA catheter sits deep. The best way is then to engage the RCA with a guide and place a 0.014-inch guidewire in the distal RCA, pull back the guide to the ostium, use a second guidewire to "fish" for the ostium of the LCx, and then direct the guide to the LCx or use a guide extension catheter over the LCx guidewire (**Figure 9**).

POSTERIOR ORIGIN OF THE LEFT MAIN FROM THE LEFT CUSP OR NONCORONARY CUSP

If the left main origin is at the posterior edge of the left cusp, it can be challenging to engage. The best way is to use guide catheters, such as EBU or CLS, rather than diagnostic catheters, and use the back of the 0.35-inch wire to lower the tip of the guide and move the guide posterior (**Figure 10 and** Video 6). The same technique is effective to engage a left main coronary artery arising from the noncoronary cusp (4) (**Figure 11**).

ANOMALOUS LEFT MAIN CORONARY ARTERY FROM THE RIGHT CUSP

Anomalous left main origin of the coronary arteries occurs in up to 0.7% of the general population (5). Anomalous left main coronary arteries from the right cusp can have an interarterial, septal (subpulmonic),

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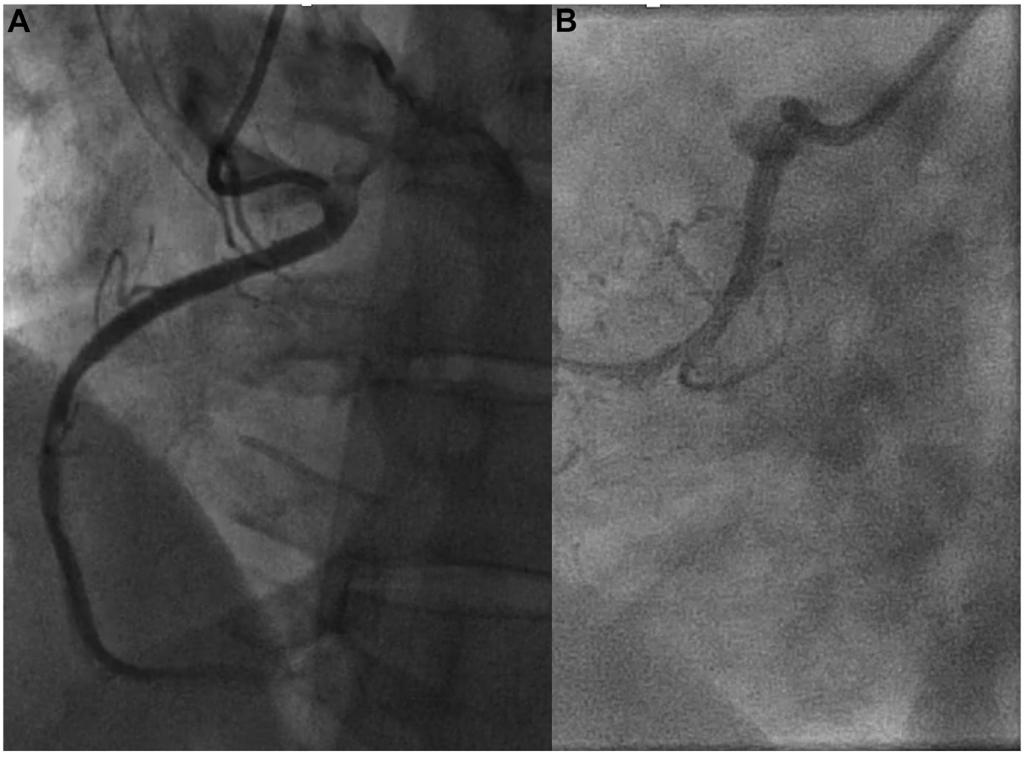
FIGURE 5 Anomalous Right Coronary Artery From Right Cusp Engaged With Amplatz Right and 3DRC Guide

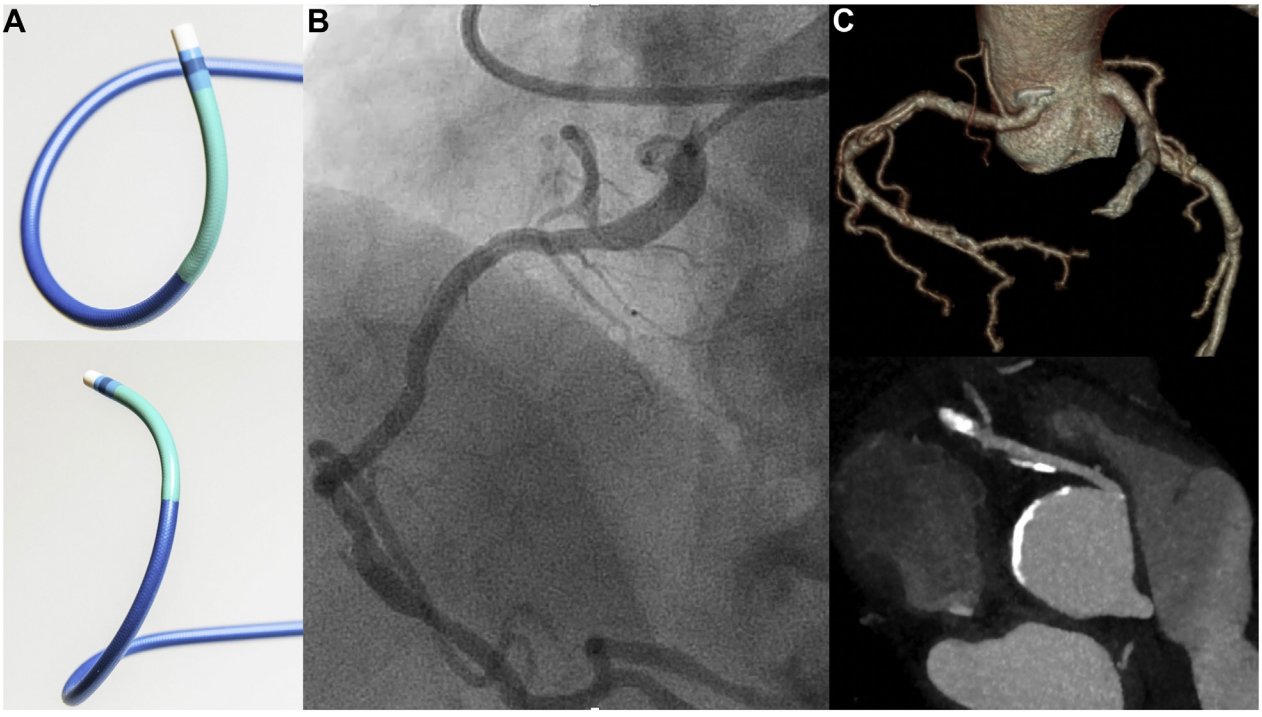


Anomalous right coronary artery from right cusp superior or toward the left cusp engaged with **(A)** Amplatz right and **(B)** 3DRC guide.

retroaortic, or anterior-pulmonic course. Selective catheter engagement of anomalous left main coronary arteries is easier than for an anomalous RCA. Usually, the same catheter, such as a Judkins 4 right or hockey stick, can be used to engage the RCA and simply rotate toward the left cusp. The best angiographic view to ascertain the course of the left main coronary artery is the right anterior oblique caudal view. In this view, a left main coronary artery going upward is anterior to the pulmonary artery, a left main coronary artery looping downward is posterior to the pulmonary artery, and a left main coronary artery going straight most likely has an interarterial course between the aorta and pulmonary artery (**Figure 12**). Similar CT images show the same concept in a caudal view: left main coronary artery going up anterior, down posterior, and straight in between (**Figure 13**). Another method to angiographically ascertain the left main coronary artery course is by placing a Swan-Ganz catheter in the pulmonary artery and obtaining a lateral 90° view. This view allows operators to assess whether the left main coronary artery is anterior to the pulmonary artery, in between the pulmonary artery and aorta, or retroaortic (**Figure 14 and** Video 7).

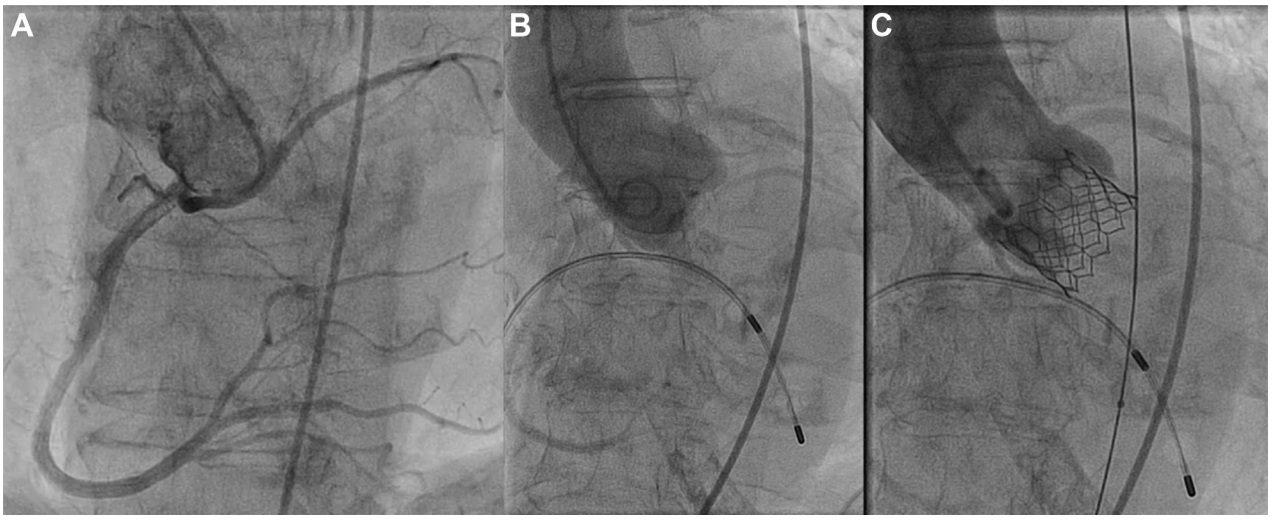
The interarterial course with an intramural segment is associated with sudden death. One hypothesis is that exercise leads to expansion of the aortic root and pulmonary trunk, which may increase the existing angulation of the coronary artery, decreasing the luminal diameter. Another hypothesis is that the vessel has an aberrant course within the aortic wall and is often hypoplastic and exposed to a lateral compression over the entire proximal intramural tract; however, perplexingly, the exact etiology remains controversial. CT angiography is the best tool to assess the course of the left main. CT imaging can demonstrate the vessel course as well as the anatomy of the ostium. The presence of a "slitlike" orifice, an acute takeoff angle, and the length of the intramural segment all are associated with higher risk for sudden cardiac death. Three-dimensional models were superior in representing ostial characteristics (6). **Figure 15** and

FIGURE 6 **Anomalous RCA From Right Cusp Engaged With Sherpa NX Balanced Guide**



(A) Sherpa NX Balanced guide. **(B)** Anomalous RCA from right cusp superior and toward the left cusp engaged with Sherpa NX Balanced guide. **(C)** Computed tomographic images of anomalous right coronary artery from right cusp superior and toward the left cusp.

FIGURE 7 **Transcatheter Aortic Valve in a Patient With Anomalous Left Circumflex Coronary Artery From Right Coronary Artery**



(A) Anomalous circumflex from right cusp retro aortic course engaged with Judkins right catheter. **(B)** Aortogram pre-transcatheter aortic valve replacement. **(C)** Aortogram post-transcatheter aortic valve replacement (Sapien S3) demonstrating risk for compression of the anomalous artery by a prosthetic valve.

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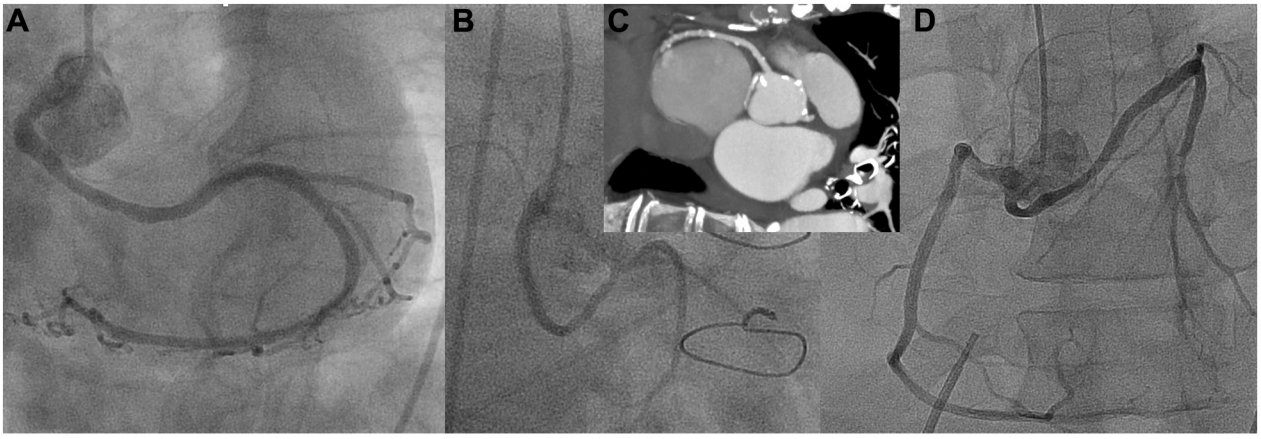
FIGURE 8 Anomalous Left Circumflex Coronary Artery From Right Cusp

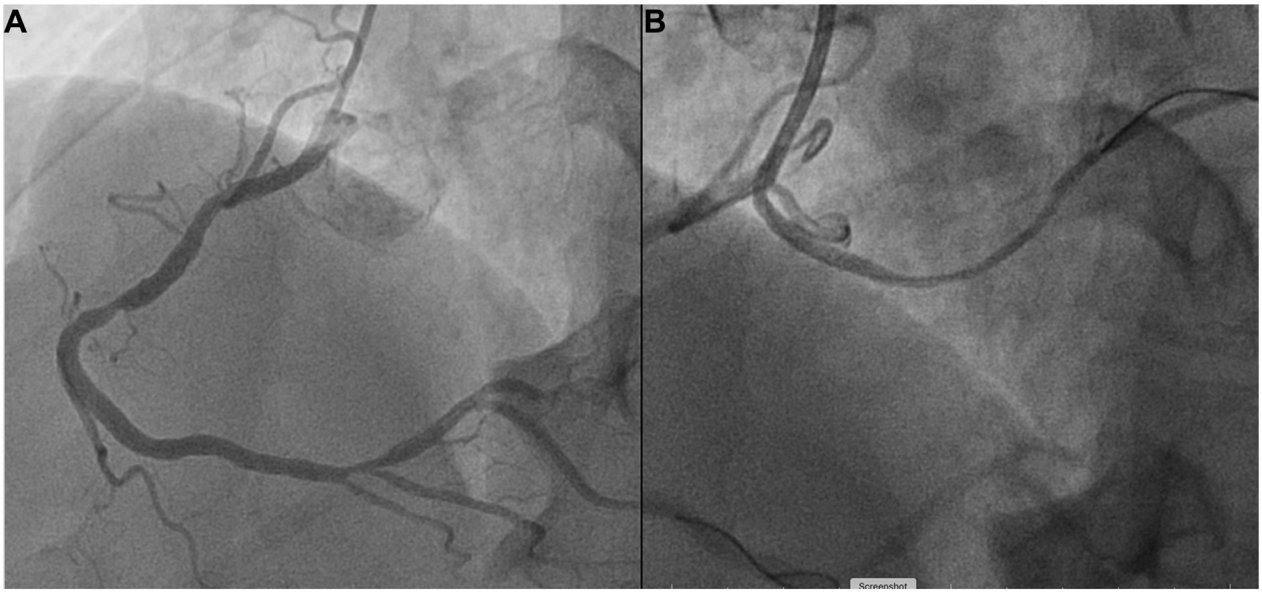


Anomalous left circumflex coronary artery (LCx) from right cusp engaged with **(A)** Amplatz right catheter. **(B)** Multipurpose catheter. **(C)** Computed tomographic imaging of anomalous LCx for right cusp with retroaortic course and **(D)** engaged with Judkins right catheter.

Video 8 show an example of a left main coronary artery with intramural course.

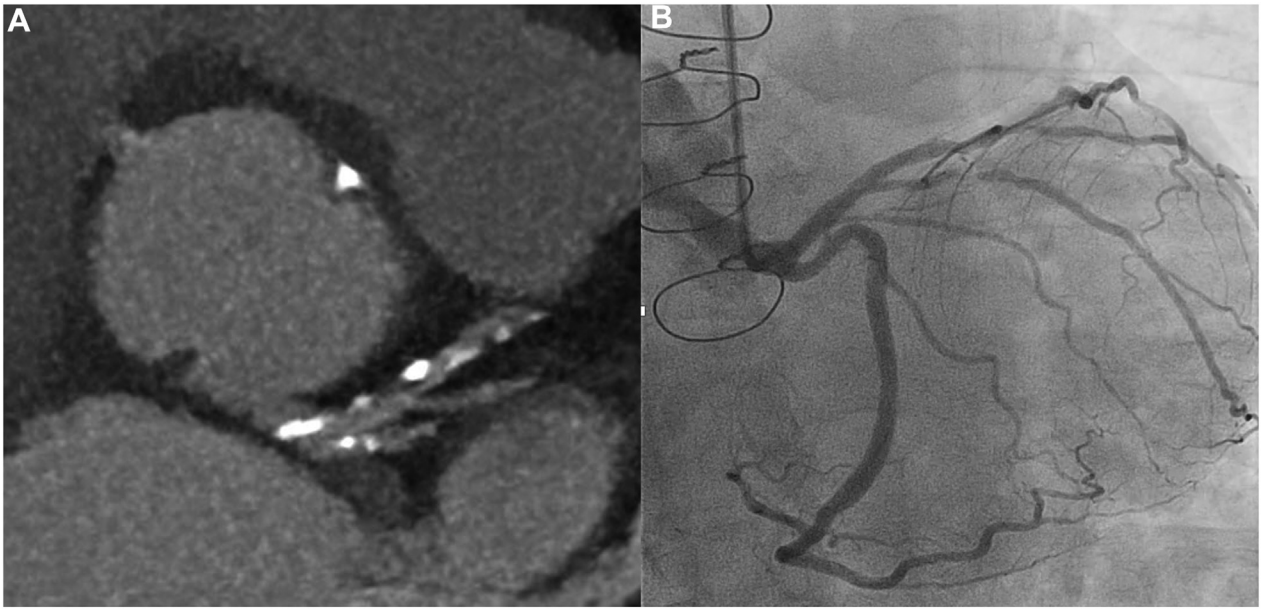
A transseptal anomaly has an inferior course. It usually runs through the septal myocardium, with downward and posterior dip of the left main coronary artery as it exits the right sinus. In **Figure 16**, the left main coronary artery traverses between the pulmonary artery and aorta, diving down through the crista supraventricularis (within the ventricular septum beneath the right ventricular infundibulum). In some cases, the LAD and LCx take different courses. In **Figure 17**, for example, the LAD is anterior to the

FIGURE 9 Catheterization Technique How to Engage Anomalous Circumplex Artery From Right Coronary Artery



(A) Image of the right coronary artery (RCA) with Judkins right seated deep, missing the ostium of anomalous left circumflex coronary artery (LCx). **(B)** Using hockey stick guide and coronary wire in distal RCA to stabilize the guide and pulling back the guide close to the ostium, and second guidewire used to direct the wire to an anomalous LCx with selective image.

FIGURE 10 Left Main Coronary Artery Origin at the Posterior of the Left Cusp



(A) Computed tomographic image. **(B)** CLS guide with back of Terumo 0.35-inch wire helps lower the tip of the guide and move the guide posteriorly for engagement.

pulmonary artery, but the LCx is posterior to the pulmonary artery.

ROLE OF IVUS OR FRACTIONAL FLOW RESERVE

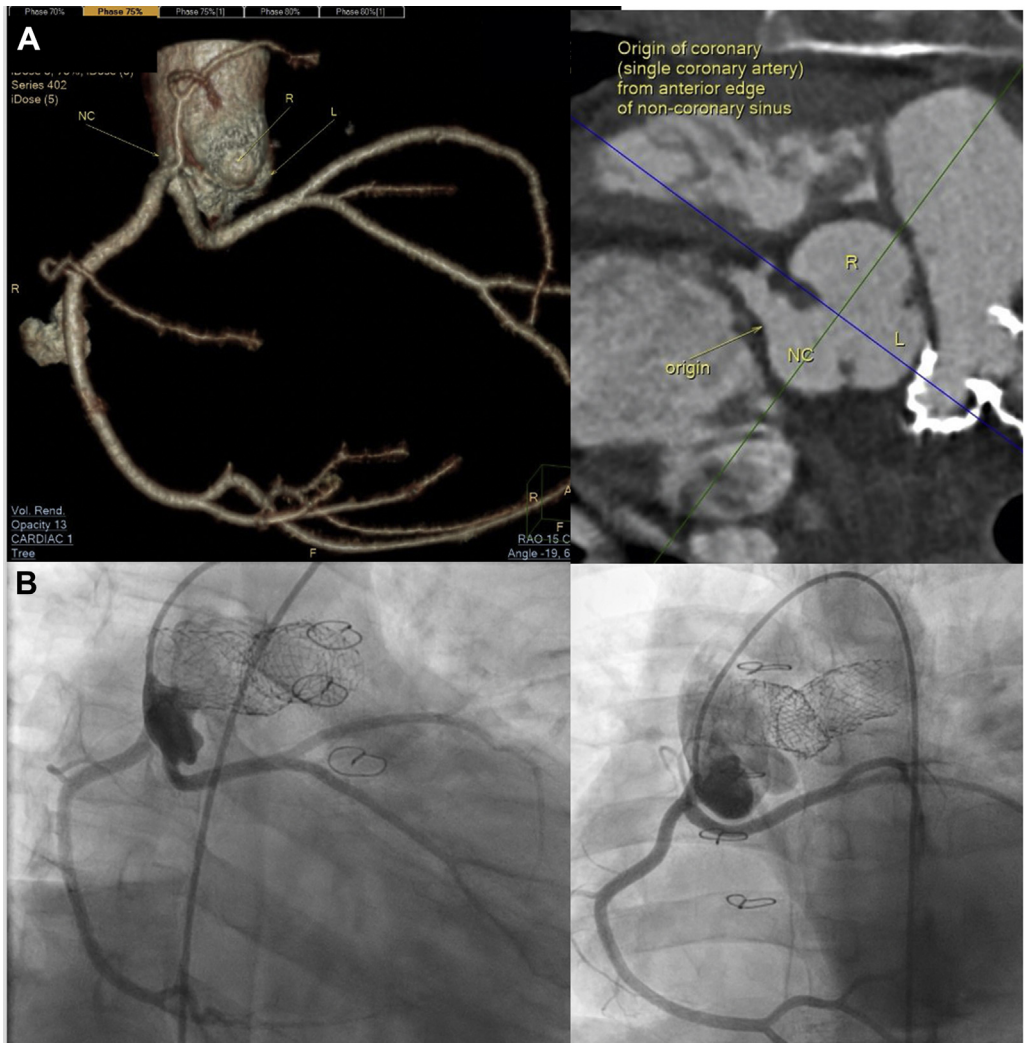
IVUS and fractional flow reserve (FFR) can be safely performed in patients with anomalous coronary arteries and may help with risk stratification, especially in cases in which CT or angiographic findings are equivocal (7). As demonstrated in Video 2, IVUS pull back shows severe lateral compression of the proximal anomalous RCA, which increases during systole. IVUS can identify ostial morphology, interarterial or intramyocardial courses, and caliber changes of coronary arteries during the cardiac cycle. Angelini et al. (8) report the use of IVUS in 67 adults with anomalous RCAs off the left sinus and showed that the severity of stenosis correlates with the occurrence of ischemic symptoms. IVUS can identify the proximal intramural course, with variable degrees of hyperplasia and lateral compression with systolic increment. Few cases report assessing the value of CT-FFR of an anomalous coronary artery to determine clinical significance (9,10).

FFR can be used to assess the significance of anomalous coronary arteries. When performing FFR measurements in patients with anomalous coronary arteries, it is important to leave coronary catheters outside the coronary ostium in the aorta if the lesions are very proximal to ensure accurate results. This is because coronary catheters may obstruct the ostium, particularly in small coronary arteries, or tent open an intramural segment, thus falsely influencing the results. One important difference when evaluating patients with anomalous coronary arteries is the dynamic nature of these lesions compared with atherosclerotic fixed lesions. In atherosclerotic lesions, adenosine is administered to induce hyperemia during FFR assessment. In adults with coronary artery anomalies, dobutamine has been used to mimic exercise changes that occur in these lesions (11).

WHEN SHOULD AN ANOMALOUS CORONARY ARTERY BE TREATED?

LEFT MAIN CORONARY ARTERY FROM RIGHT CUSP. At present, guidelines recommend surgical intervention for anomalous left main coronary artery from the right cusp with interarterial course in the following situations (12): 1) abnormal chest pain or shortness of breath with exertion, syncope, or aborted sudden cardiac death (Class 1, Level of Evidence: B); and 2) positive results on treadmill stress testing, ideally nuclear, in the correct dependent myocardial territory, in the presence of clear evidence of an "intramural" course (Class 1, Level of Evidence: B).

FIGURE 11 Left Main Coronary Artery Origin From Noncoronary Cusp



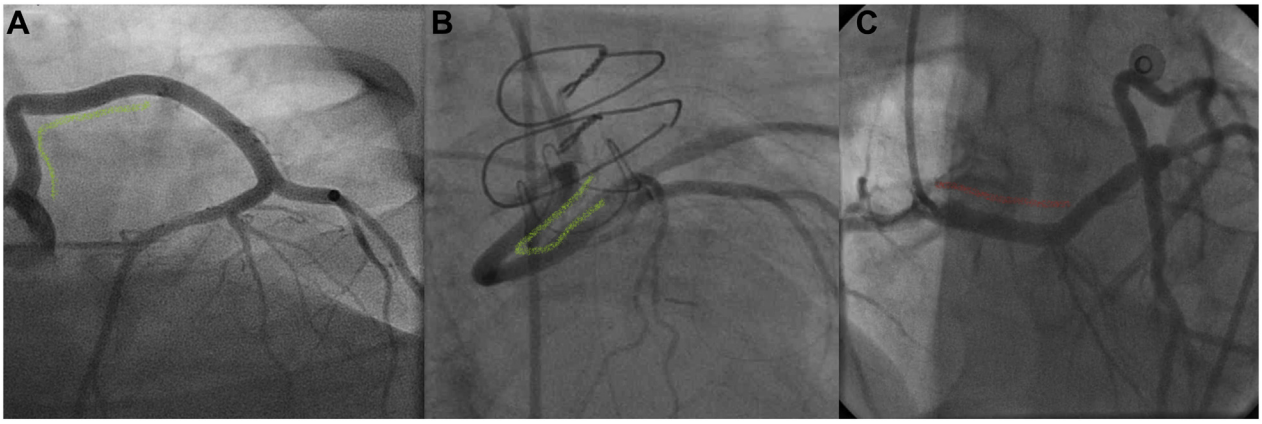
(A) Computed tomographic image. **(B)** Angiographic view engaged with Judkins right catheter. L = left coronary cusp; NC = noncoronary cusp; R = right coronary cusp.

Intervention is surgical and can be accomplished by "unroofing" of the inner side aortic media at the intramural segment to eliminate the aortic half of the stenosing channel. This is the most contemporary management strategy. Coronary artery bypass graft surgery has become obsolete in recent years because of a high incidence of graft failure due to competitive flow unless the proximal native coronary artery is ligated or diseased (13). Translocation of the anomalous arteries is not indicated because the proximal intramural course plays a significant role in the risk for coronary flow compromise.

Intraseptal anomalous aortic origin of a coronary artery is considered a relatively benign condition, and currently, most clinicians defer surgical treatment, but there are recent reports that this anomaly can be associated with ischemia (14). A novel surgical approach through an anterior right ventriculotomy, in which the posterior right ventricular muscle covering the coronary is incised and the outflow tract is reconstructed with a patch, was recently reported (15).

Outcomes of surgical intervention for anomalous aortic origin of a coronary artery were recently reported, showing that it is safe, with a low risk for mortality and other complications. Most patients are cleared for exercise post-operatively and remain asymptomatic (16).

FIGURE 12 Angiographic Views to Identify Left Main Course From the Right Cusp

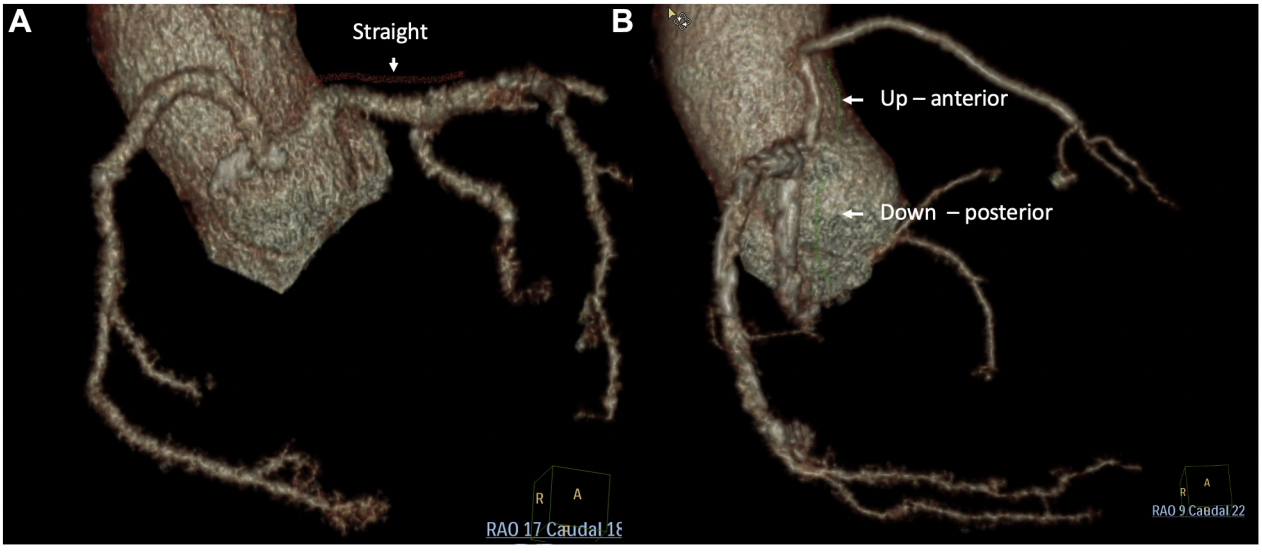


Right anterior oblique caudal views of **(A)** left main coronary artery going up anterior to pulmonary artery, **(B)** left main coronary artery down posterior to pulmonary artery, and **(C)** left main coronary artery straight in between the aorta and pulmonary artery.

RCA FROM LEFT CUSP. Anomalous origin of the RCA from the left sinus is associated with sudden death and myocardial infarction because of the interarterial course of the RCA between the aorta and the pulmonary artery. Kinking of the proximal RCA or "slitlike" orifice into the aorta is also believed to contribute to sudden death and myocardial ischemia.

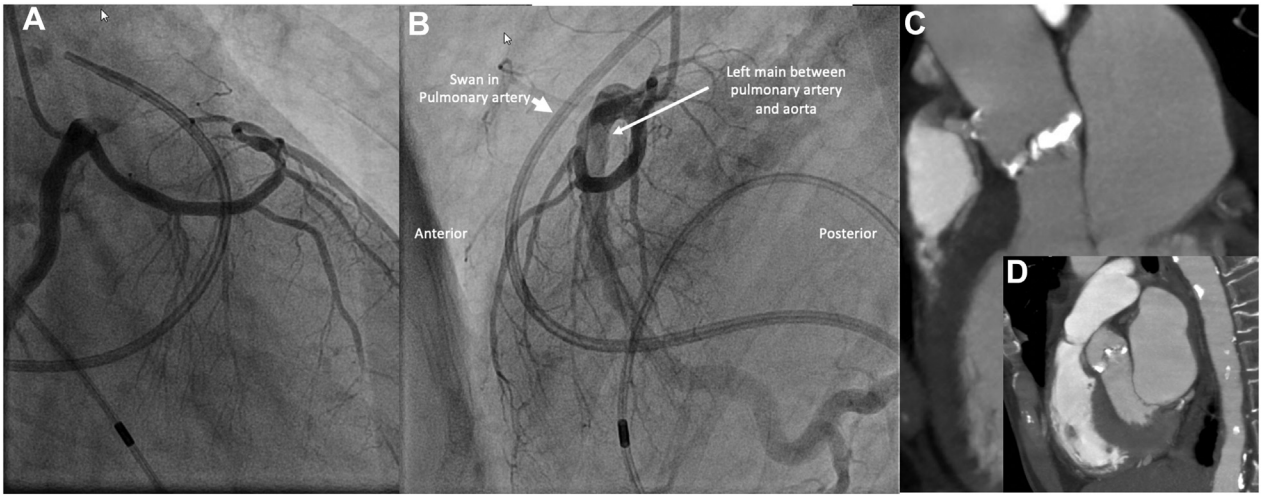
At the present time, the established guideline criteria for intervention for an anomalous RCA from the left cusp depend on patient age, symptoms, and evidence of ischemia. PCI with stents has been reported (8), with high probability of immediate resolution of stenosis and symptoms. However, long-term durability is uncertain because the interarterial course has not been corrected, and ongoing extrinsic compression can lead to stent deformation, with possible stent fracture and vessel occlusion.

FIGURE 13 Computed Tomographic Images



(A) Caudal view left main coronary artery straight between the aorta and pulmonary artery and **(B)** left anterior descending coronary artery going up anterior to pulmonary artery, left circumflex coronary artery down posterior to aorta. RAO = right anterior oblique.

FIGURE 14 Identify Left Main Course With Pulmonary Artery Catheter in Place

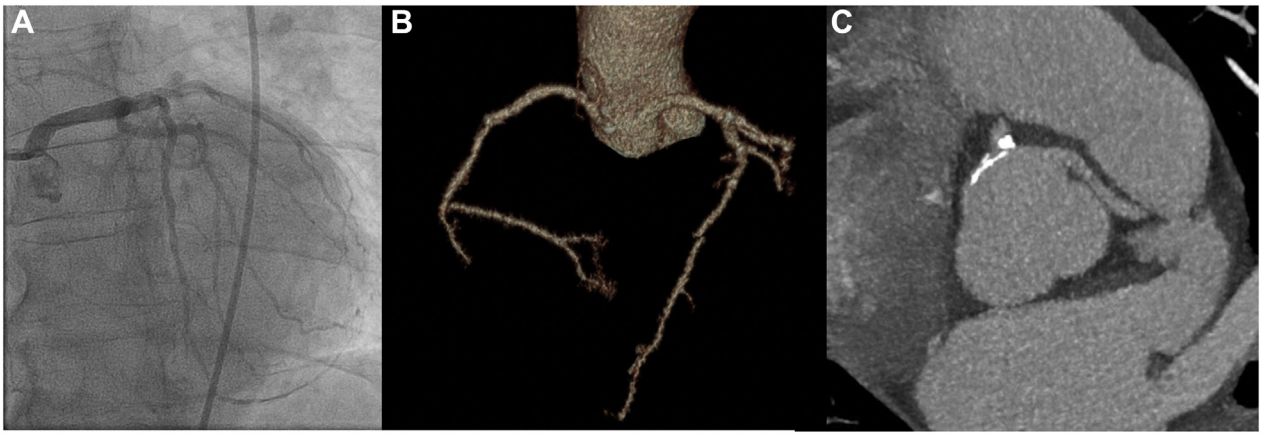


(A) Caudal view showing left main coronary artery and Swan-Ganz but unable to tell if in between pulmonary artery and aorta. **(B)** Lateral view 90° with Swan-Ganz in the pulmonary artery showing left main coronary artery in between aorta and pulmonary artery. **(C)** CT image showing left main coronary artery in between aorta and pulmonary artery diving down with intraseptal course. **(D)** Computed tomographic lateral view 90°, same projection as angiography in **(B)**.

SUMMARY

Coronary artery anomalies present technical challenges in the cardiac catheterization laboratory. There may be difficulty in locating and engaging the ostium of the coronary artery, requiring large contrast volumes and prolonged radiation exposure. In the setting of ACS, intervention can be delayed because of the inability to engage the ostium of a culprit anomalous coronary artery. In addition, there may be

FIGURE 15 Left Main From Right Cusp With Intramural Course



Left main coronary artery between the aorta and pulmonary with intramural course: **(A)** angiography, **(B)** computed tomographic (CT) 3-dimensional image, and **(C)** CT axial image.

FIGURE 16 Computed Tomography and Angiography of Left Main Coronary Artery Between the Pulmonary Artery and Aorta Diving Down Through the Crista Supraventricularis (Within the Ventricular Septum Beneath the Right Ventricular Infundibulum)

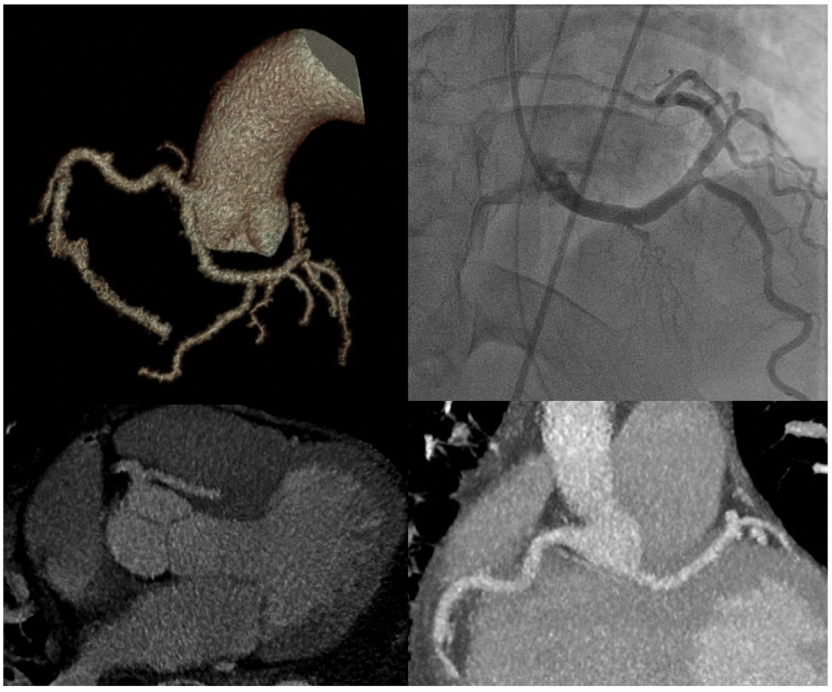
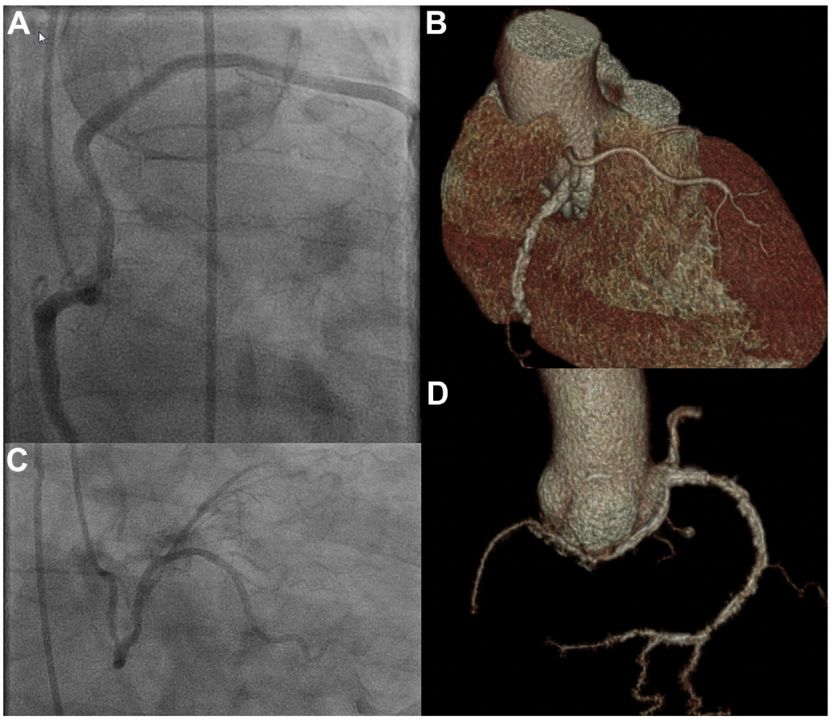


FIGURE 17 Anomalous Left Main Coronary Artery From the Right Cusp



(A) Angiography shows left anterior descending coronary artery (LAD) anterior to pulmonary artery. **(B)** Computed tomography (CT) shows LAD anterior to pulmonary artery. **(C)** Angiography shows left circumflex coronary artery (LCx) retroaortic. **(D)** CT shows LCx retroaortic.

difficulty determining the precise course of the artery. In this review, we have summarized the best angiographic views for anomalous vessel course and the catheter selection for selective engagement for each type of anomaly.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr. Rogers is a proctor and consultant for Medtronic and Edwards Lifesciences; is an advisory board member for Medtronic; and holds equity interest in Transmural Systems. Dr. Slack is a proctor for Abbott. Dr. Pichard is medical director of Abbott Structural Heart. Dr. Waksman is an advisory board member for Amgen, Boston Scientific, Cardioset, Cardiovascular Systems, Medtronic, Philips, and Pi-Cardia; is a consultant for Amgen, Biotronik, Boston Scientific, Cardioset, Cardiovascular Systems, Medtronic, Philips, and Pi-Cardia; has received grant support from AstraZeneca, Biotronik, Boston Scientific, and Chiesi; is a member of the Speakers Bureaus for AstraZeneca and Chiesi; and is an investor in MedAlliance. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS anomalous coronary artery, anomalies of coronary artery, coronary arteries anomalies, catheter selection

APPENDIX For supplemental videos, please see the online version of this paper.