

MINI REVIEW

Anomalous aortic origin of coronary artery: For a challenging diagnosis, a transthoracic echocardiogram is recommended

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Abstract

Anomalous aortic origin of a coronary artery (AAOCA), especially the interarterial course of the right or left coronary artery, predisposes paediatric patients to myocardial ischaemia. This rare condition is a leading cause of sudden cardiac death. General paediatricians face challenges when diagnosing this anomaly, and they should pay particular attention to the recurrence of exercise-related syncope without prodromal symptoms, chest pain and dyspnoea. An accurate transthoracic echocardiogram with Doppler colour flow mapping is the best method to use to identify AAOCA.

Conclusion: Identifying an AAOCA is challenging, and we provide advice on clinical red flags and diagnostic approaches for general paediatricians.

KEYWORDS

anomalous origin, coronary artery, exertional syncope, sudden cardiac death, transthoracic echocardiogram

1 | BACKGROUND

Anomalous aortic origin of a coronary artery (AAOCA) is when both coronary arteries arise from the same aortic sinus, a single ostium or two separate ostia. It is called anomalous left coronary artery from the right sinus (ALCA-R), when the left main coronary artery arises from Valsalva's right sinus and ARCA-L when the right coronary artery originates from the left sinus. Apart from the origin, anomalous coronary can develop differently. AAOCA can be characterised by one of five-course subtypes: interarterial, subpulmonic (intraconal or intraseptal), prepulmonic, retroaortic or retrocardiac. Whilst most coronary anomalies are benign, the two most common courses predisposing to myocardial ischaemia and sudden cardiac death (SCD) in young people are the anomalous right interarterial coronary artery and the anomalous left interarterial coronary artery. In both

conditions, the anomalous coronary artery passes between the aortic root and the pulmonary trunk. The interarterial vessel is usually intramural, travelling in the aorta wall with an acute-angle take-off of the anomalous vessel ostium. These features determine the blood flow reduction to the myocardium during physical effort and the development of ventricular arrhythmia (Figure 1).¹ However, how this occurs is still a matter of conjecture as patients who suddenly died have usually exercised numerous times at the same or greater level and duration without any symptoms: this evidence suggests that whatever the mechanism of sudden coronary obstruction may be, it must be intermittent and unpredictable.^{2,3} Moreover, electrocardiogram (ECG), exercise study, or echocardiography before or after a resuscitated episode of SCD are usually negative.⁴

Although AAOCA is a leading cause of SCD, general paediatricians face many challenges when caring for these patients. We will

Abbreviations: AAOCA, anomalous aortic origin of a coronary artery; ALCA-R, anomalous left coronary artery from right sinus; ARCA-L, anomalous right coronary artery from left sinus; ECG, electrocardiogram; SCD, sudden cardiac death.

Tessitore Antimo and Caiffa Thomas contributed equally to this work.

[Correction added on 14 May 2022, after first online publication: CRUI-CARE funding statement has been added.]

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not deal with the anomalous origin of the left coronary artery from the pulmonary artery, an infants' disease characterised by heart failure in the first months of life.

2 | EPIDEMIOLOGY

Several studies have attempted to quantify the prevalence of congenital coronary anomalies, with a significant difference in prevalence rates between 0.1% and 1.15% in adult and paediatric populations.⁵⁻⁸ The difference in prevalence rates can be explained by choice of imaging modality (invasive coronary angiography, transthoracic echocardiography, coronary computed tomography angiography, or magnetic resonance angiography) and patient population (symptomatic or asymptomatic, populations of athletic or non-athletic persons). The anomalous aortic origin of the coronary artery accounts for 14% of all cardiac-related deaths in young athletes.^{9,10}

Key Notes

- Anomalous aortic origin of a coronary artery, especially the interarterial course of the right or left coronary artery, predisposes patients to myocardial ischaemia and sudden cardiac death.
- The red flags that general paediatricians should pay attention to are the absence of prodromal symptoms and the occurrence of syncope during physical exercise.
- Transthoracic echocardiogram with Doppler colour flow mapping can identify the origin and course of the coronary arteries.

Interarterial ARCA-L is from three to six times more common than ALCA-R, with SCD occurring more frequently in patients with ALCA-R.^{2,3}

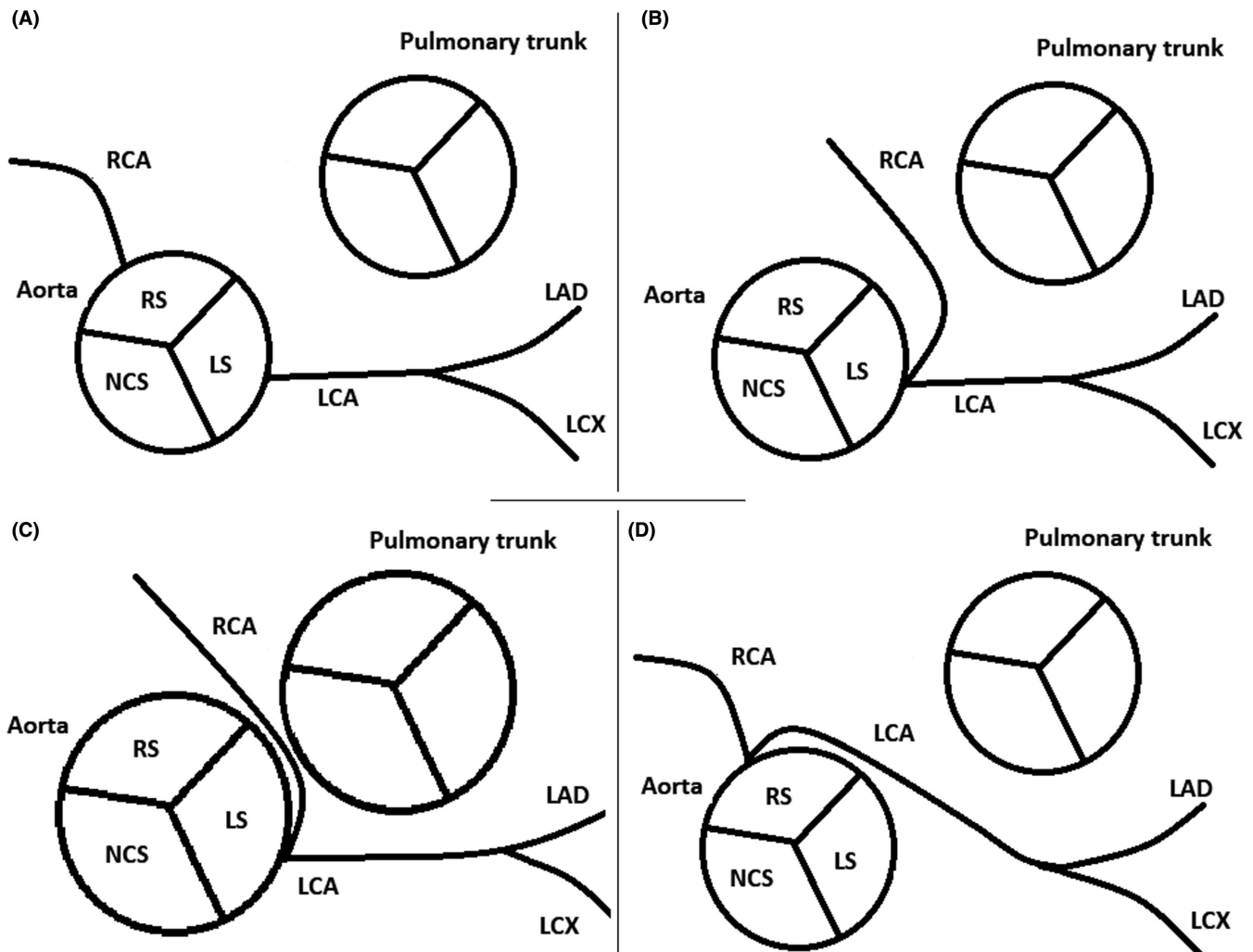


FIGURE 1 (A) A standard origin of coronary arteries. (B) An anomalous origin of the right coronary artery. (C) During physical effort, the aortic root and pulmonary trunk expansion compress the anomalous artery passing through. (D) An anomalous origin of the left coronary artery. LAD, left anterior descending artery; LCA, left coronary artery; LCX, left circumflex artery; LS, left sinus; NCS, non-coronary sinus; RCA, right coronary artery; RS, right sinus

3 | CLINICAL SUSPECT AND PRESENTATION

There is no typical presentation of patients with AAOCA. Patients are frequently diagnosed in the absence of symptoms, when the anomaly is found incidentally on an echocardiogram or computed tomography angiogram performed for another reason, such as a heart murmur or an abnormal electrocardiogram. When symptoms are present, they are usually related to exertion and comprehend chest pain, palpitations, shortness of breath, dizziness or syncope on exertion.¹¹ An episode of aborted SCD or SCD might also be the initial event. According to one study,⁴ only 36% of the cases presenting with AAOCA related SCD had symptoms before the event. Another investigation¹² reported that 52% of military recruits with AAOCA who presented with SCD had previous symptoms of syncope, chest pain, and dyspnoea.

Chest pain is a common complaint in general paediatric clinics, emergency departments and paediatric cardiology clinics. In a study cohort of 3700 children, a cardiac cause was detected in 37 patients (1%), amongst whom two patients had anomalous right coronary arteries from the left sinus with chest pain at rest.¹³ Chest pain should be considered ischaemic if accompanied by evidence of myocardial injury, such as ST-segment depression on ECG, ventricular arrhythmias, blood pressure not increasing or decreasing with exercise, hypokinesia or akinesia of heart walls on echocardiography, lack of perfusion on a nuclear scan, or evidence of past fibrosis or late gadolinium enhancement by cardiac magnetic resonance imaging.¹¹ Syncope is also a widespread disorder with a bimodal incidence in children and adolescents, peaking in females aged 15–19 years. Benign aetiologies are the most common cause of syncope in children, with neurocardiogenic syncope accounting for up to 75%–80% of cases. Typical features of neurocardiogenic syncope are prominent prodromal symptoms, including nausea, dizziness and tunnel vision, along with pallor and brief loss of consciousness.¹⁴ Remarkably, these prodromal symptoms are absent in cardiogenic syncopes, such as those due to anomalies of coronary artery origin and channelopathies.¹⁵

4 | DIAGNOSIS

A screening ECG is unreliable for suspecting or recognising AAOCA, and stress tests do not help identify these abnormalities early, before an adverse event occurs.^{4,12}

Cardiac catheterisation with coronary angiography, due to the invasive nature of the test and its inherent exposure to ionising radiation, is rarely used in the paediatric population. The best method for initially identifying AAOCA is a transthoracic echocardiogram with Doppler colour flow mapping to characterise the origin and course of the coronary arteries.^{16,17} Coronary computed tomography angiography or cardiac magnetic resonance imaging is commonly employed to visualise the coronary artery anatomy better to confirm the diagnosis.¹⁸ Once the anatomy has been established, a maximal exercise stress test with ECG is performed to assess the ischaemic potential of the anatomic variant, especially in competitive athletes.¹⁹ A nuclear perfusion test can also be

considered. Remarkably a regular stress test is only partially reassuring due to its low negative predictive value, being diriment only if positive.

5 | MANAGEMENT

It is generally accepted that the activity restriction and surgery are indicated for any patient with AAOCA who has signs or symptoms of myocardial ischaemia or ischaemic changes inducible with exercise testing. However, the treatment dilemma arises when this diagnosis is made in the asymptomatic patient, particularly with ARCA-L. In this case, the indications recommending surgery depend on the anatomy of the coronary artery: high-risk anatomies, like high orifice, ostial stenosis, fish-mouth-shaped orifice, acute-angle take-off, intramural or interarterial course, and hypoplasia of the proximal coronary artery, has been associated with myocardial ischaemia and has been proposed as indications for surgical correction.

The clinical suggestion for intervention is based on the calculated risk of SCD. Most children over 10 years of age with interarterial ALCA-R, even if asymptomatic, are referred for surgery due to their increased risk of exertional SCD.²⁰ On the other hand, there is less agreement that patients with ARCA-L without symptoms should undergo the operation. In addition, ALCA-R with intraseptal, prepulmonic, or retroaortic courses is generally considered benign with only rare case reports of ischaemia. Therefore, these patients are generally not referred for surgery and have no exercise restriction but must perform a stress test before admission to competitive athletics to rule out inducible ischaemic or syncope events.

There are several surgical approaches based on patient characteristics and coronary anatomy. Even though surgical procedures may remove one of the hypothesised mechanisms of ischaemia leading to SCD, the long-term impact of the surgical procedure on the coronary arteries is unknown. In particular, it is unrevealed whether these procedures place the patient at long-term risk of coronary stenosis due to scarring or accelerated atherosclerosis.¹¹

When surgery is not considered, exercise restriction from competitive athletics is the most common medical strategy. The recommendation to avoid competitive athletics in AAOCA patients depends on the high mortality rate associated with this anomaly and the evidence that SCD usually occurs during or after peak exercise.^{2,10} Despite this, asymptomatic children are encouraged to participate in physical education classes and other recreational activities. Contrary to previous recommendations, the current guidance of the American Heart Association/American College of Cardiology allows individuals with unrepaired asymptomatic and negative exercise stress test ARCA-L to participate in competitive sports.²¹

Cardiology follow-up for patients with AAOCA depends on the strategy chosen and is lifelong, particularly for those undergoing surgical repair, whose long-term results are mainly unknown. Nevertheless, short- and mid-term complications have been noted, including mild aortic valve insufficiency, severe aortic valve insufficiency requiring valve replacement, pericardial effusions, and ischaemic changes on provocative postoperative testing.^{22,23}

According to the recent American Heart Association/American College of Cardiology guidelines, patients who received corrective surgery may return to competitive sports three months after surgery and only if an exercise stress test reveals no signs of myocardial ischaemia or ventricular arrhythmias.²¹ However, those presenting with aborted SCD appear to remain at increased risk, even after surgical repair.²⁴ For these patients, a study¹¹ recommends a return to competitive sports after one year and only if with no symptoms and with a negative exercise stress test. Indeed, whether a patient with a previous surgery should return to competitive sport or not is still the subject of debate, and each patient must be analysed based on his or her history. The evaluation for the issue of the sports medical certificates must preferably be managed by a sports medicine physician expert in coronary anomalies in collaboration with the cardiac surgery centre that performed the corrective surgery.

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CONFLICTS OF INTEREST

The authors declare that there is no conflict of interest.

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