

TABLE 2. Recommended follow-up for AAOCA, surgical repair

Postoperative short-term follow-up (timeline respective to surgical date)
• 7-10 d: Cardiology appointment with ECG and echocardiogram
• 4-6 wk: Cardiology appointment with ECG and echocardiogram
• 3 mo: Cardiology appointment with exercise stress test with imaging
• 6 mo: Cardiology appointment with ECG and cardiac MRI, if available
• Remain on 1 baby aspirin daily indefinitely
Postoperative long-term medical follow-up
• Cardiology follow-up annually
• ECG annually
• Echocardiogram annually
• Exercise stress test every 1-3 y, depending on activity level
• If participating in high-level recreational or competitive sports, exercise stress test annually
• With nuclear perfusion or stress echocardiogram only if new symptoms
• Holter monitor as needed, if symptoms
Physical activity
• No competitive athletics until at least 3 mo after surgery and after a complete evaluation has been performed by the pediatric cardiologist
• No competitive athletics for at least 12 mo after surgery and after a complete evaluation has been performed by the pediatric cardiologist in those whose presentation was sudden cardiac arrest/aborted sudden death
• Patients should be counseled that they are to cease exercise until further evaluation by the cardiologist if any symptoms develop, such as chest pain, palpitations, or fainting, during or just after exercise
• Patient should be allowed to rest if he or she gets tired
• Patient should stay well-hydrated with water, aiming for at least 60 ounces of water daily, or more, based on his or her activity level

AAOCA, Anomalous aortic origin of a coronary artery; ECG, electrocardiogram; MRI, magnetic resonance imaging.

SECTION IV: FOLLOW-UP

Ongoing Risk

The follow-up for patients with AAOCA will depend somewhat on the management chosen; however, all patients should be followed by a cardiologist for their lifetime. This should include a transition from a pediatric to an adult cardiologist, if necessary. Lifelong follow-up is especially important for those who undergo surgical repair, as long-term outcomes from surgical repair are largely unknown. Short- and mid-term complications have been noted, including mild aortic valve insufficiency, severe aortic valve insufficiency requiring aortic valve replacement, pericardial effusions, and ischemic changes on postoperative provocative testing.^{96,109,111,145} Postoperative death has been reported rarely in the literature in both the pediatric and adult age groups.^{82,146,147} The authors are aware of a handful of other pediatric deaths postoperatively that have not been published. It is interesting to note that, at least in the young, if a patient presents with SCD or SCA and survives the surgery, he or she may still be at increased risk for an SCD event once the patient returns to sport.¹⁴⁶

TABLE 3. Long-term follow-up and physical activity recommendations, unrestricted

Medical follow-up
• Cardiology follow-up annually
• Electrocardiogram annually
• Echocardiogram every 1-2 y
• Exercise stress test every 1-3 y, depending on activity level
• If participating in high-level recreational or competitive sports, exercise stress test annually
• With nuclear perfusion only if new symptoms
• Holter monitor as needed, if symptoms
Physical activity
• Full participation in competitive athletics
• Recommend automated external defibrillator at all team practices and sporting events
• Patients should be counseled that they are to cease exercise until further evaluation by a cardiologist if any symptoms develop, such as chest pain, palpitations, or fainting, during or just after exercise
• Patient should be allowed to rest if he or she gets tired
• Patient should stay well-hydrated with water, aiming for at least 60 ounces of water daily, or more, based on his or her activity level

Those who have undergone surgery will need close follow-up in the initial postoperative period, but will be able to lengthen the interval between visits after the first postoperative year. According to the recent AHA/ACC guidelines, these patients may return to competitive sports at least 3 months postoperatively and if an exercise stress test reveals no evidence of myocardial ischemia or ventricular arrhythmias.^{104,105} However, those who present with true aborted SCD appear to remain at increased risk, even after surgical repair.¹⁴⁶ We would recommend those

TABLE 4. Follow-up and activity recommendations for AAOCA, restricted from competitive athletics

Medical follow-up
• Cardiology follow-up annually
• Electrocardiogram annually
• Echocardiogram every 1-2 y
• Exercise stress test every 1-3 y, depending on activity level
• With nuclear perfusion only if new symptoms
• Holter monitor as needed, if symptoms
Physical activity
• No competitive sport at the middle school level or higher that requires moderate to vigorous activity levels (<40% maximum oxygen uptake and <20% maximal voluntary contraction)
• Examples include running, soccer, tennis, swimming, basketball, baseball, football
• Participation at the middle school level or higher is allowed in those sports that require low activity levels
• Examples include golf, bowling, cricket, archery, equestrian, karate
• Participation is allowed in recreational athletics, including gym class
• Patient should be allowed to rest if he or she gets tired
• Patient should stay well-hydrated with water, aiming for at least 60 ounces of water daily, or more, based on his or her activity level