

## Recommendations

1. Individuals with suspected AAOCA should undergo transthoracic echocardiography to identify the origin and course of the proximal coronary arteries. (*Class I, Level of Evidence B—supporting references*<sup>11,27,32,53-65</sup>)
2. Additional imaging studies, such as coronary CT angiography or cardiac MRI are reasonable to better visualize the coronary artery anatomy and to confirm the diagnosis. (*Class IIa, Level of Evidence B—supporting references*<sup>40,46,52,59,63,67,69-87,89-95,100</sup>)
3. In those individuals without a history of ischemic chest pain or aborted SCD, exercise stress testing combined with nuclear perfusion scan or echocardiographic imaging should be used to help assess the potential ischemic burden of the anatomic variant. (*Class I, Level of Evidence B—supporting references*<sup>27,32,40,98,99</sup>)
4. Cardiac catheterization should be performed in those individuals with anomalous origin of a coronary artery if the anatomy cannot be defined with noninvasive imaging, and in adults with risk factors for coexistent atherosclerotic coronary artery disease. (*Class I, Level of Evidence B—supporting references*<sup>1,33-52</sup>)

5. Individuals with AAOCA and symptoms of ischemic chest pain or syncope suspected to be due to ventricular arrhythmias, or a history of aborted SCD, should be activity restricted and offered surgery. (*Class 1; Level of Evidence B—supporting references*<sup>6,18,21-23,29,32,40,58,72,82,105,113,123,124</sup>)
6. Individuals with AAOCA and symptoms of ischemic chest pain or syncope suspected to be due to ventricular arrhythmias, or a history of aborted SCD, should be activity restricted and if deemed prohibitively high risk for surgery, catheter-based intervention may be considered. (*Class IIb; Level of Evidence C*)
7. Individuals with or without symptoms with an unrepaired anomalous origin of a left coronary artery from the right sinus of Valsalva, with an interarterial course, should be restricted from participation in all competitive sports. (*Class 1; Level of Evidence B—supporting references*<sup>6,18,21-23,29,105</sup>)
8. Individuals without symptoms with anomalous origin of a left coronary artery from the right sinus of Valsalva with an interarterial course should be offered surgery. (*Class 1; Level of Evidence B—supporting references*<sup>17-24</sup>)
9. Individuals with an anomalous origin of a right coronary artery from the left sinus of Valsalva should be evaluated for inducible ischemia, using an exercise stress test with additional imaging, including stress echocardiography or nuclear perfusion imaging. For those without symptoms concerning for ischemia or a positive exercise stress test, and after counseling concerning the risk of SCD, participation in competitive athletics is permissible. (*Class IIa; Level of Evidence C*)

10. Surgery for repair of AAOCA from the opposite sinus of Valsalva should include elimination of the intramural course and any associated ostial narrowing by unroofing, ostioplasty, or reimplantation. (*Class I; Level of Evidence B—supporting references*<sup>56,59,65,82,117,123</sup>)
11. Repositioning of the pulmonary artery confluence away from the anomalous artery (laterally or anteriorly) may be considered as an adjunctive procedure. (*Class IIb; Level of Evidence C*)
12. After surgical repair of an anomalous coronary artery, individuals without a history of aborted SCD should be offered the opportunity to return to competitive or recreational athletics after waiting at least 3 months after surgery, provided they have remained without symptoms concerning for ischemia or arrhythmia and an exercise stress test does not show evidence of myocardial ischemia or concerning arrhythmia. (*Class I; Level of Evidence C*)
13. After surgical repair of an anomalous coronary artery, in an individual who presented with aborted SCD, it

is reasonable to permit return to competitive athletics after a longer waiting period of 12 months after surgery, provided the patient has remained without symptoms concerning for ischemia or arrhythmia and an exercise stress test does not show evidence of myocardial ischemia or concerning arrhythmia. (*Class IIa; Level of Evidence C*)

14. After surgical repair of an anomalous coronary artery, in an individual who presented with aborted SCD, it is reasonable to permit return to recreational sports, including physical education class, 3 months after surgery, provided the patient has remained without symptoms concerning for ischemia or arrhythmia and an exercise stress test does not show evidence of myocardial ischemia or concerning arrhythmia. (*Class IIa; Level of Evidence C*)
15. An automated external defibrillator with trained personnel should be immediately available during competition and training. (*Class I; Level of Evidence B—supporting references*<sup>131-143</sup>)