

Coronary artery fistula draining into pulmonary artery and optimal management: a review

ABSTRACT

Coronary artery fistula is a rare congenital malformation of high variability. The disease is illustrated with a description of a case example. The management of patients with coronary artery fistulas remains controversial. Both spontaneous regression and life threatening complications have been described. The fistula can be ligated or embolised; however, there are no long term outcome data regarding management. Intraoperative risk of myocardial infarction is less than 5% and death rate varies between 0% and 6%. Due to a small number of cases being described in the literature and a lack of evidence on optimal management, further research is needed in order to determine the best treatment options.

INTRODUCTION

Coronary artery fistulas are complex and highly variable pathologies that may present with unsuspected symptoms and complications. Illustration of the problems of this condition is presented in a case example.

A 79-year-old female presented with chest pain, fatigue and a progressive increase in exercise intolerance. Chest pain was present in the posterior and distal aspects of her thorax, as well as in the clavicular region bilaterally. It has been associated with arthritis. A significant increase in fatigue and inability to walk more than 500 yards was noted. The patient denied the presence of any palpitations, loss of consciousness or breathlessness. She suffers from vertigo and light headedness on numerous occasions.

PREVIOUS HISTORY

The patient has progressive aortic stenosis with a normal ventricular size and function. In 2010, paroxysmal atrial fibrillation with troponin 0.66 was detected. She was diagnosed with temporal arteritis in 2009 (prescribed prednisolone 7 mg). She also has a long history of multiple lumbar disc surgeries including five laminectomies and one lumbosacral fusion with 'cage'. The patient has been anaemic in the past due to a duodenal ulcer and diverticulum bleeds in 2009. A terminal colostomy was created following the diverticular bleed. She had small bowel obstruction due to adhesions in 2006.

ON EXAMINATION

The patient appeared mildly cushinoid. A moderately slow rising pulse was detected with BP 152/84. Venous pressure was normal. Neither ventricle was palpable. The second heart sound was barely audible in pulmonary area and inaudible elsewhere. There was a harsh 3/6 ejection systolic murmur throughout the pericardium and radiating to both carotids. The chest was clear and no peripheral oedema was noted.

CARDIAC HISTORY DETAILS

The patient was followed up for aortic stenosis until the transthoracic echocardiogram gradient reached a peak of 95 mm Hg (mean 53 mm Hg) and the need for aortic valve replacement was established. Ventricular size and function were normal (3.3 cm and 5.5 cm) and there was mild septal hypertrophy

(1.3 cm). The left atrium was mildly dilated (4.5 cm) and the root diameter was normal (3.1 cm). The aortic valve area was 0.3 cm² by the continuity equation. Mitral valves were mildly thickened with mild mitral regurgitation present. The size of both atria and right ventricle was normal. Ventricular systolic pressure was 33 mm Hg plus the right atrial mean. An echo revealed an aortic gradient of 81 mm Hg (mean 50 mm Hg, aortic valve area 0.6 cm²).

The ECG showed sinus rhythm, normal axis, PR interval and QRS complexes. The only abnormality detected was non-specific T wave flattening in aVL lead. The chest radiograph was normal.

The coronary angiogram showed that the left main stem, left anterior descending and left circumflex were all angiographically smooth. The right coronary artery was a dominant vessel with minor coronary disease. There was also an abnormal fistula arising from the left anterior descending and draining into the pulmonary artery (figures 1 and 2).

DISCUSSION

Arterio-venous fistulas in the coronary system occur in 0.1%–0.3% of the adult population.¹ They are congenital malformations of coronary arteries which may drain into the cardiac chambers, great veins or pulmonary arteries. The fistulas usually drain into the right ventricle (41%).^{2–3} Other cases include drainage into right atrium (26%), pulmonary artery (17%), coronary sinus (7%), left atrium (5%), left ventricle (3%) and superior vena cava (1%).^{2–3} The variety is clearly shown by how differently each case is described in the literature.^{4–9} They are usually small and asymptomatic and arise more frequently from the left coronary circulation according to some sources⁶ and more frequently from the right in other papers (50% right coronary artery, 42% left coronary artery, 5% both vessels).³ Symptoms vary with patient age and size of fistula. The main and most common symptoms include angina, myocardial

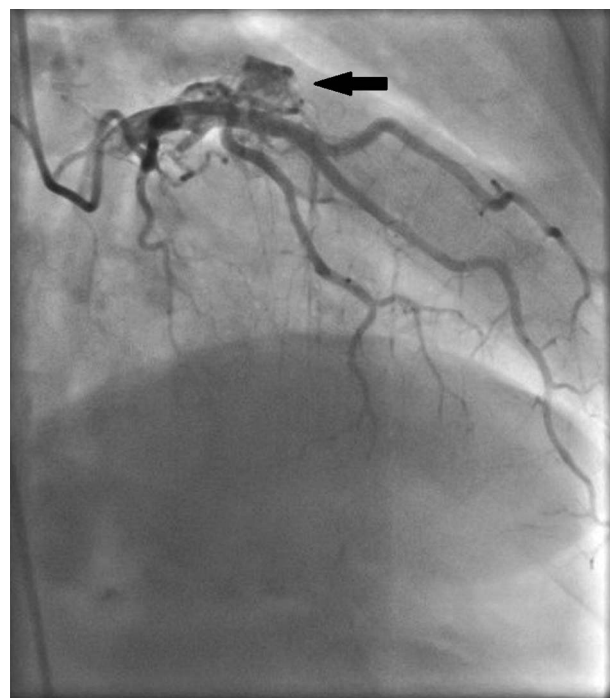


Figure 1 Picture of a coronary angiogram showing an abnormal coronary fistula arising from the left anterior descending artery and draining into the pulmonary artery.

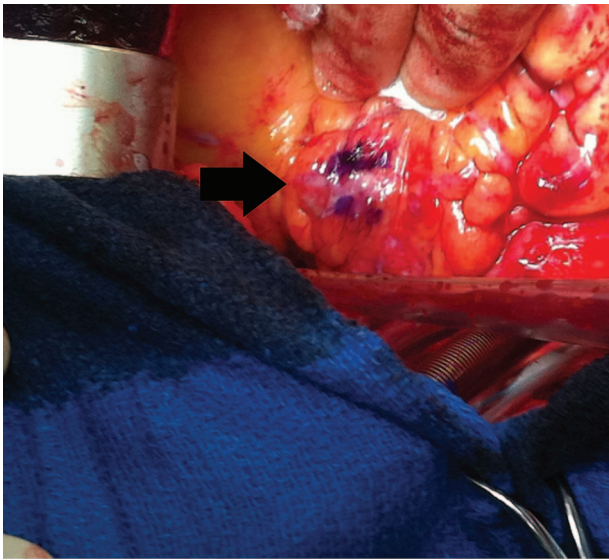


Figure 2 Intraoperative view of the coronary artery fistula arising from the left anterior descending artery and draining into the pulmonary artery. The origin is coloured with purple marker.

infarction, heart failure, arrhythmias and endocarditis.⁷ An aneurysm is formed in association with the fistula in 19% of cases and multiple fistulas form in 16% of cases.⁵ Unfortunately, diagnosis is often accidental while patients are being investigated for other cardiac diseases.

The management of patients with coronary artery fistulas remains controversial. Both spontaneous regression as well as life threatening complications have been described.⁶ The fistula can be ligated or embolised; however, there are no long term outcome data regarding this management. Percutaneous occlusion has also been described being used in some cases.^{8–10–12} The future of such interventions could be a percutaneous procedure using a robotic catheter in order to enhance accuracy, safety and limit complication rate by reduced trauma to surrounding tissues.¹³ Intraoperative risk of myocardial infarction is less than 5%¹⁴ and death rate varies between 0% and 6%.⁸

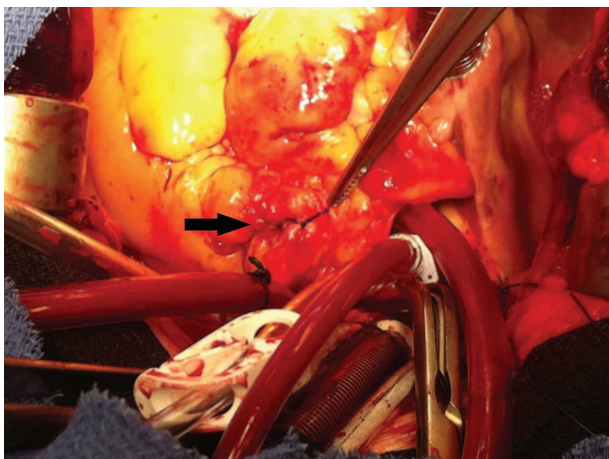


Figure 3 The fistula has been closed of using a slip knot. It was applied in the most proximal aspect of the fistula (closest to left anterior descending artery).

In this case, the fistula was closed off during aortic valve replacement. The proximal aspect of the defect was occluded using sutures and after no signs of cardiac ischaemia were detected on ECG it was firmly tied off. Figure 3 shows the fistula after it had been closed off.

CONCLUSIONS

Coronary artery fistula is a rare congenital malformation of high variability. Due to a small number of cases being described in the literature and even less evidence on optimal management, further research is needed to determine the best treatment option. In the future, new technologies such as percutaneous techniques and robotics may be the solution for these patients, improving safety, minimising trauma and enhancing optimal visualisation for the operator.

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