Multi-imaging evaluation and long-term outcome of a patient with chest pain and an anomalous right coronary artery arising from pulmonary artery: a case report

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Background
Anomalous right coronary artery from pulmonary artery (ARCAPA) is a rare coronary anomaly. Adult patients usually present with few symptoms due to extensive collateral network from left coronary artery, with little/absent symptoms. Few data exist regarding surgical vs. conservative strategy for paucisymptomatic cases. Moreover, consensus is lacking.

Case summary
We describe the case of a 52-year-old male patient with undiagnosed ARCAPA, who acceded to our emergency department with suspected acute myocardial infarction and was discharged with medical therapy after demonstration of mild ischaemia at myocardial perfusion imaging with dipyridamole and bicycle exercise test.

Discussion
The patient completed 2-year event-free follow up. After complete imaging assessment and thorough clinical evaluation, medical management could be regarded as valid alternative to surgery for paucisymptomatic ARCAPA patients with evidence of limited ischaemia.

Keywords
Coronary anomalies • Cardiac catheterization • Scintigraphy • Coronary artery disease • Case report

ESC Curriculum
2.1 Imaging modalities • 3.1 Coronary artery disease

Learning points
• Evaluation of inducible ischaemia is useful to assess the best management of ARCAPA patients.
• Myocardial perfusion imaging with bicycle exercise test and dipyridamole could explore different mechanisms of ischaemia in ARCAPA patients.
• ARCAPA is commonly treated by surgery. In an adult patient with mild burden of ischaemia, a conservative management may be considered.

Introduction
Anomalous right coronary artery from pulmonary artery (ARCAPA) is a rare congenital heart disease accounting for 0.12% of all coronary anomalies. Typically, the anomalous right coronary originates from the pulmonary trunk and it receives retrograde collateral circulation from the left coronary artery, due to the pressure gradient between the coronary and the pulmonary circulation. One-third of the patients with ARCAPA are asymptomatic. In symptomatic patients, symptoms may include stable angina, dyspnoea, and heart failure and usually
manifest early after birth or between the age 40 and 60 years.1,2 We report the case of an adult patient admitted in the emergency department for the onset of chest pain following a vasovagal syndrome who underwent multi-imaging evaluation and ultimately received conservative management.

**Timeline**

| Presentation | The patient presented to the emergency department with chest pain, sweating, and vomiting. |
|-------------|--------------------------------------------------------------------------------------------------|
| In-hospital management and evaluation | Coronary angiography revealed ARCAPA, confirmed by computed tomography angiography. Myocardial perfusion imaging showed a reversible mild inferior perfusion defect. A conservative strategy was pursued. |
| Two years follow up | The patient was asymptomatic, no clinical events were present, and burden of ischaemia remained unchanged. |

**Case report**

A 52-year-old man was admitted in our institutional emergency department for chest pain, sweating, and vomiting. The patient had no medical history or known cardiovascular risk factors and denied any other symptoms. Blood pressure was 85/50 mmHg and heart rate 54 b.p.m. Intravenous fluid administration was started to manage hypotension. Twelve-lead electrocardiogram showed sinus rhythm with only non-specific ST-segment changes (Figure 1). Trans-thoracic echocardiogram showed normal global and regional left and right ventricular function and normal size of left and right chambers. In particular, left ventricular ejection fraction was 60%, left ventricular end-diastolic diameter was 45 mm, and right ventricular end-diastolic diameter was 38 mm. Troponin T levels were mildly elevated (3.0 and 5.0 ng/L at 0 and 3 h, respectively).

The clinical presentation was consistent with non-ST-segment elevation acute coronary syndrome (ACS). Aspirin was administered and patient underwent coronary angiography, which revealed a patent large left anterior descending coronary artery providing collaterals to a large dominant right coronary artery draining into the pulmonary artery (Figure 2A). Non-obstructive coronary artery disease was found. Right and left cardiac catheterization was concomitantly performed, showing a normal pulmonary artery pressure (24/10 mmHg, average 12 mmHg) and a non-significant left-to-right shunt (pulmonary venous oxygen saturation 95%; pulmonary artery oxygen saturation 80%; mixed venous oxygen saturation 76%; QP/QS: 1.3). Then, the patient was transferred to our Cardiac Care Unit for monitoring.

After 2 days, a cardiac computed tomography (CT) angiography was performed to better define the coronary anatomy. CT angiography confirmed ARCAPA, excluded any other coronary anomalies and showed normal calibre of pulmonary arterial vessels (diameter of pulmonary trunk: 26 mm; Figure 2B). Finally, a 99mTc-Technetium-gated single-photon emission computerized tomography (SPECT) with bicycle exercise test and then a 201TlThallium-gated SPECT with dipyridamole were performed to assess the functional implication of the coronary anomaly (Figure 3), showing a reversible mild inferior perfusion defect without any significant difference between the two tests. Stress test was maximal: it lasted 9 min and the patient attained 154 b.p.m. (91% of target heart rate), 6.8 estimated metabolic equivalents and 180/85 mmHg as peak blood pressure. The patient experiences a subsequent event-free hospital stay, and was discharged on aspirin 100 mg and bisoprolol 2.5 mg. The patient was educated on actions to prevent future vaso-vagal episodes, including keeping adequate fluid intake (2 L/day), recognizing prodromes and lying down and performing counter manoeuvres in case of vaso-vagal symptoms. Also, in order to minimize gastrointestinal triggers the patient was visited by a gastroenterologist and diet recommendations were made.

At 2-year follow up, the patient was asymptomatic and repeated myocardial scintigraphy with dipyridamole stress test was performed after beta-blocker washout. The test confirmed the reversible mild inferior perfusion defect, previously observed. Bisoprolol dosage was up-titrated to 3.75 mg daily and aspirin was maintained.

**Discussion**

Consensus regarding ARCAPA treatment and management is lacking. The main aim of the correction is the prevention of ‘coronary steal’, which could lead to myocardial ischaemia and failure due to left-to-right shunt.1 The 2018 American Heart Association and 2020 European Society of Cardiology guidelines3,4 both concluded that surgery, preferably by aortic re-implantation of the ARCAPA, is indicated in patients with symptoms (class of recommendation I, level of evidence C) and should be considered in asymptomatic patients with ventricular dysfunction, or myocardial ischaemia attributable to coronary anomaly (class of recommendation IIa, level of evidence C). However, the overall operative mortality of 2.5% is not negligible.1 Moreover, the exact burden of ischaemia is difficult to quantify and robust scientific evidence is lacking. Conservative treatment is seldom an option. In a recent review of 193 cases of ARCAPA, surgery was not performed just in 17 cases (7.6%).1 Consequently, risk/benefit of this approach is still unclear. In this case, a conservative management strategy was decided after heart team discussion. This decision took into account the mild reversible perfusion defect at scintigraphy, the absence of symptoms during both stress types, the normal cardiac function, and the haemodynamically irrelevant left-to-right shunt.

We hypothesize that, in our patient, the severe hypotension during the vaso-vagal syndrome induced a reduction of the coronary perfusion pressure in the collateral-dependent right coronary artery territory, leading to myocardial ischaemia with typical chest pain. On the counterpart, also in stress conditions like exercise or hypertension, the increase in oxygen demand could lead to myocardial hypoperfusion. To investigate both these triggers, we decided to perform the scintigraphy with pharmacological and exercise imaging stress. The pathophysiological mechanism of myocardial hypoperfusion reproduced during scintigraphy with dipyridamole stress test is based on the reduction of systemic arterial pressure and distal vessel resistance in the collateral-dependent myocardium and is likely to mimic the coronary steal induced by the vaso-vagal syndrome. On the other hand, with effort stress test, we tried to reproduce ischaemia deriving from stress triggers, which is more common in daily activities. Subsequently, we deduced that from the clinical standpoint one single pharmacological imaging stress test would have been sufficient in our case, thus avoiding an excessive radiation exposure. Similarly, repeating a scintigraphy at 2-year follow up may not be indicated in an asymptomatic patient. In addition, stress cardiac magnetic resonance imaging in this context of subendocardial scarring and ischaemia would have provided a superior spatial resolution, without radiation exposure. In conclusion, the patient was discharged on aspirin as a
Figure 1  Twelve-lead electrocardiogram showing alterations attributable to inferior ischaemia.

Figure 2  Coronary angiography shows left anterior descending artery collateralizing to a large right coronary artery, finally draining into the pulmonary artery (A). Three-dimensional computed tomography reconstruction confirms this finding (B). Arrow shows the position of Swan–Ganz catheter, placed to confirm right coronary artery drainage into pulmonary artery. Asterisk indicates the point of drainage from right coronary artery to pulmonary artery.
secondary prevention to ACS, although its utility is unclear in the absence of any thrombotic/atherosclerotic disease.

Conclusion
This is a case of a medically treated ARCAPA undergoing multi-imaging evaluation, with a good mid-term outcome. Although further evidence and long-term follow up are required, medical management may represent a reasonable alternative to surgery in selected patients with ARCAPA.

Supplementary material
Supplementary material is available at European Heart Journal – Case Reports online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary material.

Consent: The authors confirm that written consent for submission and publication of this case report, including images and associated text, has been obtained from the patient in line with COPE guidance.

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