



Dr. Shabib Alhadheri, MD
Advanced Pediatric Cardiology
Compassionate Care for Healthy Tomorrows

Anomalous Aortic Origin of a Coronary Artery (AAOCA)

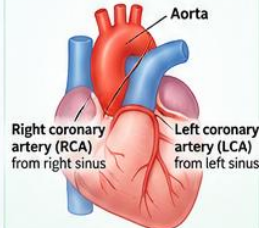
UNDERSTANDING THE CONDITION • GUIDING SAFE AND ACTIVE LIVES

What is AAOCA?

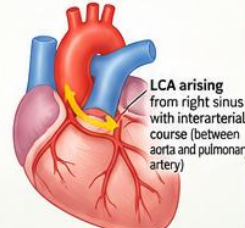
Anomalous aortic origin of a coronary artery (AAOCA) is a congenital heart condition where one of the coronary arteries arises from an abnormal location in the aorta rather than its usual position. In some cases, the abnormal artery follows a course between the aorta and pulmonary artery, which may reduce blood flow to the heart, especially during or after intense physical activity.

Normal vs. Anomalous Coronary Anatomy

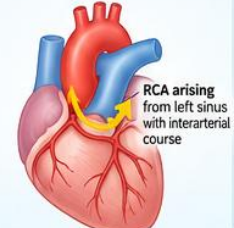
Normal Coronary Anatomy



Anomalous Left Coronary Artery (from right sinus – higher risk)

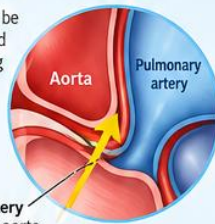


Anomalous Right Coronary Artery (from left sinus – more common)



Why Can AAOCA Be Concerning?

The abnormal coronary artery may be compressed between the aorta and pulmonary artery, especially during exercise, which can reduce blood flow to the heart muscle and increase the risk of myocardial ischemia and abnormal heart rhythms.



Coronary artery between the aorta and pulmonary artery

How Common is AAOCA?



Overall, AAOCA is rare.

Anomalous Right Coronary Artery (ARCA)
0.06% – 0.9%
(more common)

Anomalous Left Coronary Artery (ALCA)
0.025% – 0.15%
(less common)

Although ARCA is more common, ALCA has been responsible for up to 85% of AAOCA-related sudden cardiac deaths in some studies.

Symptoms

Many individuals have no symptoms. When symptoms occur, they often happen during or after vigorous exercise and may include:

- Chest pain
- Shortness of breath
- Heart palpitations
- Dizziness or fainting (syncope)
- In rare cases, the first sign can be sudden cardiac arrest.

How is AAOCA Diagnosed?

1



Echocardiogram
Initial test to look for abnormal coronary origins and evaluate heart function.

2



Cardiac CT or MRI
Provides detailed images of the coronary origin, course, and relationship to the aorta and pulmonary artery.

3



Exercise Stress Testing
Evaluates exercise capacity, heart rhythm, and symptoms.

4



Myocardial Perfusion Imaging
Stress echocardiogram, nuclear perfusion imaging, or stress cardiac MRI may be used to look for reduced blood flow to the heart.

5



Cardiac Catheterization (selected cases)
Rarely needed, but may provide additional information in complex cases.

Risk Factors for a Concerning Course



Anomalous left coronary artery from the right sinus (higher risk)



Interarterial course between the aorta and pulmonary artery



Intramural course within the aortic wall



Slit-like or narrowed coronary ostium



Proximal narrowing or acute-angle takeoff



Symptoms with exercise (chest pain, syncope)



Evidence of myocardial ischemia or ventricular arrhythmias

Evidence-Based Management Recommendations



Surgery is generally recommended for:

- Anomalous left coronary artery from the right sinus with interarterial/intramural course (even if asymptomatic)
- Symptomatic patients
- Evidence of myocardial ischemia or arrhythmias
- Other high-risk anatomic features



Non-surgical management may be considered for lower-risk anatomy, based on individual evaluation, symptoms, and testing results.



Exercise restriction is recommended for patients with high-risk anatomy, symptoms, or positive testing, especially while awaiting surgery.



Lifelong follow-up with a pediatric cardiologist is essential, even after successful surgical repair.



Safety Pearls

- ✓ Many individuals are asymptomatic, but serious events can occur.
- ✓ The risk of sudden cardiac events is highest during childhood, adolescence, and early adulthood.
- ✓ A normal stress test does not completely eliminate risk.
- ✓ Decisions about surgery and activity should be individualized based on anatomy, symptoms, and test results.
- ✓ Even after surgical repair, ongoing follow-up is important.
- ✓ Encourage a heart-healthy lifestyle and report new symptoms promptly.



Exercise and Sports

- Patients with high-risk anatomy or symptoms should avoid vigorous and competitive sports while completing evaluation or awaiting surgery.
- Some lower-risk patients may be able to participate in sports after careful evaluation and discussion with their cardiologist.
- After surgical repair, return to sports is considered only after an appropriate recovery period and reassurance with follow-up testing.
- Recommendations are individualized and should be discussed with your cardiologist team.



Family Screening & Support

- ✓ More than one affected child has been reported in some families.
- ✓ It is reasonable to consider echocardiogram screening for siblings to evaluate for coronary or other cardiac abnormalities.
- ✓ A diagnosis can be emotionally challenging. We encourage open discussion and provide support for patients and families.



Dr. Shabib Alhadheri, MD
Advanced Pediatric Cardiology



1478 W Elliot Rd, Suite 102
Gilbert, AZ 85233



480-855-1339





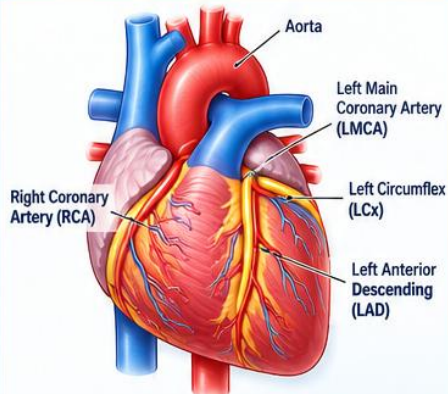
Understanding Congenital Coronary Artery Abnormalities

What Parents and Patients Need to Know



Congenital coronary artery abnormalities (CCAAs) are conditions present at birth that affect the origin, course, size, or structure of the coronary arteries (the blood vessels that supply blood to the heart muscle).

Normal Coronary Arteries



The heart is supplied by two main coronary arteries: right coronary artery (RCA) and left main coronary artery (LMCA), which divides into the left anterior descending (LAD) and left circumflex (LCx) arteries.

Common Types of Congenital Coronary Artery Abnormalities

1. Anomalous Aortic Origin of a Coronary Artery (AAOCA)



A coronary artery arises from the wrong aortic sinus, which may travel between the aorta and pulmonary artery (interarterial course) and can be associated with ischemia, especially during exercise.

2. Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA)



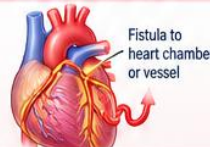
The left coronary artery arises from the pulmonary artery instead of the aorta. This can cause heart failure in infancy and requires surgical repair.

3. Anomalous Right Coronary Artery from the Pulmonary Artery (ARCAPA)



The right coronary artery arises from the pulmonary artery. Often diagnosed later in childhood or adulthood.

4. Coronary Artery Fistulas



An abnormal connection between a coronary artery and a heart chamber or major vessel.

5. Single Coronary Artery



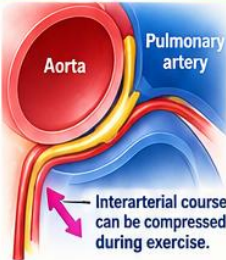
Only one coronary artery originates from the aorta and supplies the entire heart.

6. Coronary Artery Atresia or Hypoplasia



Underdevelopment or absence of a coronary artery segment. May cause ischemia or ventricular dysfunction.

Why Are Some Anomalies Dangerous?



High-risk features:

- Interarterial course
- Intramural aortic segment
- Slit-like coronary ostium
- Acute-angle takeoff
- Dynamic compression during exercise

Symptoms



Infants

- Poor feeding
- Excessive sweating
- Rapid breathing
- Failure to thrive
- Heart failure



Older Children and Adolescents

- Chest pain during exercise
- Dizziness
- Syncope (fainting)
- Palpitations
- Exercise intolerance
- Unexplained cardiac arrest

Diagnosis



Echocardiography

First-line imaging study in children.



Cardiac CT (CCTA)

Best visualization of coronary origins and vessel course.



Cardiac MRI

Ventricular function and myocardial scarring.



ECG



Holter monitor



Exercise testing

Nuclear imaging (in selected cases)

Treatment



Observation

- For benign variants
- Asymptomatic low-risk anomalies



Catheter-Based Closure

- Used for selected coronary fistulas.



Surgical Repair

- AAOCA: unroofing, reimplantation, ostioplasty
- ALCAPA: coronary reimplantation
- Large fistulas: surgical ligation or transcatheter closure

Sports and Activity Recommendations



Generally Allowed



Benign coronary variants without evidence of ischemia.



Restricted Until Evaluation/Treatment

- AAOCA with interarterial course
- Exercise-induced symptoms
- Evidence of ischemia
- Prior aborted sudden cardiac arrest

Competitive sports participation should follow comprehensive evaluation by a pediatric congenital cardiologist and exercise testing.

Long-Term Follow-Up



Patients may require lifelong surveillance for:

- Coronary artery patency
- Ventricular function
- Myocardial ischemia
- Arrhythmias
- Exercise tolerance

Follow-up intervals depend on the specific anomaly and treatment history.

Prognosis



Excellent Prognosis

- Benign coronary variants
- Successfully repaired fistulas
- Treated ALCAPA with preserved ventricular function



Higher-Risk Conditions

- Left AAOCA with interarterial course
- Untreated ALCAPA
- Significant ischemia
- Previous ventricular arrhythmias



Key Take-Home Points

- ✓ Coronary artery abnormalities range from harmless variants to life-threatening conditions.
- ✓ Most important lesions: AAOCA, ALCAPA, ARCAPA, and coronary fistulas.
- ✓ Exercise-related chest pain, syncope, or unexplained cardiac arrest should prompt evaluation.
- ✓ Echocardiography is the initial test, but cardiac CT often provides the best anatomical definition.
- ✓ Early recognition and appropriate intervention can improve outcomes and reduce the risk of sudden cardiac death.



Dr. Shabib Alhadheri, MD
Advanced Pediatric Cardiology



1478 W Elliot Rd, Suite 102
Gilbert, AZ 85233



480-855-1339



www.apcardio.com