

Table 1. ACC/AHA 2025 GL on ACHD. Comparison of Anomalous Pulmonary Venous Connections With Atrial Septal Defect

	Anomalous Pulmonary Venous Connections	Atrial Septal Defect
Shunt direction	Left-to-right only	Left-to-right, right-to-left, or bidirectional
Risk for paradoxical embolism	No	Yes
Risk for hypoxemia	No	Yes
Magnitude of shunt change over time	No	Yes
Right ventricular volume overload	Yes	Yes
Risk for pulmonary arterial hypertension	Yes	Yes
Transcatheter option	No	Yes

Table 2. ACC/AHA 2025 GL on ACHD. Recommendations for Anomalous Pulmonary Venous Connections (APVC).

Diagnostic

For adults with APVCs, CMR imaging is recommended to assess pulmonary venous anatomy, to evaluate RV size and function, and to determine shunt fraction (Qp:Qs).	1 B-NR
For adults with APVCs who do not have an indication for repair, routine cardiac imaging is recommended for serial evaluation of RV size and function and for assessment of pulmonary hypertension.	1 B-NR
For adults with repaired APVCs and dyspnea on exertion and/or reduced functional capacity, CMR or CT angiography is recommended to evaluate for venous obstruction or residual shunt.	1 B-NR
For adults with unrepaired APVCs and noninvasive evidence of, and/or risk factors for, PAH, cardiac catheterization can be useful to evaluate hemodynamics and aid in determining indication for repair	2a B-NR

Therapeutic

For adults with APVCs who have cardiac symptoms and/or reduced functional capacity with RV volume overload, and whose Qp:Qs is ≥ 1.5 , surgical repair is recommended to improve symptoms and clinical outcomes.	1 B-NR
For adults with APVCs undergoing surgical repair for an ASD, repair of the APVC(s) is recommended, when technically feasible, to reduce left-to-right shunting and RV volume overload.	1 B-NR

For adults with APVCs who are asymptomatic; whose Qp:Qs is ≥ 1.5 ; and who have severe or progressive RV volume overload, reduced RV systolic function, PAH by invasive measurement, or objective exercise intolerance, surgical repair is reasonable in order to improve hemodynamics and/or stabilize RV size and function.

2a B-NR

For adults with APVCs who have a normal to mildly enlarged right ventricle and whose Qp:Qs is < 1.5 , surgical repair is not indicated, given the absence of data to support benefit.

3 Harm B-NR

Table 3. ACC/AHA 2025 GL on ACHD. Routine Follow-up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36-60	24	6-12	3-6
Electrocardiogram	36-60	24	12	12
Transthoracic echocardiogram	36-60	24	12	12

Anomalous Aortic Origin of the Coronary Artery

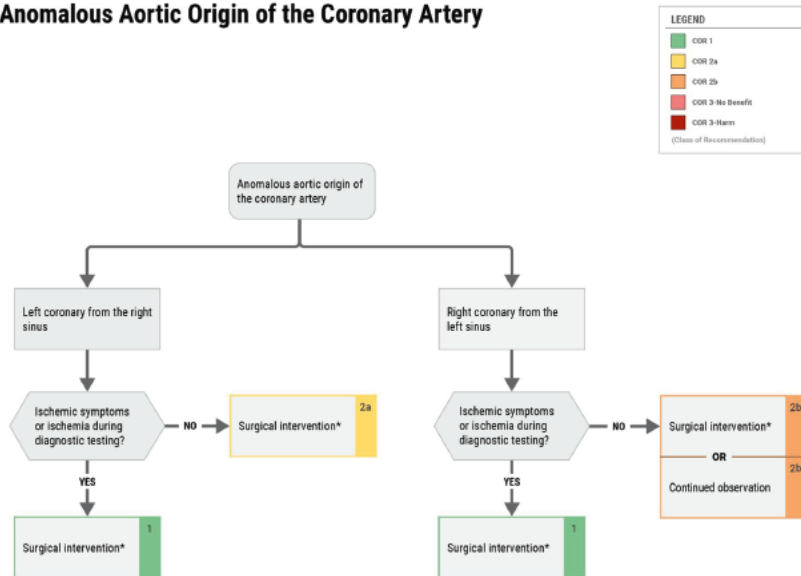


Figure 1. ACC/AHA 2025 GL on ACHD. Anomalous Aortic Origin of the Coronary Artery

*Surgical approaches include coronary unroofing with or without osteoplasty, coronary reimplantation, and pulmonary translocation. Coronary artery bypass graft is reserved for patients with coexisting coronary artery disease.

Table 4. ACC/AHA 2025 GL on ACHD. Recommendations for Anomalous Aortic Origin of a Coronary Artery

Diagnostic

In adults with suspected anomalous origin of a coronary artery (AAOCA), coronary CT angiography or MR angiography (if CT angiography is contraindicated) is recommended to confirm the anatomic diagnosis and guide management. 1 B-NR

In adults with an AAOCA from the opposite sinus, risk stratification with evaluation for physiological and/or ischemic changes should be performed to guide management. 1 B-NR

Therapeutic

In adults with symptomatic AAOCA or diagnostic evidence consistent with myocardial ischemia attributable to the anomalous artery, surgery is recommended. 1 B-NR

In adults with asymptomatic anomalous origin of the left coronary artery, surgery is reasonable in the presence of high-risk anatomy.* 2a C-EO

In asymptomatic adults with an AAOCA from the opposite sinus and without evidence of ischemia or evidence of compromised coronary perfusion, the benefit of surgery is not well established, and continued observation or surgery may be reasonable.

*High-risk findings such as ostial or proximal stenosis, slit-like orifice, acute angle of takeoff, or intramural course.

Follow-up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	24–36	12	12	12
Electrocardiogram	24–36	12	12	12
Trans thoracic echocardiogram	24–36	24	12	12

Table 5. ACC/AHA 2025 GL on ACHD. Recommendations for Anomalous Coronary Artery Arising from the Pulmonary artery

In adults with an anomalous left coronary artery arising from the PA, surgery is recommended to improve cardiac function and reduce the risk for myocardial ischemia 1 B-NR

In symptomatic adults with an anomalous right coronary artery arising from the PA and with evidence of ischemia, surgery is recommended to reduce ischemia, to reduce the risk for SCD, and to improve cardiac function. 1 B-NR

In asymptomatic adults with an anomalous right coronary artery arising from the PA and without evidence of cardiac dysfunction or ischemia, surgery may be considered to avoid the development of cardiac dysfunction. 2b B-NR

Table 6. ESC 2020 GL on ACHD. Recommendations for the management of patients with anomalous coronary arteries

Non-pharmacological functional imaging (e.g. nuclear study, echocardiography, or CMR with physical stress) in patients with coronary anomalies to confirm/exclude myocardial ischaemia. I C

Anomalous coronary arteries from the pulmonary artery

Surgery in patients with ALCAPA. I C

Surgery in patients with ARCAPA and symptoms attributable to anomalous coronary artery. I C

Surgery for ARCAPA in asymptomatic patients with ventricular dysfunction, or myocardial ischaemia attributable to coronary anomaly. IIa C

Anomalous aortic origin of the coronary artery

Surgery for AAOCA in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischaemia in a matching territory or high-risk anatomy.^a I C

Surgery in asymptomatic patients with AAOCA (right or left) and evidence of myocardial ischaemia. IIa C

Surgery in asymptomatic patients with AAOLCA and no evidence of myocardial ischaemia but a highrisk anatomy.^a IIa C

Surgery for symptomatic patients with AAOCA even if there is no evidence of myocardial ischaemia or highrisk anatomy.^a IIb C

Surgery for asymptomatic patients with AAOLCA without myocardial ischaemia and without high-risk anatomy^c when they present at young age (<35 years). IIb C

Surgery is not recommended for AAORCA in asymptomatic patients without myocardial ischaemia and without high-risk anatomy.^a III C

AAOCA = anomalous aortic origin of a coronary artery; AAOLCA = anomalous aortic origin of the left coronary artery; AAORCA = anomalous aortic origin of the right coronary artery; ALCAPA = anomalous left coronary artery from the pulmonary artery; ARCAPA = anomalous right coronary artery from the pulmonary artery; CMR = cardiovascular magnetic resonance.

^a High-risk anatomy includes features such as an intramural course and orifice anomalies (slit-like orifice, acute-angle take-off, orifice >1 cm above the sinotubular junction).

Table 7. ESC 2022 GL on VA and SCD. Ischaemic heart disease and coronary artery anomalies

Recommendations for sudden cardiac death prevention in patients with coronary anomalies

Diagnostic evaluation

Cardiac stress imaging during physical in addition to cardiopulmonary exercise test in patients with anomalous aortic origin of a coronary artery with an interarterial course to confirm/exclude myocardial ischaemia. I C

Cardiac stress imaging during physical exercise in addition to cardiopulmonary exercise test after surgery in patients with anomalous aortic origin of a coronary artery with a history of aborted CA.	I C
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Treatment

Surgery in patients with anomalous aortic origin of a coronary artery with CA, syncope suspected to be due to VAs, or angina when other causes have been excluded.	I C
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Surgery in asymptomatic patients with anomalous aortic origin of a coronary artery and evidence of myocardial ischaemia or abnormal aortic origin of the left coronary artery with high-risk anatomy.*	IIa C
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*: High-risk anatomy is defined as interarterial course, slit-like shaped orifice, high orifice, acute-angle take-off, and intramural course and its length.

Table 8. ESC 2020 GL on Sports. Recommendations for exercise in young individuals/athletes with anomalous origins of coronary arteries

Evaluation with imaging tests to identify high-risk patterns and an exercise stress test to check for ischaemia	IIa C
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In asymptomatic individuals with an anomalous coronary artery that does not course between the large vessels, does not have a slitlike orifice with reduced lumen and/or intramural course, competition may be considered, after adequate counselling on the risks, provided there is absence of inducible ischaemia.	IIb C
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After surgical repair of an AOCA, participation in all sports, at the earliest 3 months after surgery, if they are asymptomatic and there is no evidence of inducible myocardial ischaemia or complex cardiac arrhythmias during maximal exercise stress test.	IIB C
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Participation in most competitive sports with a moderate and high cardiovascular demand among individuals with AOCA with an acutely angled take-off or an anomalous course between the large vessels is not recommended. ^a	III C
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AOCA = anomalous origin of coronary arteries.

a This recommendation applies whether the anomaly is identified as a consequence of symptoms or discovered incidentally, and in individuals <40 years of age.

Table 9. ESC 2020 GL on Sports. Recommendations for exercise/sports in individuals with myocardial bridging

Participation in competitive and leisure-time sports of asymptomatic individuals with myocardial bridging and without inducible ischaemia or ventricular arrhythmia during maximal exercise testing.	IIa C
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Competitive sports are not recommended in individuals with myocardial bridging and persistent ischaemia or complex cardiac arrhythmias during maximal exercise stress testing.	III C
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Table 10. ACC/AHA 2025 GL on ACHD. Anatomy of Vascular Rings and Pulmonary Artery Slings

Complete ring
■ Right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum
■ Right aortic arch with mirror image branching and left ligamentum arteriosum extending from descending aorta to main pulmonary artery
■ Left aortic arch with aberrant right subclavian artery and right ligamentum arteriosum
■ Double aortic arch
■ Circumflex aorta
Incomplete ring
■ Left aortic arch with aberrant right subclavian artery
■ Right aortic arch with aberrant left subclavian artery and right ligamentum arteriosum
Other
■ Innominate artery compression syndrome
■ Left pulmonary artery sling

Table 11. ACC/AHA 2025 GL on ACHD. Recommendations for Vascular Rings

Diagnostic

For adults with unexplained symptoms of airway or esophageal compression that raise concern for vascular ring or PA sling, anatomic assessment with cross-sectional imaging of the aortic arch and PAs is recommended. 1 B-NR

For adults with persistent symptoms of airway or esophageal compression after repair of vascular ring or PA sling, cross-sectional advanced imaging is recommended to evaluate postoperative anatomy and indications for additional intervention. 1 B-NR

Therapeutic

For adults with symptoms of airway or esophageal compression and a vascular ring or PA sling, surgical repair* is reasonable to improve symptoms. 2a B-NR

For asymptomatic adults with a vascular ring or PA sling, surgical repair is not indicated. 3 No Benefit B-NR

*A cardiothoracic surgeon with expertise in ACHD should perform the repair.