# Extensive multiple coronary artery to left ventricular fistulas – a 10-year case history

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**Summary**

**We report the 10-year case history of a 50-year-old woman who presented with angina due to extensive, bilateral, multiple coronary artery to left ventricular fistulas (MCALVF). $^{201}$Thallium myocardial scintigraphy revealed reversible ischaemia due to coronary 'steal'. Cardiac catheterisation showed left ventricular dilatation due to high cardiac output from significant coronary to left ventricular shunt. This is the first case report where the decision not to perform surgical ligation of the fistulas has provided a unique example of the natural history of a MCALVF, documented by serial data over 10 years.**

**Key words:** coronary artery, fistula, left ventricle, dilatation, ischaemia, $^{201}$Thallium scintigraphy.

## Case report

A 40-year-old female smoker presented in 1987 with classical angina, shortness of breath and fatigue at 200 metres. She had had a left lower lobectomy for bronchiectasis at the age of 12, three uncomplicated pregnancies and was pre-menopausal. Her mother had died of a myocardial infarction aged 75. Cardiovascular and general examination was normal. Blood pressure was 140/90 mmHg. ECG revealed T-wave inversion in leads, AVL, and $V_{4-6}$. Chest X-ray (CXR) showed only her previous thoracic surgery. Full blood count, biochemical profile and thyroid function were normal, but fasting serum cholesterol was 7.3 mmol/L. A maximal Bruce exercise ECG was strongly positive with ST-depression before the end of the second stage. $^{201}$Thallium scintigraphy confirmed antero-septal and antero-lateral reversible ischaemia. Transthoracic echocardiography revealed no evidence of left ventricular (LV) dilatation or dysfunction and showed normal valves and Doppler flows.

Cardiac catheterisation in 1988 showed good LV contraction, with massive dilation of both coronary arteries (left main stem > 15 mm) but no evidence of coronary atheroma. Both coronary arteries supplied an extensive network of fistulous communications which rapidly filled the left ventricular cavity. Left and right heart pressures were normal (table 1). Central venous, pulmonic and aortic arterial blood lactate samples taken during fast atrial pacing were unremarkable (table 1); these were taken because the coronary sinus could neither be identified or cannulated, so no coronary sinus values were obtained.

A diagnosis of coronary artery fistula causing myocardial ischaemia due to coronary 'steal' was made. Surgical advice was sought but, owing to the size and complexity of the lesion, surgical ligation was felt unlikely to be successful and medical therapy was recommended. She was started on isosorbide mononitrate 20 mg b.d. and nifedipine 10 g b.d., later changed to verapamil SR 240 mg with some improvement in her symptoms. She was found to be intolerant of beta blockers due to wheeze.

The patient coped well, living within her anginal limits up until 1995 with no change in her symptoms. At this time, a deterioration in symptoms was observed, predominantly shortness of breath. This culminated in an admission with unstable angina in 1996. The patient was noted to have ankle oedema, but no other clinical features of cardiac failure. ECG was unchanged, as was the CXR. Lung function tests were normal (FVC 2.59 L, FEV 1.78 L/sec). There was no serological evidence of myocardial infarction. An angiotensin-converting enzyme inhibitor was commenced at this time which caused a symptomatic improvement in her dyspnoea but was stopped soon after for intolerable cough.

**Table 1.** Cardiac catheter findings from 1988 and 1996

| Site | 1988 Pressure (mmHg) | 1988 Mean | 1988 $O_2$ (%) | 1988 Lactate (mmol/L) | 1996 Pressure (mmHg) | 1996 Mean | 1996 $O_2$ (%) |
|---|---|---|---|---|---|---|---|
| Aorta | 110/75 | – | – | 1.2 | 110/73 | – | 97 |
| LV | 115/15 | – | – | – | 110/21 | – | 97 |
| PA | 25/12 | 16 | – | 1.4 | 33/20 | 23 | 74 |
| RV | 20/10 | – | – | 1.5 | 31/6 | – | 74 |
| HRA | – | – | – | – | – | – | 75 |
| MRA | – | 3 | – | – | – | 6 | 77.3 |
| LRA | – | – | – | – | – | – | 73 |
| IVC | – | – | – | 1.3 | – | – | 83.4 |
| SVC | – | – | – | – | – | – | 69 |

**Key:** HRA = high right atrium; IVC = inferior vena cava; LRA = low right atrium; LV = left ventricle; MRA = mid right atrium; PA = pulmonary artery; RV = right ventricle; SVC = superior vena cava

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**Figure 1.** Left ventriculogram in diastole (RAO view), showing mild dilatation of the left ventricle and massive dilatation of the left coronary artery and branches

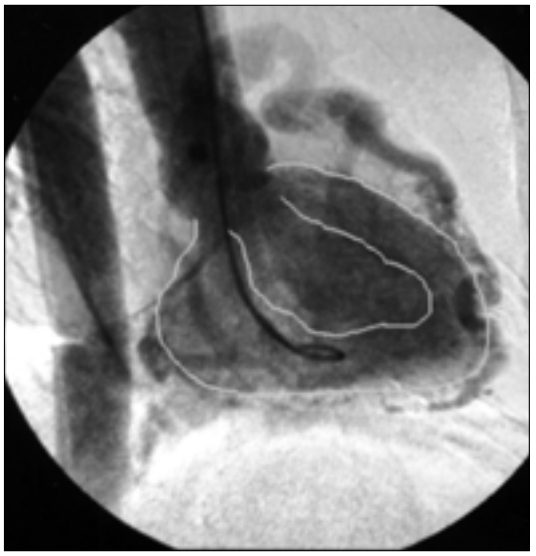
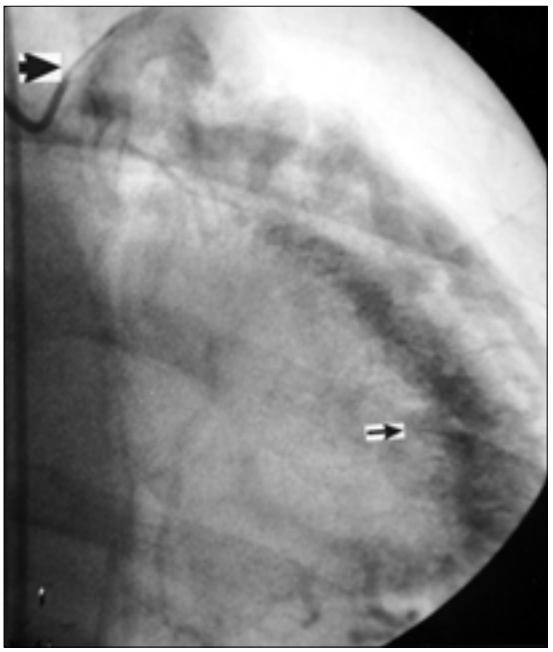


**Figure 2.** Selective left coronary angiogram (RAO view), showing dilated left main stem (large arrow) and multiple coronary artery to left ventricle fistulas



Continuing symptoms led to repeat cardiac catheterisation in June 1996. Although left ventricular systolic function was preserved, the ventricle was mildly dilated compared with the previous study and end diastolic pressure had increased (table 1). Coronary artery anatomy was unchanged (figures 1 and 2). Again, no coronary sinus was seen during the venous phase. There was no step up in oximetry from mixed venous return to pulmonary artery, excluding significant left to right shunt (table 1). Cardiac output measured by thermodilution at rest was 6.2 L/min and cardiac index was 3.55 L/min/m$^2$. Transoesophageal echocardiography performed in 1998 confirmed mild dilation (6 cm in diastole) but good function of the LV and normal valve structure and function. Both atria were normal in size and structure. There was no atrial or ventricular septal defect and no anomalous venous drainage. The coronary sinus could not be identified.

The patient continues to have controlled angina on medical therapy: verapamil SR 240 mg o.d., frusemide 40 mg o.d., isosorbide mononitrate SR 60 mg o.d., and nicorandil 20 mg b.d.

## Discussion

Multiple coronary artery to left ventricular fistulas (MCALVF) are extremely rare. To-date, only 13 cases have been reported.[1,2] This is the first case where the decision not to perform surgical ligation provides a unique example of the natural history of a MCALVF, documented by serial catheter data over 10 years.

Although extra cardiac coronary anastamoses were originally described by Von Haller[3] in 1803, widespread recognition of coronary artery fistulas followed the increased availability of coronary angiography. They are rare and usually discovered incidentally at coronary angiography during investigation for other cardiac disease. A retrospective review of 14,708 adult coronary angiograms revealed 20 fistulas (0.13%), only one of which was multiple.[4] Other workers have confirmed a similar prevalence (0.1–0.2%); multiple fistulas representing less than 5% of this total.[5,6]

Presentation depends on the site of origin, size, multiplicity, and site of drainage.[4] The majority of coronary artery fistulas drain to the pulmonary arteries; they can drain to any cardiac chamber, the least likely being the left ventricle. They are most commonly seen in children but have been reported in adults.[4] Clinically minor lesions are often silent but presentation with coronary ischaemia due to coronary 'steal' is well recognised.[1-4] Cardiac arrhythmia, syncope, myocardial infarction and endocarditis have all been reported with this anomaly.[4] Occasionally, a continuous murmur has been reported in association with coronary artery fistulas.[1-4] In more extensive cases, the high flow may lead to significant left to right or left to left shunting. The latter leads to volume overload of the left ventricle in a manner likened to aortic regurgitation leading to LV dilatation,[1] as documented in this case along with an increase in end diastolic pressure. A proportion of this patient's dyspnoea may be due to high LV output failure. Indeed, her symptoms appeared to improve with the institution of an angiotensin-converting enzyme inhibitor, however she was unable to tolerate this medication due to side effects. We have shown reversible ischaemia with $^{201}$Thallium scintigraphy suggesting coronary 'steal' and believe this explains her anginal symptoms. Coronary artery to bronchial artery fistula 'steal' syndrome has been shown previously with $^{201}$Thallium scintigraphy.[3]

**Key messages**

- Multiple coronary artery to left ventricular fistulas is rare
- Transcatheter vascular occlusion is the accepted treatment option for this condition
- This case illustrates the role of medical therapy in this condition when surgical treatment is difficult, e.g. when there are multiple communications

The high coronary flow in more extensive cases causes dilation and tortuosity of the supplying epicardial artery, as demonstrated by this case.[7] MCALVF is believed to result from persistence of the embryological myocardial microcirculation (myocardial sinusoids) leading to arterio-cameral communications.[7] These are distinct from the Thebesian veins which connect the myocardial venous system to the cardiac cavities. The connections can be a single large connection or a series of small fistulas forming a complex network. The high flow and low resistance cause these communications to develop and enlarge over time.[7]

Although surgical ligation of the supplying vessels in localised areas of fistulous communication or transcatheter vascular occlusion with permanent occlusive agents are the accepted treatment options,[8] the treatment of coronary fistulas is less clear when there are multiple communications. This case provides a further dilemma as there is a possibility of abnormally developed venous run off. We have not been able to demonstrate the mechanism of venous return from the myocardium. We speculate that the 'apparent absence' of the coronary sinus is a result of significantly reduced myocardial venous return due to the extensive fistulous connection to the left ventricle. True atresia or unroofing of the coronary sinus is normally associated with atrial isomerism and persistence of the left superior vena cava and other cardiac malformations,[9] excluded in this case.

Despite evidence of ischaemia, medical therapy has maintained this lady with stable angina over a 10-year period. Symptomatic treatment with medical therapy appears to have been the best alternative in this case.

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