Coronary artery anomalies – a short review

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Abstract: Coronary artery anomalies are rare but sometimes important findings in the evaluation of the coronary artery. The majority of the anomalies are of benign prognosis, but others can be associated with cardiac symptoms and syndromes (angina, dyspnea, syncope, congestive heart failure, myocardial infarction and sudden death). The potentially serious anomalies include: ectopic coronary origin from the pulmonary artery; ectopic coronary origin from the opposite aortic sinus; and large coronary fistulae. Appropriate diagnosis is critical for recognition and management. Treatment can be performed by surgical or percutaneous approach. However, management is conservative in the majority of them. Contemporary diagnosis and clinical management of these anomalies are briefly reviewed and discussed in this article.

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1 Introduction

Coronary artery anomalies are abnormalities in the origin, course or distribution of the coronary arterial circulation. There are various definitions of coronary anomalies. The most comprehensive of them is that proposed by Angelini [1]. Normal is any morphological feature observed in more than 1% of an unselected population and anomaly is a morphological feature seen in less than 1% of that population. According to this definition, coronary anomalies affect approximately 1.3% of the population undergoing coronary angiography [2] and 0.3% of autopsies [3]. The true incidence is unknown. 87% of these have anomalies of origin and course and 13% have coronary artery fistulae. The majority of anomalies are of benign prognosis; others, however, can cause cardiovascular morbidity.

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and mortality. These anomalies have a high clinical profile and they are the second most common cause of sudden cardiac death related to sports, behind hypertrophic cardiomyopathy. These anomalies account for approximately 12% of such cases, compared with 1.2% of sudden death cases not related to sports (which is similar to the background prevalence in the population) [4].

2 Anatomy, pathology and classification

Normally, the left coronary artery (LCA) arises from the left aortic sinus and divides into the left anterior descending artery (LAD) and the left circumflex artery (Cx) after a short length known as the left main artery (LM). Typically (85% of cases), the right coronary artery (RCA) arises from the right sinus as the “dominant” artery of the heart, defined as the artery of origin for the posterior descending artery. LCA is the “dominant” artery (the posterior descending artery branches from the Cx) in 10% of cases, whereas the remaining 5% cases have “co-dominant” circulation (the posterior descending artery branches from both the RCA and Cx). If the aortic valve is bicuspid, the origins of both arteries are in the typical site as in the tricuspid valve.

Despite the occasional association of coronary anomalies with other congenital heart defects, there is no known genetic predisposition to coronary artery anomalies. The most often associated congenital defects are mitral valve prolaps, bicuspid aortic valve, tetralogy of Fallot, corrected and uncorrected transposition of the great arteries, univentricular heart and aortic coarctation [5].

Various schemes of classification of coronary anomalies have been proposed. We prefer the classification based on origin, course and distribution of the arteries. The division between benign and potentially malignant anomalies is clinically relevant. The potentially serious anomalies include: ectopic coronary origin from the pulmonary artery; ectopic coronary origin from the opposite aortic sinus; and large coronary fistulas. This article is about these anomalies.

3 Clinical presentations and diagnosis

Despite the majority of the anomalies being asymptomatic, others can be associated with serious cardiac symptoms and clinical syndromes. Cardiac symptoms include angina, shortness of breath and syncope. It is very important that clinical syndromes are serious ones, such as myocardial infarction, heart failure and sudden cardiac death. The main problem is low sensitivity and low specificity of these symptoms and syndromes. Only one third of anomalies had clinical manifestations, while the majority (62%) of cases of sudden death was previously asymptomatic [6]. Physical examination can reveal systolic murmurs, which can be associated with large fistulas and abnormal origin of the coronary artery from the pulmonary artery with large left-to-right shunt.

In the last few years we can see the interesting challenge in diagnostics of coronary artery anomalies. Multidetector row computer tomography and magnetic resonance imag-
ing have rapidly evolved from the research setting to become useful clinical methods [7, 8]. These methods allow acquisition of images of a whole heart and subsequent 3-dimensional reconstruction of the arteries and heart structures (Figure 1, 2). The relation between anomalous coronary arteries and heart structures is very important for good management of patients; in addition these methods are non-invasive. We suppose that this is an optimal method in the hands of an experienced team with good technical support.

![Figure 1](image1.png)

**Fig. 1** Two-dimensional view of the heart at level of proximal right coronary artery showing the course between aorta and pulmonary artery.

On the other hand, because anomalies are mostly accidental findings during coronary angiography, this method is still the “gold standard” for the evaluation [9]. In the majority of cases, coronary angiography is still sufficient and further evaluation is not needed. There are several angiographic signs to distinguish them and we can use the pulmonary catheter during the coronary angiography to perform ventriculography or aortography [10].

Echocardiography may also provide useful information for diagnosis, especially as a screening method in children and young patients [11]. There are several reports of successful examination of different anomalies [12]. Transesophageal approach helps to display the origin of LCA in approximately 90%. Visualization of RCA origin is worse.

### 4 Benign coronary anomaly

Most of coronary anomalies are of benign prognosis. Their importance is mostly attributed to difficult catheterization. The question is whether or not they predispose to early atherosclerosis due to altered coronary flow. Large angiographic series and analysis
of the Coronary Artery Surgery Study (CASS) registry do not suggest such an association [13]. Only the trend for increasing stenosis in anomalous Cx was seen, although this is not significant. Overall, it seems unlikely that some anomalies predispose to accelerated atherosclerosis.

The most frequent benign anomaly (or anomaly in general) is the anomalous origin of Cx from the right aortic sinus. The Cx can arise from the proximal part of RCA or from the separate ostium. This anomaly counts for 28% of all recognized anomalies or 0.3% of all coronary catheterization findings [2]. Awareness of this coronary anomaly is important during cardiac surgery to avoid problems with myocardial protection, or during placement of prosthetic valves.

![Three-dimensional reconstruction of the heart after cutting the pulmonary artery and partially the right ventricle with the course between aorta and pulmonary artery; aorta and coronary artery in red.](image)

(RCA – right coronary artery, LAD – left anterior descending artery, AO – aorta).

### 5 Origin of coronary artery from contralateral aortic sinus

The origin of coronary artery from another sinus, including the origin of Cx from the right aortic sinus, is most the common anomaly. The majority of them are benign without relevant clinical outcomes, but some of them are potentially serious due to association with sudden cardiac death. The origin of coronary artery from contralateral aortic sinus belongs to the potentially malignant ones. There are several possible courses between their anomalous origin and their territory of distribution. We distinguish several pathways in relation to surrounding cardiac structures: 1) retrocardiac – behind the mitral and tricuspid valves; 2) retroaortic – behind the aorta; 3) interarterial - between the aorta and pulmonary artery; 4) intramuscular – also between the aorta and pulmonary artery but inside myocardial muscle; and 5) prepulmonary – before the pulmonary artery (Figure 3).
Sometimes, the retroaortic and retrocardiac course are commonly named as retrocardiac. It is important to note that the only course between the aorta and pulmonary artery is potentially malignant [10]. There are several angiographic signs to distinguish different courses, such as the typical oblique course around the aortic root in lateral or right anterior oblique view for retroaortic pathway, or muscular bridges and septal branches from proximal part of anomalous artery for intramuscular course (Figure 4). The 3-dimensional reconstruction with computer tomography or magnetic resonance is the optimal method for resolution in some questionable cases, especially between prepulmonary and interarterial courses (Figure 2) [9].

**Fig. 3** The diagram of courses of the coronary artery from the contralateral aortic sinus. (1 – retrocardiac, 2 – retroaortic, 3 – between aorta and pulmonary artery, 4 – intramyocardial, 5 – prepulmonary course, LAD – left anterior descending artery, Cx – circumflex artery, RCA – right coronary artery, PA – pulmonary artery, L/R/N – left/right/non-coronary aortic sinus, M – mitral valve, T – tricuspid valve).

Sudden death may result from one or several mechanisms that compromise flow, but the true mechanism is still discussed. Flow may be compromised by a slit-like coronary orifice, kinking or torsion of the vessel, or frank compression of the artery between the aorta and pulmonary artery.

Origin of the LCA or LAD from the right aortic sinus with a course between the aorta and pulmonary artery is an anomaly that was first described in a young healthy boy found dead after a cross-country race in 1962 [14]. Many similar cases have been reported since then. There is a general agreement that risk falls with increasing age, and death is mostly related to sports. It is also generally accepted that myocardial ischemia occurring during stress tests constitutes high-risk lesion; however, there are several reports about low sensitivity of the stress tests [15]. Some suggest that the male gender or previous symptoms constitute high-risk patients [15]. This anomaly occurs with an incidence of 1
in 12,500 coronary angiographies [2].

Surgical treatment for this anomaly is generally recommended, but some authors do not recommend this approach for all patients [16]. There are several surgical techniques. Coronary artery bypass grafting, employing both internal mammary artery and saphenous vein grafts, is most commonly used. Newer techniques are based on reimplantation of anomalous artery or “unroofing” the anomalous artery from intramural course, allowing modification of the slit-like ostium of the artery, which becomes fully patent after reconstruction [17, 18]. However, long-term patency of both arterial and vein grafts is still problematic and reimplantation is also associated with high occlusion rates [19]. Successful percutaneous treatment with stent implantation was also reported [20] but this method is not generally recommended due to unexplained pathophysiology of sudden death.

Fig. 4 Coronary angiogram in right anterior oblique view with intramuscular course of left anterior descending artery with typical septal branches in proximal part. (LAD – left anterior descending artery, RS – septal branches, RCA – right coronary artery).

The conservative approach should be individually considered for older asymptomatic patients with a negative stress test. Medical treatment is based on \( \beta \)-blockers although rigorous data are lacking. Guidelines on physical activity prescribe exclusion from all “competitive sports” [21]. However, the definition of “competitive” is vague.

Origin of the RCA from the left coronary sinus is also a potentially serious anomaly and is more common than origin of the LCA from the right aortic sinus. There are also the same 5 possible pathways as in the previous case, and the course between the aorta and pulmonary artery is potentially serious. This anomaly is believed to be benign; however, some reports have associated it with sudden cardiac death [6]. In our opinion, we reserve the surgical treatment for younger patients with symptoms or ischemia during
stress tests.

6 Coronary artery arising from pulmonary trunk

The anomalous origin of one or two coronary arteries from the pulmonary artery is a very rare malformation accounting for less than 1% of all occurrences [2]. In the case of coronary artery originating from the pulmonary artery, there is a left-to-right shunt due to a drop in pulmonary pressure after birth. Big coronary “steal” collaterals of the coronary circulation especially in the myocardial septum, and the remaining artery must supply the whole heart. We described two types of flow in anomalous artery. The “infant” type is characterized by minimal or bi-directional flow, and the “adult” type by fully developed left-to-right shunt.

Origin of both coronary arteries from the pulmonary artery is rare – there are approximately only 30 reported cases – and is usually combined with another congenital cardiac defect. The prognosis is very bad and survival beyond 2 weeks is uncommon.

Origin of the LCA from the pulmonary artery (also known as “Bland-White-Garland syndrome” or ALCAPA – Anomalous origin of the Left Coronary Artery from the Pulmonary Artery) occurs in approximately 1 in 300,000 births and is the most often seen variety of anomalous origin from the pulmonary artery [22]. It is a very commonly fatal anomaly because up to 90% of untreated cases die in the first year due to myocardial ischemia, systolic dysfunction and mitral regurgitation resulting in heart failure and sudden death. Only 10% of patients with expressive RCA dominancy survive childhood but their life expectancy still remains poor and they often die within 35 years of age [23]. In these patients a continuous murmur may present and symptoms or syndromes may occur, including angina, dyspnea, myocardial infarction, syncope and sudden death. Surgical treatment is mandatory and involves closure of the LCA with or without concomitant bypass. An alternative approach is primary reimplantation of the ostium of the LCA to the aorta or subclavian artery and secondary establishment of a double coronary artery system. Long-term results from this method appear favorable [22]. The procedure is often accompanied by mitral valve reconstruction; however, mild mitral regurgitation usually decline after surgery. Mitral regurgitation is also a prognostic marker of survival [24].

Origin of the RCA from the pulmonary artery is approximately ten-fold rarer than the previous circumstance. The patients survive usually to adulthood and they have the adult type of flow in anomalous artery. Most of the patients with this anomaly are asymptomatic, although angina and other symptoms may occur. The surgical approach is also recommended [25, 26]. Creation of a double coronary circulation is preferred to the ligation of the anomalous vessel with or without subsequent bypass graft [27].

Origin of other arteries than the LCA or RCA is also very rare. Origin of only the LAD (20 cases in literature) presents symptomatic ischemia and heart failure as with ALCAPA, and warrants surgical repair. After successful surgical corrections, relief of ischemic symptoms and improvement of systolic function can occur [28]. Origin of just the Cx was reported in adult patients only once. This patient was treated conservatively.
Sometimes, but very rarely, we can see origin of small accessory arteries with otherwise normal origin and course of the main coronary arteries. These findings are of benign prognosis. Patients are usually asymptomatic and we treat them conservatively.

7 Coronary artery fistulas

Coronary artery fistulas are unusual congenital or sometimes acquired (traumatic or post-operative) coronary artery anomalies in which blood is shunted into a cardiac chamber or great vessel, bypassing the myocardial capillary network. Congenital fistulous connections between the coronary system and cardiac chamber appear to represent persistence of embryonic intertrabecular spaces and sinusoids. They are usually seen as dilated and tortuous communications (Figure 5). There is a slight predominance for fistulas involving the RCA or terminating in the pulmonary artery [29]. These terminate to low-pressure structures, and can be clinically important due to their creation of left-to-right shunt. We found them in approximately 0.1-0.2% of coronary angiograms [2]. Large fistulae can be present as continuous cardiac murmur.

![Coronary angiogram in anterior-posterior view with fistulae from circumflex artery.](image)

(Fistula – tortuous dilated fistulae from circumflex artery to territory of right pulmonary artery).

The majority of fistulas are typically asymptomatic. The clinical presentations include fatigue, dyspnoe, angina and very rarely endocarditis, rupture and compression of surrounding structures. Myocardial ischemia can occur from decreased coronary blood flow distal to the fistulae (“steal phenomenon”), but myocardial infarction is very rare. Large fistulae can cause right ventricle overload.

The natural history of coronary fistulas is variable. Some of them close spontaneously, while others are only incidental. The clinical management is controversial and is different
between adults and children. In young patients we are more aggressive to prevent occurrence of symptoms and complications [30]. However, for adults we reserve closure only for patient with symptoms related to fistulae confirmed by diagnostic tests (ischemia on stress test, signs of volume overload of the right ventricle on echocardiography and in hemodynamic data). Asymptomatic patients are treated conservatively. Antiplatelet therapy and also prevention of bacterial endocarditis are recommended. The indications for closure are severe clinical symptoms confirmed by diagnostic tests. Cardiac surgery (epicardial and endocardial ligation) is a traditional option and is relatively safe and effective with good results. We reserve it for patients with other indications to surgery (another congenital heart defect, coronary atherosclerosis etc.). The catheter-based closure is a non-surgical treatment option using a variety of techniques, including controlled-release coils, detachable balloons and umbrellas, patent ductus arteriosus plug and various chemicals [31–33]. The treated vessel after occlusion becomes thrombosed. The condition for reliability of percutaneous intervention is based on anatomical features (ability to cannulate the distal fistulae, thin “neck” for fixation embolisation instruments). Results from the catheter and surgical literature show that both approaches have similar early effectiveness, morbidity and mortality [34]. However, important complications were reported by very experienced teams and so the closure is reserved for symptomatic patients [30].

Table 1 Summary of radical treatments for coronary artery anomalies.

<table>
<thead>
<tr>
<th>Type of coronary artery anomalies</th>
<th>Indication for radical treatment</th>
<th>Type of treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anomalous origin of coronary artery from pulmonary artery</td>
<td>For all cases except accessories coronary arteries</td>
<td>Surgical</td>
</tr>
<tr>
<td>Anomalous origin of coronary artery from opposite coronary sinus</td>
<td>Interarterial course and Young age or active athlete (ambiguous for older patients) and LMCA or RCA or LAD</td>
<td>Surgical</td>
</tr>
<tr>
<td>Coronary fistulas</td>
<td>Large left-to-right shunt or Detection of important “steal phenomenon”</td>
<td>Percutaneous if possible Surgical for rest of them</td>
</tr>
</tbody>
</table>

8 Conclusion

Coronary anomalies are mostly benign divergences. But some of them are associated with important clinical events including sudden cardiac death. Appropriate diagnosis is critical for recognition and management. Treatment can be performed by surgical or percutaneous approach. The recommendations for surgical or percutaneous approach is summarized in Table 1. However, the management is conservative in the majority of coronary artery anomalies. We recommend the “step-by-step” approach in existing practice. The first step is good anatomical diagnosis. Coronary angiography is frequently
sufficient, but computer tomography or magnetic resonance is the method of choice, especially in cases of equivocal results from the coronary angiography. The second one should be functional quantification (detection of large left-to-right shunt or important “steal phenomenon” for coronary fistula or determination of the range of regions supplied by coronary arteries with anomalous origin). The last step is risk stratification, based mainly on the age and physical activity of the patient.

Coronary artery anomalies are a very interesting and important clinical topic for cardiologists and pediatric cardiologists. The more important and latest studies are summarized in Table 2.

<table>
<thead>
<tr>
<th>Coronary artery anomalies generally</th>
<th>Yamanaka et. al. 1990</th>
<th>126595 pts.</th>
<th>incidence anomalies among coronary angiography</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Burke et. al. 1991</td>
<td>690 autopsy</td>
<td>analysis of sudden cardiac death in youngs</td>
</tr>
<tr>
<td></td>
<td>Taylor et. al. 1992</td>
<td>242 autopsy</td>
<td>analysis of sudden cardiac death due to coronary anomalies</td>
</tr>
<tr>
<td>Anomalous origin CA from pulmonary trunk</td>
<td>Schwartz et. al. 1997</td>
<td>42 pts.</td>
<td>LCA from pulmonary trunk – surgery and prediction of outcome</td>
</tr>
<tr>
<td></td>
<td>Williams et. al. 2006</td>
<td>7 pts.</td>
<td>RCA from pulmonary trunk - surgery</td>
</tr>
<tr>
<td>Anomalous origin CA from opposite sinus</td>
<td>Basso et. al. 2000</td>
<td>27 autopsy</td>
<td>analyse of sudden death</td>
</tr>
<tr>
<td></td>
<td>Frommelt et. al. 2003</td>
<td>10 patients</td>
<td>surgery and diagnosis</td>
</tr>
<tr>
<td></td>
<td>Romp et. al. 2003</td>
<td>9 patients</td>
<td>surgery</td>
</tr>
<tr>
<td></td>
<td>Erez et. al. 2006</td>
<td>9 patients</td>
<td>surgery and diagnosis</td>
</tr>
<tr>
<td></td>
<td>Armsby et. al. 2002</td>
<td>33 pts.</td>
<td>percutaneous closure</td>
</tr>
<tr>
<td></td>
<td>Behera et. al. 2006</td>
<td>13 pts.</td>
<td>Amplatzer duct occluder</td>
</tr>
<tr>
<td></td>
<td>Liang et. al. 2006</td>
<td>18 pts.</td>
<td>mid-term outcome of coil occlusion</td>
</tr>
</tbody>
</table>

(CA – coronary arteries, LCA – left coronary artery, RCA – right coronary artery)

References


