# Coarctation of the aorta: a life-long disease of the entire vascular system

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**Abstract**

**Coarctation of the aorta is an important differential diagnosis in adults with hypertension. Unfortunately, simply removing the obstruction does not restore cardiovascular normality. Patients may continue to be hypertensive, demonstrate abnormalities of endothelial function and remain at risk of premature coronary artery disease and other vascular disease. Therapy therefore requires both relief of the mechanical obstruction and long-term follow-up to deliver optimal antihypertensive therapy, vascular risk factor modification and detection and management of complications (such as bicuspid aortic valve and cerebral aneurysms). This paper discusses the management of three cases of this condition.**

**Key words:** adult congenital heart disease, coarctation of the aorta, hypertension.

## Patient 1

A 55-year-old woman presented to her general practitioner complaining that her head was being "blown off". She was found to be severely hypertensive (250/130 mmHg) and was referred for further assessment. She had an ejection systolic murmur and absent femoral pulses. A diagnosis of coarctation of the aorta with associated bicuspid aortic valve was made. The coarctation was confirmed by the MRI scans (figure 1). The black area seen just after the coarctation is due to the high velocity jet of blood that has moved into the imaging plane which has not been excited by the radiofrequency energy.

Cardiac catheterisation was performed to evaluate her coronary arteries that were normal. An aortogram was also obtained (see figure 2) as a digital subtraction image. Her hypertension was treated medically and she was referred to a cardiac centre for surgical repair. Post-operatively she did well and her blood

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**Figure 1.** MRI scan of patient 1 showing coarctation at the classic site

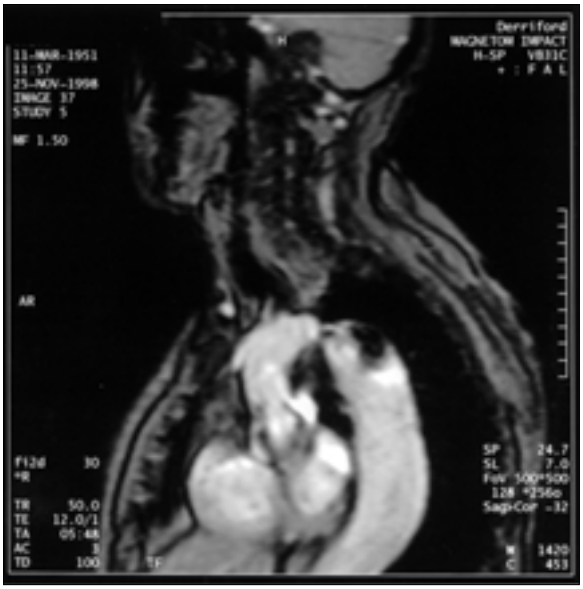



**Figure 2.** Aortogram of patient 1 also showing the coarctation and dilated head and neck vessels

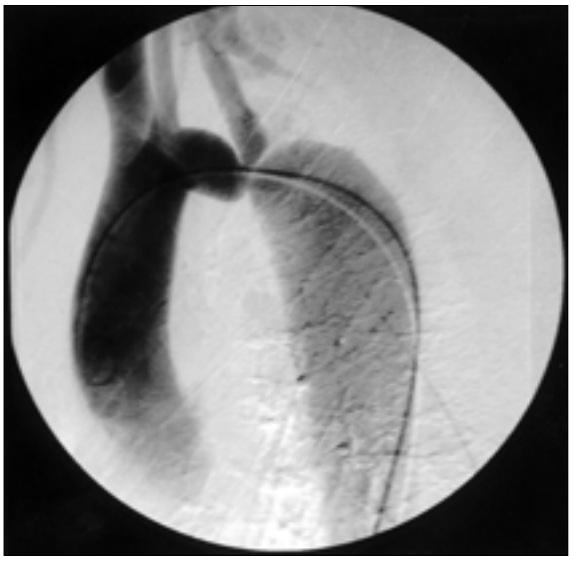


**Figure 3.** Aortogram of patient 2 prior to intervention

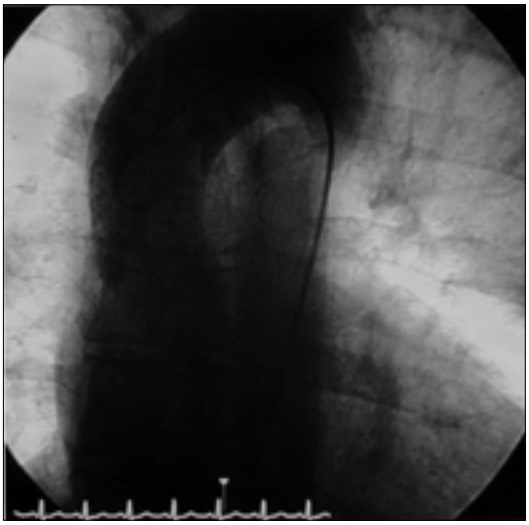
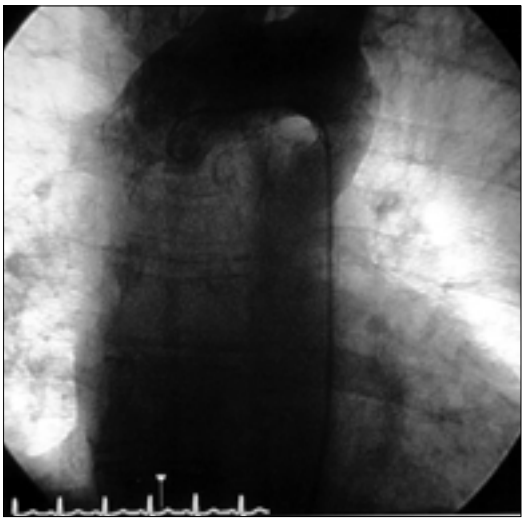


**Figure 4.** Aortogram after stent implantation showing some residual narrowing but relieving the obstruction



pressure improved but she continued to require antihypertensive drugs.

Several years later she suffered further headaches and a MRI scan of her head was performed. This showed a large aneurysm, which was not amenable to surgery and was deemed to be too high risk for percutaneous intervention. The estimated risk of rupture is 1% per year and therefore the neurosurgeon has elected to observe the aneurysm for any rapid increase in size; her blood pressure is also being controlled aggressively.

## Patient 2

A 62-year old man who had been treated for hypertension for many years was found to have coarctation of the aorta during pre-operative assessment for orthopaedic surgery. He suffered with bilateral leg claudication and non-specific sub-mammary chest pain, unrelated to exertion. His upper limb pulses were easily palpable but his femoral pulses were weak. Blood pressure was 178/88 mmHg and there was a systolic murmur audible over the scapulae. His ECG showed left ventricular hypertrophy with some repolarisation abnormality and evidence of left atrial enlargement. Cross-sectional imaging showed his coarctation to be discrete and some distance from the left subclavian artery origin. Due to the increased risk of coronary disease in coarctation patients,[1] coronary angiography was performed and demonstrated normal epicardial coronary arteries but with evidence of mild left ventricular dysfunction. As the lesion was technically suitable, he was offered stenting of the coarctation instead of surgery. Prior to stenting, the gradient across the coarctation (during general anaesthesia) was 40 mmHg. A Jomed non-covered stent was implanted using a 23 mm balloon. This abolished the pressure gradient across the coarctation. The pre- and post-procedural aortograms are shown (see figures 3 and 4). Long-term therapy will include antihypertensive drugs and cholesterol-lowering measures.

## Patient 3

A 49-year-old woman was referred by her general practitioner because of hypertension, systolic murmur and a previous diagnosis of coarctation of the aorta. She had not been followed-up and also described palpitations. She had an ejection systolic murmur with a slow-rising pulse. Femoral pulses were palpable but radio-femoral delay was detected. Blood pressure was 140/80 mmHg despite antihypertensive therapy. Echocardiography demonstrated a thickened bicuspid aortic valve with severe aortic stenosis, coarctation and left ventricular hypertrophy. CT showed this to be a severe coarctation with virtual obliteration of the lumen. Due to the increased prevalence of coronary disease among patients with coarctation, she underwent coronary angiography from the right radial artery that demonstrated normal coronaries and severe coarctation.

She was referred for surgery to her aortic valve and repair of her coarctation. The surgeon used a median sternotomy incision and a mechanical aortic valve to relieve the aortic stenosis. In order to avoid a left thoracotomy to access the coarctation, it was elected to bypass the narrowing using a conduit from the ascending aorta to the mid-thoracic descending aorta. Post-operatively her blood pressure was 120/70 mmHg on a combination of an ACE inhibitor and thiazide diuretic.

## Discussion

Coarctation of the aorta can present in childhood with heart failure and absent femoral pulses. It may also present in adulthood, as in the patients described, most commonly with hypertension that may be malignant. Other presentations include: cerebral haemorrhage, coronary artery disease, aortic stenosis due to

**Key messages**

- Coarctation in adults usually presents as hypertension
- It is associated with bicuspid aortic valve and cerebral aneurysms
- All patients should be followed up for local and remote complications
- Coarctation is a life-long disease of the entire vascular system

bicuspid aortic valve, or as a chance finding. In young children, femoral pulses may be absent but in adults they are often present (due to the formation of collateral vessels). All hypertensive patients should be examined carefully for radio-femoral delay that is usually present but can be subtle. Some cases will be identified on chest X-ray or on echocardiography. All patients with a bicuspid aortic valve should have imaging of the aortic arch by echocardiography or other modality to exclude coarctation.

Whilst a definite diagnosis can often be made by echocardiography, cross-sectional imaging with MRI or CT gives better anatomical information. Aortography is rarely necessary unless cardiac catheterisation is indicated for other reasons. Assessment of the severity of coarctation can be difficult but it should be considered significant if it is causing hypertension (even if only on exercise) or if the pressure gradient is > 20 mmHg.

Therapeutic options in adults are surgical, percutaneous stenting[2] or medical therapy alone to control hypertension. Relief of the mechanical obstruction is highly desirable but is not curative as the risk of complications remains. Even when repair is undertaken early, vascular dysfunction remains.[3] Optimal blood pressure control is necessary. Patients may retain a tendency to develop severe hypertension on exercise that can be detected on a treadmill test. An exaggerated hypertensive response to exercise also warrants aggressive antihypertensive treatment and investigations to confirm that there is no residual obstruction following repair. A holistic response to vascular risk factors is important and may include drug therapy for hypercholesterolaemia, smoking cessation, healthy diet and avoidance of obesity.

Long-term complications of surgery or stenting also need to be considered. Some repairs are at risk of aneurysm formation or recoarctation, necessitating regular routine cross-sectional imaging (CT/MRI). Thus, all patients who have had coarctation (even those repaired as young children) need long-term follow-up to optimise their risk factor profile and to monitor for other treatable complications. Coarctation is not merely an anatomical obstruction – it is rather a life-long disease of the entire vascular system.

## Editors' note

This article continues our series on grown-up congenital heart (GUCH) disease, which began in the February issue. Previous articles included:

- adult congenital heart disease: time for a national framework (*Br J Cardiol* 2002;**9**:65–67)
- grown-up congenital heart disease: experience in a district general hospital (*Br J Cardiol* 2002;**9**:92–98)
- atrial septal defects: a differential diagnosis for breathlessness in adults and the elderly (*Br J Cardiol* 2002;**9**:99–102)
- patent foramen ovale: a normal variant or a congenital abnormality requiring treatment? (*Br J Cardiol* 2002;**9**: 223–5).


## Acknowledgement

We are grateful to Dr J De Giovanni for his help with patient 2.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