# Anomalous Aortic Origin of a Coronary Artery is Always a Surgical Disease

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**Anomalous aortic origin of a coronary artery is a congenital anomaly in which a major coronary artery arises from the wrong sinus of Valsalva (left coronary from right sinus or right coronary from left sinus) and courses between the great arteries before reaching its normal epicardial course. Although the risk of sudden death is clearly established, the indications for surgery remain controversial. The risk of sudden death is increased in symptomatic patients, in anomalous left coronary artery, as well as in the presence of some risk factors (intense physical activity, young age [<35 years], aggravating anatomical features [intramural interarterial course, slit-like ostium, acute angle of take-off]). As far as is currently known, surgical management using an anatomical repair can prevent sudden death, provided that normal coronary anatomy and function are achieved and that extensive return-to-play testing is performed. A precise evaluation of the benefit/risk ratio is mandatory on an individual basis, but surgery may be indicated in the vast majority of patients.**
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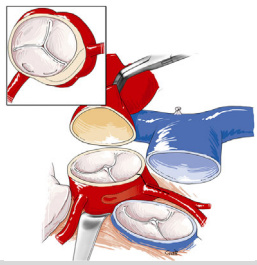
## Introduction

Anomalous aortic origin of a coronary artery (AAOCA) is a congenital anomaly in which a major coronary artery arises from the wrong sinus of Valsalva and courses between the great arteries before reaching its normal epicardial course. Although the incidence is difficult to ascertain, it is estimated to be in the range of 0.1% to 0.3% of the general population. The pathophysiology is elucidated badly; however, it is clearly established that such anomalies can be associated with an increased risk of sudden death and cardiac morbidity, particularly in young athletes during or after strenuous exercise. The diagnosis is, more and more often, made during a routine systematic cardiac evaluation. The only effective treatment is surgical, and various techniques have been described.

Management of AAOCA remains controversial. Surgical intervention is generally warranted in patients with symptoms suggestive of myocardial ischemia and in patients with anomalous left coronary artery (ALCA). Less clear is the optimal treatment of asymptomatic patients, particularly those with anomalous right coronary artery (ARCA). The prevalence and natural history of this anomaly are not well established.



Anatomical repair of anomalous aortic origin of a coronary artery.

**Central Message**

Anatomical repair may prevent sudden cardiac death in young patients with anomalous aortic origin of a coronary artery. Surgery is (nearly) always indicated.

Surgical results are reported from small series with relatively short follow-up. In the present lack of established evidence-based data, the treatment should be individualized and the benefit/risk ratio should be evaluated for every patient.

## Surgical Treatment

### Surgical Techniques

Various surgical techniques have been used, including coronary bypass grafting, pulmonary translocation, and

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coronary reimplantation. More recently, unroofing of the anomalous vessel along its intramural segment has become the preferred management option. Even if the pathophysiology of the anomaly is not completely understood, some aggravating anatomical factors have been identified: 1) intramural course with stenosis, particularly at the level of the valvar commissure; 2) slit-like deformation of the coronary ostium with potential flap closure; and 3) acute (non-orthogonal) angle of take-off with potential kinking of the coronary artery as it actually exits from the aorta. Optimal surgical repair should address all these anatomic features. This is not the case with the usual operations. Particularly, in the unroofing procedure, the more distal part of the intramural segment, at the point where the coronary artery actually leaves the aortic wall, is left intact and potentially stenotic.

Karl[1] proposed a more physiological approach by opening and patching the abnormal segment from the ostium down to the normal epicardial artery. The procedure takes into account most of the potential mechanisms that can generate ischemia. It enlarges a slit-like ostium, augments the diameter of the proximal coronary, and improves the acute angulation at take-off. However, the intervascular course between the aorta and the pulmonary artery is left in place, thus justifying the need for associated pulmonary translocation.

We described a modification of Karl's technique that provides a more "anatomical" repair, by creating a new enlarged coronary ostium in the appropriate sinus.[2] This technique

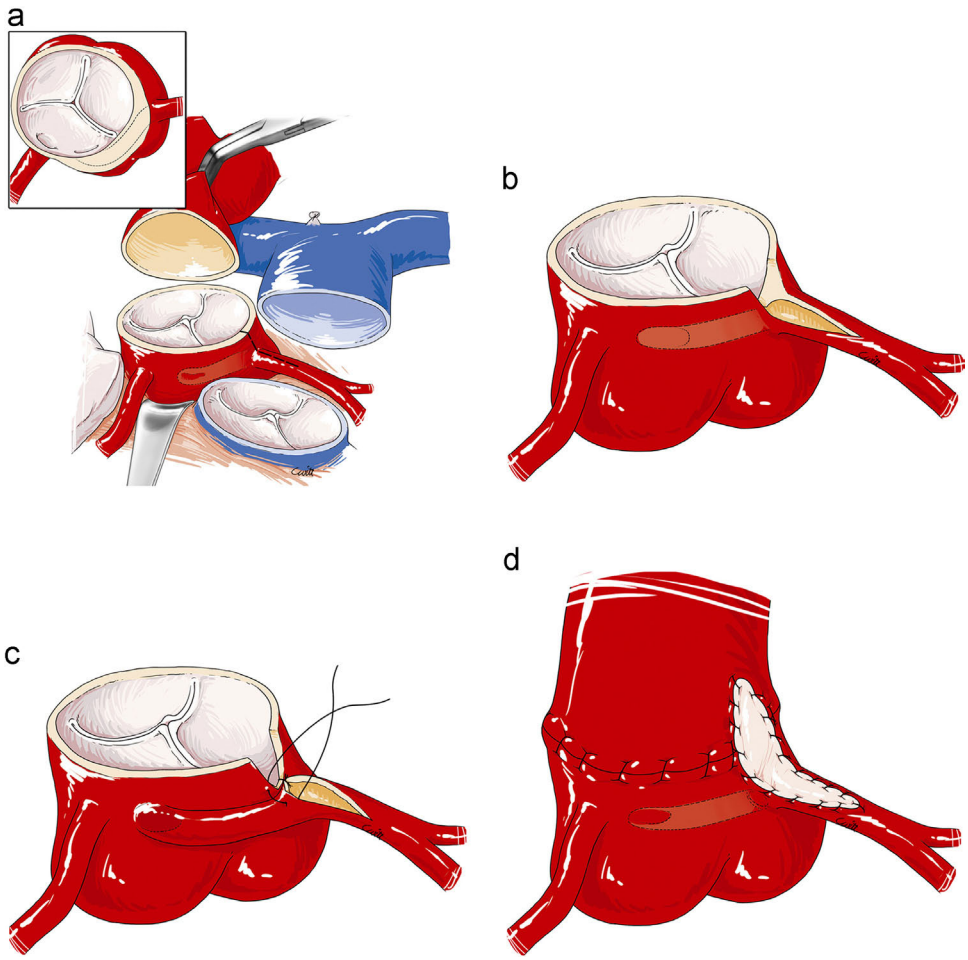


**Figure 1** "Anatomical" repair of AAOCA. (A) The left coronary ostium arises from the right sinus; the initial course of the left coronary artery is intramural, within the aortic wall; the left coronary artery reaches a normal epicardial course, opposite the left sinus (see inset). The pulmonary trunk is divided to expose the epicardial course of the left coronary artery. The aorta is transected at the level of the sinotubular junction. The initial epicardial course of the left coronary artery in incised longitudinally. Beginning from the cut edge of the aorta, a vertical incision is made in the left sinus towards the incised epicardial left coronary artery. (B) The two incisions, aortic and coronary, are joined together at the point where the coronary artery leaves its intramural course to become epicardial. The intramural segment of the coronary artery is, thus, left intact but bypassed. (C) When the abnormal left coronary artery courses between the great arteries, without an intramural course, the coronary and aortic incisions must be approximated with a few interrupted sutures. (D) A patch of fresh autologous pericardium is sutured into the aorto-coronary incision to create a new coronary ostium in the appropriate left sinus. The patch is incorporated into the aortic suture line. (Reprinted with permission from Gaudin et al. Anomalous aortic origin of coronary arteries : " anatomical " surgical repair. Multimed Man Cardiothorac Surg 2014;2014:mmt022.[2])

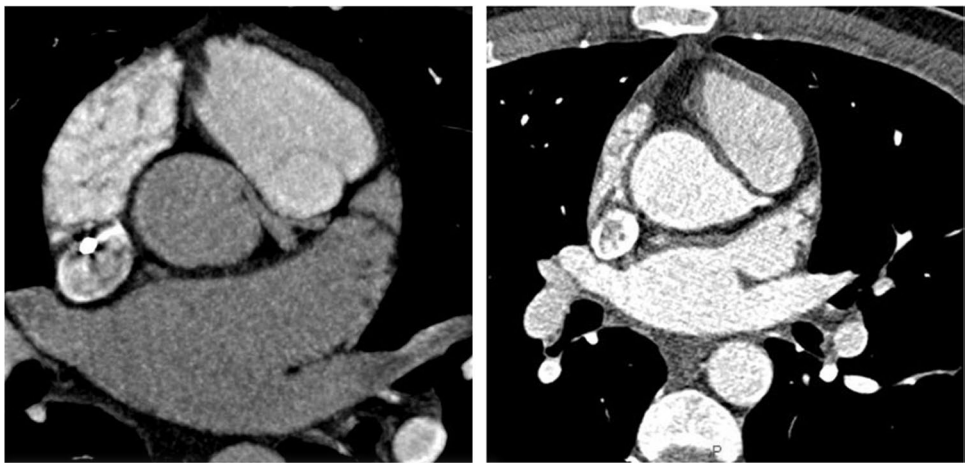

**Figure 2** Preoperative and postoperative coronary computed tomography. (Left) Preoperative: the *arrow* indicates the left coronary artery arising from the right aortic sinus and coursing between the aorta and the pulmonary artery. (Right) Postoperative: after "anatomical" repair, a new left coronary ostium is created in the appropriate sinus with a normal angle of take-off and a complete disappearance of the interarterial abnormal course.

creates a large coronary ostium, completely eliminates the intramural segment, and restores a normal angle of take-off.

## "Anatomical" repair

The anatomical repair is described for an anomalous origin of the left coronary artery from the right sinus (Fig. 1). The same technique can be transposed for an ARCA.

After aortic clamping and induction of cardioplegic arrest, the ascending aorta is divided at the level of the sinotubular junction. The anomalous coronary ostium and the abnormal intramural segment are identified. The aortic and pulmonary roots are separated from each other down to the annular level, to get a perfect exposure of the epicardial course of the anomalous coronary artery; in most cases this step is greatly facilitated by the division of the main pulmonary trunk. The normal epicardial course of the left coronary artery, opposite its appropriate sinus, is clearly identified by removing all surrounding connective tissue. The left main coronary artery is opened longitudinally. The incision is begun distally, close to the coronary bifurcation, and carried out upstream toward the aortic wall, at the point where the artery leaves the aorta after its intramural course.

Beginning from the cut edge of the aorta, a vertical incision is made in the left sinus to reach the coronary incision. The two incisions are joined together, exactly at the point where the coronary artery leaves the aortic wall. In rarer cases in which the abnormal interarterial coronary artery has no intramural segment, the coronary and aortic incisions must be approximated together, using a few interrupted sutures. A patch (fresh autologous pericardium) is implanted to create a new large left coronary ostium. The abnormal intramural segment is therefore left intact. The aortic anastomosis is completed, incorporating the top edge of the patch into the anastomotic suture line. When the pulmonary trunk has been transected, it is reconstructed after extensive mobilization of the pulmonary branches. In all cases, the absence of any residual compression of the reconstructed coronary artery by the pulmonary artery is ascertained.

A similar technique is used for ARCA from the left sinus. Division of the main pulmonary artery is not necessary.

## Results of "Anatomical" Repair

Between 2005 and 2014, 19 patients underwent "anatomical" repair for ALCA (seven patients) or ARCA (12 patients). Mean age was 14 years (range, 5 to 37 years). Symptoms (present in 13 patients; 68%) included resuscitated sudden cardiac death (five patients), chest pain (five patients), syncope (two patients) and myocardial infarction (one patient). No symptoms were present in six cases of serendipitous diagnosis. It is noteworthy that three patients with ARCA (25%) had aborted sudden death.

There was no early and late mortality. All patients had patent coronary flow by Doppler and normal echocardiography after a mean follow-up of 4.8 years. Coronary computed tomography was performed in 10 cases and showed a patent coronary artery in all (Fig. 2). One patient underwent reoperation at 6 months for a progressive dilatation of the coronary patch. No patient had activity restrictions. None had evidence of residual myocardial ischemia at stress test.

## Critical Appraisal of Surgical Treatment

Excellent early and late results have been reported using various techniques. Coronary transfer and reimplantation into the appropriate sinus is a satisfactory technique that achieves anatomical repair in anomalies without an intramural course.[3] In the more frequent patients with an intramural course, the unroofing procedure is usually advocated. Excellent results have been reported.[3,4] However, a single study reported a significant incidence of subclinical and inconsistently reproducible evidence of myocardial ischemia after the unroofing

procedure.[5] This prompted the search for more anatomical repairs.[1,2,6].

Most sudden deaths occur during or immediately after strenuous exercise. Even if the exact mechanism is not fully understood, it is likely that acute myocardial ischemia under stress plays a major role. It is therefore essential for the surgical repair to address all the anatomic features that can be incriminated, including presence of interarterial course, slit-like deformation of the ostium, stenosis of the intramural segment, and acute angle of take-off. We believe that this is the case with the "anatomical" repair that we use. After surgery, extensive evaluation must be performed to ascertain the normality of coronary anatomy and to rule out residual ischemia under stress before return to normal physical activity can be recommended. Although more data are needed, it is likely that sudden death at exertion is prevented by adequate and successful surgical repair. The forthcoming results of a multicenter study will provide essential information.[7]

In the current literature a single case of sudden death at exertion after a successful surgical operation and despite extensive return-to-play testing has been reported.[8] There are also a few reports of sudden death at rest in unoperated patients.[9] It is hypothesized that cumulative episodes of myocardial ischemia lead to patchy myocardial necrosis and fibrosis that can create the substrate for lethal ventricular arrhythmias. This theory is supported by the histologic evidence of chronic ischemic changes in areas of myocardium supplied by the anomalous artery.[9] It is clear that surgical repair, even optimal, cannot prevent such events.

## Risk of Sudden Death with AAOCA

There are a few hard data: 1) AAOCA can cause sudden death; and 2) AAOCA represents one of the main causes of cardiac death in young athletes.[9,10] There are, above all, many uncertainties.[11]

The true prevalence of the condition is not known. However, the anomaly is not rare. It is generally accepted that incidence ranges from 0.1% to 0.3% of the population, with ARCA being six to 10 times more frequent that ALCA. Diagnosis can be ascertained by careful echocardiographic examination, performed with special attention to the origin of the coronary arteries. Most patients (>50%) are asymptomatic. Diagnosis is typically made as an incidental finding, during a cardiac evaluation performed for another indication or during a systematic screening, particularly in young athletes.

The risk of sudden death is difficult to determine. It is higher in patients with ALCA, but it does exist in patients with ARCA. The true risk cannot be estimated from autopsy data, as it has been too often reported. Penalver and colleagues[11] provided a critical review of the data observed in large cohorts of military recruits and young athletes. From this analysis, it can be estimated that the risk of sudden death is high in ALCA (2% to 5%), but much lower in ARCA (0.1% to 0.2%). From the same data, incremental risk factors can be identified: intense physical activity, young age (<35 years), and aggravating anatomical features (intramural interarterial course, slit-like ostium, acute angle of take-off). Both the prevalence and the natural history of AAOCA remain to be clearly delineated.

# Conclusions: Suggested Recommendations

In the most recent guidelines for the management of adults with congenital heart disease, the American College of Cardiology/American Heart Association task force recommended surgical treatment in patients with ALCA and in patients with AAOCA (left or right) and documented coronary ischemia (symptoms and/or evidence of inducible ischemia). On the other hand, it is stated that a conservative approach is reasonable in patients with ARCA with no evidence of myocardial ischemia; it is even recommended that such patients may participate in athletics, provided appropriate counseling of the athlete and/or the athlete's parent(s) is given.[12]

This creates conflicting situations for both patients and care providers. ARCA is, by far, more frequent than ALCA; most patients with ARCA have no symptoms and no inducible ischemia. The risk of sudden death is probably low, even very low, but it is not absent. In our own small series, there were three patients with ARCA who did not experience previous symptoms and who had aborted sudden death. More and more often diagnosis is made as an incidental finding during a routine cardiac evaluation using echocardiography. Once the diagnosis is made, the patient must face major consequences. Recreational activities that approach the level of exertion (such as that seen in organized sports) are restricted. Participation in competitive sports is impossible because a sport license cannot be delivered. High-level professional athletes must stop their activities and completely change their life goals. All other patients must modify their way of life and accept life with a permanent risk of sudden death at physical exertion. This may be difficult to bear for many patients. Particularly in adolescents and young adults, behavior and psychosocial development may be severely impaired. The only solution to restore a normal quality of life is surgical treatment, which can be performed with an extremely low risk. As far as is currently known, surgical management can prevent sudden death, provided that normal coronary anatomy and function are achieved and that extensive return-to-play testing is performed.

To conclude, we believe that AAOCA is always (or nearly always) a surgical disease. Surgery is indicated: 1) in all patients with ALCA; 2) in all patients with anomalous origin of the left anterior descending artery; 3) in patients with ARCA with symptoms and/or inducible ischemia; 4) in patients with ARCA without symptoms and/or inducible ischemia, but with potentially dangerous anatomical features (slit-like orifice, stenosis of intramural segment, or documented flow restriction via intravascular ultrasound); and 5) in asymptomatic patients with ARCA who cannot accept the behavioral and psychosocial consequences of the diagnosis and who wish a completely normal quality of life. The surgical procedure must address all the anatomical features of the anomaly and normal coronary

anatomy must be restored. We believe there is no place for percutaneous stenting of the anomalous artery, a procedure that leaves in place most of the anatomical risk factors.[13] Extensive postoperative evaluation must reveal no exercise-induced ischemia, ventricular arrhythmia, or ventricular dysfunction during maximal exertion.

A conservative approach appears to be a valuable option in only a small minority of patients. This includes, in our opinion, asymptomatic patients with ARCA, without any evidence of inducible ischemia and/or aggravating anatomical features who are over the age of 40 to 50 years and who can accept limitations in their physical activities.[14]

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