

Coronary anomalies: single centre experience

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Summary

Coronary anomalies may occur in up to 1% of general population. The clinical interest in revealing coronary anomalies relates to their occasional association with clinical symptoms or major cardiac events (death and myocardial infarction). We screened 10 340 patients who underwent diagnostic coronary angiography within the last four years. Classification, and several interesting clinical cases illustrated with angiograms are shown and discussed below.

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Coronary anomalies may occur in up to 1% of general population [1] and refers to a wide range of congenital abnormalities involving the origin, course, and structure of epicardial coronary arteries [2]. More detailed classification modified from Angelini et al [3] is presented below:

Classification of coronary anomalies

Anomalous origin Abnormalities of the origin of coronary arteries with subsequent normal epicardial course relate to the anomalous location of one or both coronary ostia. These include:

- The origin of the left main (LM), left anterior descending (LAD), left circumflex (LCx) or right coronary artery (RCA) from the pulmonary trunk (the left or right ventricles, the bronchial, internal mammary, subclavian, right carotid, or innominate arteries or the aortic arch or descending thoracic aorta);
- High takeoff of the left or right coronary ostia, defined as the location of the ostium of the left or right coronary artery more than 1 cm above the sino-tubular junction;

- The anomalous location of the coronary ostium within the aortic root, near the proper aortic sinus of Valsalva (for each artery) or outside normal “coronary” aortic sinuses;
- The absent left main trunk (split origination of the left coronary artery (LCA)).

Anomalous origin and course The entire coronary artery system may originate from a single ostium located in the left or right coronary sinus of the aorta:

- The LM originates from the RCA, or vice versa taking aberrant pathways;
- Separate origin of the LAD and LCx from the right coronary artery;
- Both the left and right coronary arteries may arise from a separate ostia located in the same, either left or right, sinus of the aorta.

Anomalous course and termination

- An intramyocardial course (i.e., the myocardial bridge).

Major epicardial coronary arteries may terminate abnormally into one of the cardiac chambers (the right or left atrium, the right or left ventricle), coronary sinus, superior vena cava, pulmonary

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artery, pulmonary vein and, thus, produce fistulas originating from:

- the left (50–60%);
- the right (30–40%);
- or both (2–5%) coronary artery systems.

Abnormal coronary structure

- Congenital epicardial coronary artery stenosis usually is caused by a membrane or a fibrotic ridge.
- Coronary artery atresia is characterized by the presence of an ostial dimple in the left or right aortic sinus that terminates in a cordlike fibrotic structure without a patent lumen.
- Hypoplastic coronary arteries have a small luminal diameter (usually <1 mm) and reduced length. The latter is often associated with the absence of the posterior descending coronary artery.
- Coronary ectasia or aneurysm.
- The absent coronary artery.

In adults, the clinical interest in revealing coronary anomalies relates to their occasional association with clinical symptoms for instance chest pain, dyspnea, syncope or major cardiac events (death and myocardial infarction) [2,4]. We suppose it is interesting to present data obtained in our centre on this quite rare pathology. Therefore, we screened all patients who underwent diagnostic coronary angiography within the last four years. Several interesting clinical cases illustrated with angiograms are shown and discussed here.

Clinical cases

Between January 2000 and December 2004, 10 340 coronary angiograms were performed and screened on purpose to identify the presence of coronary anomalies. In total 185 coronary anomalies (1.8%) were identified, grouped according to the classification and presented in Table 1.

The intramyocardial course of the coronary artery was the most frequent finding (32% of cases) among all anomalies identified on angiograms. The anomalous origin of the left coronary artery from the pulmonary artery and from the right coronary artery occurred in 2 (1%) and 33 cases (18%), respectively.

Case 1

A 63-year-old patient with a history of arterial hypertension presented with acute chest pain with significant ST segment elevation on the ECG

Table 1.
The incidence of coronary anomalies

Coronary anomalies	Total n = 185
<i>Intramural coronary artery (muscular bridge)</i>	60 (32.4%)
<i>Separate ostium of LAD and LCx</i>	25 (13.5%)
<i>Left coronary artery from right coronary sinus</i>	33 (17.8%)
RCx	22
LAD	6
LM	5
<i>Coronary ectasia or aneurysm</i>	33 (17.8%)
LCx	7
LM	4
RCA	12
LAD	8
All coronaries	2
<i>Fistulas from RCA or LCA artery to:</i>	19 (10.3%)
Right ventricle from RCA	1
Right atrium from LCx	2
Coronary sinus from LCx	2
Pulmonary artery from:	3
LCx	5
LAD	3
RCA	3
LCA arising from pulmonary artery	2 (1.1%)
Anomalous origin of RCA from non-coronary sinus	10 (5.4%)
Kinking	3 (1.6%)

LAD – left anterior descending; LCA – left coronary artery; LCx – left circumflex; LM – left main; RCA – right coronary artery.

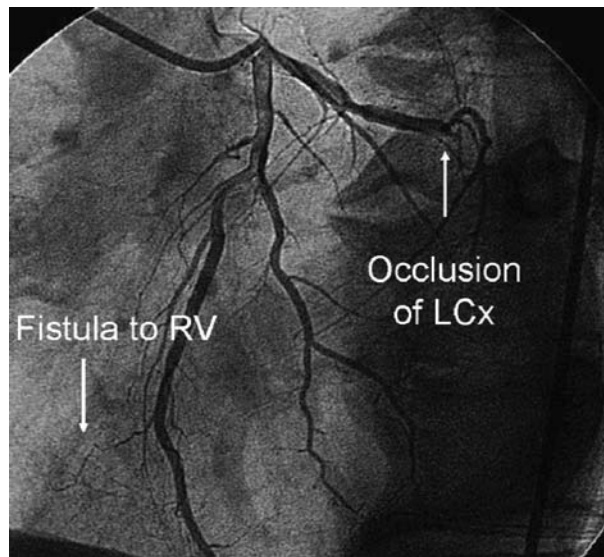


Figure 1. Angiogram of the left coronary artery: the occlusion of the LCx and fistulae from the LAD to the right ventricle. LAD – left descending coronary artery; LCx – left circumflex coronary artery; RV – right ventricle.

and elevated Troponin. An angiogram demonstrated the occlusion of the mid LCx and fistulas: from the LAD to the right ventricle (RV) (Figure 1) and from the RCA to the RV (Figure 2). Primary

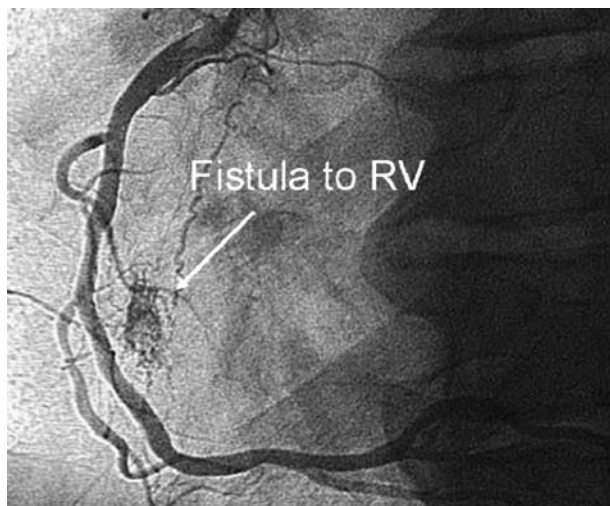


Figure 2. Angiogram of the right coronary artery: the fistula from the RCA to the right ventricle. RV – right ventricle.

percutaneous transluminal coronary angioplasty (PTCA) was performed and TIMI 3 flow was restored in the myocardial territory of the LCx. His post myocardial infarction (MI) hospital course was unremarkable. The patient was discharged in stable condition.

Case 2

A 77-year-old female presented to the emergency room with chest discomfort, fatigue and shortness of breath at rest. Over the previous few weeks she had experienced progressive dyspnea on exertion and fatigue. The patient was diagnosed with a grade III mitral valve insufficiency, the dilated left ventricle and atrial fibrillation. The patient was transferred for cardiac catheterization prior to mitral valve surgery. Coronary angiography revealed the presence of hemodynamically non-significant fistulas originating from the LCx to the coronary sinus and the pulmonary artery (Figure 3 and Figure 4).

Case 3

A 74-year-old female patient with hyperlipidemia and arterial hypertension was admitted to the hospital with acute anterior and lateral infarction of 5-hour duration. The ECG showed severe ST segment elevation in leads V1–V6, ST depression and T-wave inversion in leads II, III, and aVF. Urgent coronary angiography was performed and anomalous anatomy of coronary arteries revealed. The origin of the left main came from the RCA (Figure 5). In addition occlusions of the mid RCA and the large diagonal artery were identified (Figure 5). Primary balloon angioplasty was applied successfully in the diagonal and the RCA (Figure 6). TIMI-3 flow was established. Electrical cardioversion was needed during PTCA because

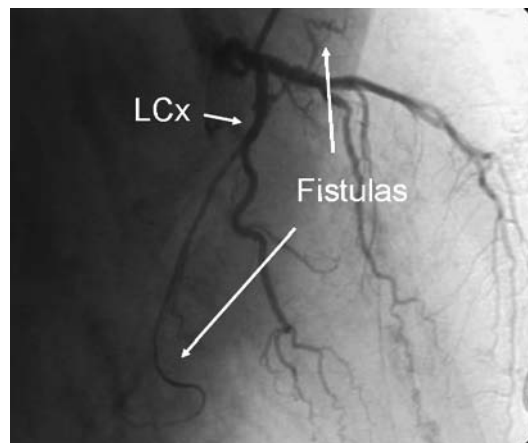


Figure 3. Left coronary artery: fistulae originating from the LCx to the coronary sinus and the pulmonary artery. LCx – left circumflex coronary artery.

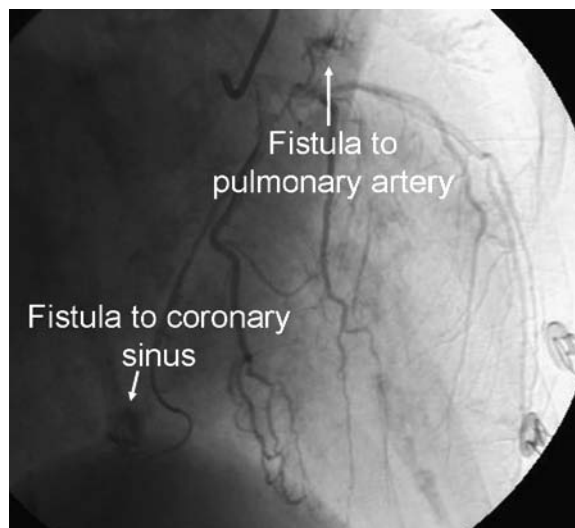


Figure 4. Fistula to the pulmonary artery and the coronary sinus.

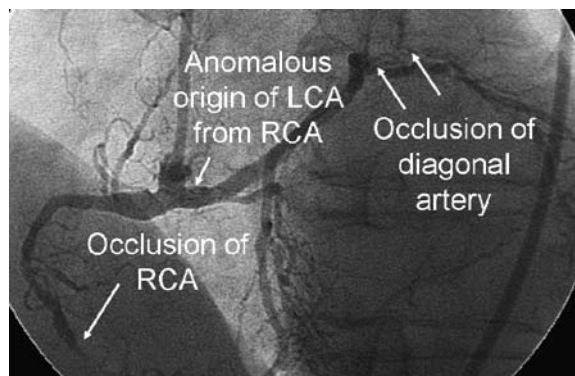


Figure 5. Angiogram shows anomalous origin of the LCA from the RCA, the occlusion of the RCA and the diagonal artery. LCA – left coronary artery; RCA – right coronary artery.

of ventricular fibrillation after RCA recanalization. The patient was discharged in stable condition.

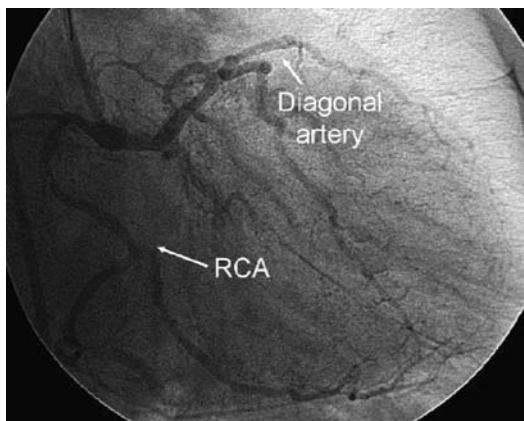


Figure 6. Successful PTCA of the RCA and the diagonal artery was performed. PTCA – percutaneous transluminal coronary angioplasty; RCA – right coronary artery.

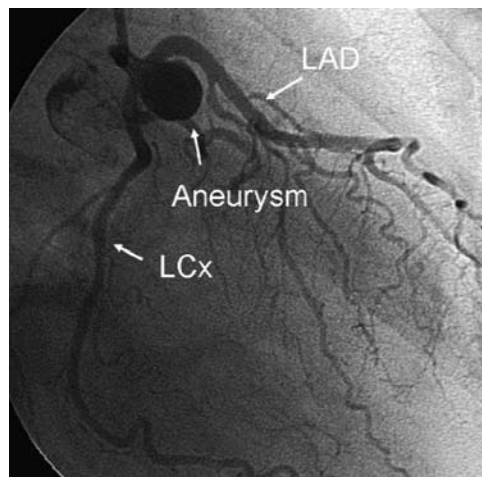


Figure 8. Aneurysm of the LCA – the same patient as on Figure 7, another view. LAD – left anterior descending coronary artery; LCx – left circumflex coronary artery.

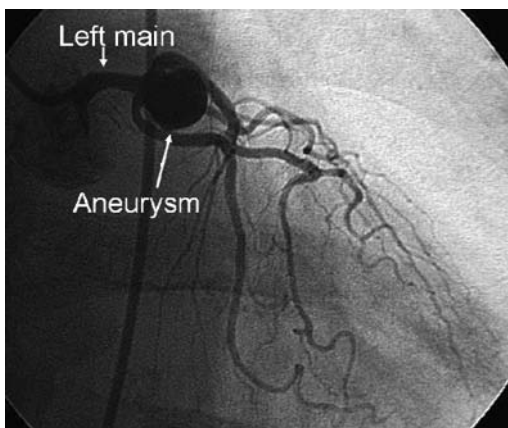


Figure 7. Aneurysm of the LCA. LCA – left coronary artery.

Case 4

A 71-year-old female with hyperlipidemia and arterial hypertension was admitted to the hospital with acute chest pain irradiating to the left hand and back. Over the previous three days she had frequent episodes of chest pain lasting up to 15 minutes. The ECG was inconclusive for myocardial ischemia, the troponine test was negative. Coronary angiography showed the presence of an aneurysm (9 mm in diameter) in the LCA (Figure 7 and Figure 8). Heart surgery was not indicated and the patient was discharged for medical treatment.

Case 5

A 58-year-old-male with arterial hypertension suffering from dyspnea was admitted for aortic valve replacement surgery. Past medical history showed the dilated ascending aorta and severe aortic insufficiency grade III. Coronary angiography revealed a fistula connecting the LAD and RCA (Figure 9), the normal origin of the RCA (Figure 10). The patient was transferred for aortic valve replacement surgery.

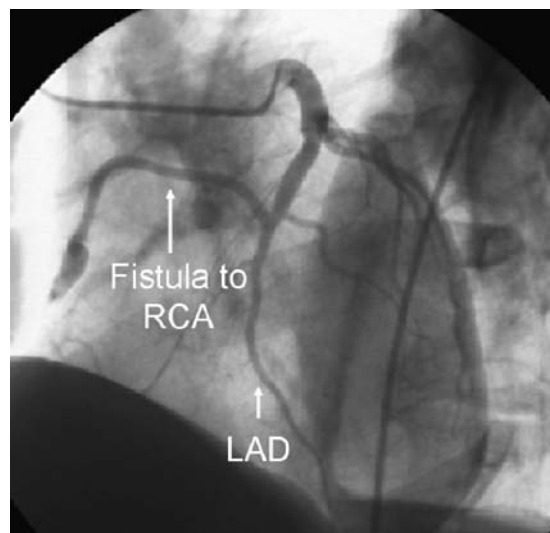


Figure 9. Fistula originating from the LAD to the RCA. LAD – left anterior descending coronary artery; RCA – right coronary artery.

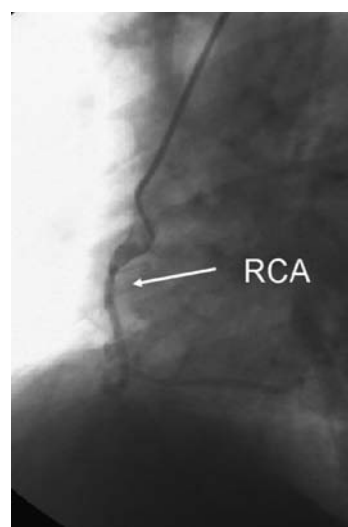


Figure 10. Origin of the RCA from the right coronary sinus. RCA – right coronary artery.

Discussion

Most coronary artery anomalies are clinically silent and usually are found incidentally during angiographic evaluation for other cardiac diseases. Certain types of anomalies that are associated with impaired myocardial perfusion can result in angina, congestive heart failure, myocardial infarction, cardiomyopathy, ventricular aneurysms, or sudden death. Some data indicated the relationship between the anomalous origin of the left coronary artery from the pulmonary artery and acute anterolateral myocardial infarction in newborns, between myocardial bridges [5] or the anomalous origination of the left coronary artery from the right sinus and sudden death [6]. Therefore, the identification of these types of coronary anomalies can be life saving if surgical or percutaneous treatment is applied. The patient described in case 1 presented with acute myocardial infarction and fistulae were found incidentally. Primary PCI restored good flow in the territory of the LCx and the patient was discharged in stable condition. An identified fistulae possibly were not hemodynamically important as the patient was free of myocardial ischemia previous to an acute coronary event. Moreover, there is a lack of evidence about the association of the coronary anomalies and accelerated coronary atherosclerosis. The second and fifth cases show hemodynamically non-significant fistulae associated with the dilated left ventricle due to severe mitral regurgitation (case 2) and aortic valve insufficiency (case 5). The ratio of pulmonic to systemic flow calculation from right heart catheterisation data could be mandatory if a large coronary fistula is present. It was not done in case 2 as the patient refused cardiac surgery. Some reports indicates that hemodynamically significant coronary artery fistulae can be successfully treated percutaneously with implantable coil occlusion [7], umbrella closure devices [8,9] or a covered stent [10,11].

Cardiac surgery could be only definitive treatment choice for patients with the anomalous origin of the left coronary artery from the pulmonary artery (Bland-White-Garland syndrome or ALCAPA) which is a life threatening coronary artery anomaly. The left coronary artery originating from the pulmonary trunk is resected optimally from the pulmonary trunk and reimplanted into the ascending aorta [12]. The left main coronary artery with the anomalous origin from the right coronary can be associated with acute myocardial ischemia or sudden death, especially when it passes between the aorta and the pulmonary trunk [13]. Therefore, surgical correction or bypass surgery can be indicated.

Case 4 describes a coronary artery aneurysm with the diameter of 9 millimetres. This is a relatively infrequent abnormality, which poses a challenge to a physician regarding management as the prognosis of the coronary artery aneurysm is not well known. In our case the patient was discharged receiving the treatment with antiplatelet agents and anticoagulation and was doing fine three months later. Although surgical experience (the resection of the aneurysm with end-to-end interposition of a saphenous vein autograft) has shown an excellent outcome [14,15] most authors agree that surgery should be reserved for patients with significant coronary stenosis, or those with significant angina despite adequate medical treatment [16,17].

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