LESSON OF THE WEEK

Late hazards after repair of coarctation of the aorta

Kunadian Vijayalakshmi, Amanda Griffiths, Asif Hasan, John O'Sullivan

Freeman Hospital, Newcastle upon Tyne Hospitals NHS Trust, High Heaton, Newcastle upon Tyne NE7 7DN

Correspondence to: K Vijayalakshmi
kunadianvijay@aol.com

BMJ 2008;336:772-3
doi:10.1136/bmj.39472.693356.47

Asymptomatic patients who have had surgical correction of coarctation of the aorta need regular monitoring to prevent life threatening complications

Introduction

Coarctation of the aorta is a relatively common type of congenital heart defect. It comprises 5-8% of all congenital cardiac defects. The first coarctation repair was carried out in 1944.¹ Children operated on in the 1960s, 70s, and 80s are now adults, and it is not known how many are receiving regular specialised medical follow-up.² Aneurysms of the aorta or in the region of the aortic isthmus are the most dangerous complications because they carry the risk of life-threatening rupture, which may be the first symptom.

We describe two patients who underwent surgical repair of coarctation of the aorta in their childhood and were lost to follow-up. Both patients presented as adults with non-specific symptoms and investigations found aneurysm of the aorta. Both patients underwent successful urgent surgery.

Case 1

A 42 year old woman presented to a district general hospital with pain in the back after a trip to the supermarket. Musculoskeletal pain was initially diagnosed and she was treated with analgesics. She did not have any important medical history except repair of coarctation of the aorta (Dacron patch repair) at the age of 16 years. Coarctation of the aorta had been diagnosed incidentally at age 4 years when her general practitioner identified a heart murmur on routine clinical examination. She had been followed up routinely until age 16 and continued to be free of symptoms. She had been given the option of having surgery at the age of 16 or leaving it until she was older if she needed it (probably in her 40s) and to continue with annual check-ups. She was advised at the time that the benefits of having surgery at age 16 were that younger people recover more quickly from surgery and once she had the surgery she would no longer need to attend any "heart" clinics, so she did not have routine follow-up under the care of a cardiologist after the surgery.

She had bicuspid aortic valve, so on this admission had a computed tomography scan of her chest, which showed that the proximal descending aorta was markedly aneurysmal, with a saccular configuration

measuring 65 mm just proximal to the previous surgical site. There was no evidence of leak, dissection flap, or dissection of ascending aorta or aortic root, but the aortic root was dilated (fig 1). An echocardiogram showed moderate aortic regurgitation through the bicuspid valve. Repair of the descending thoracic aneurysm was successful and she now has regular specialist follow-up.

Case 2

A 38 year old man presented to the chest clinic with haemoptysis. In 1968, when he was 18 months old, a heart murmur had been identified, coarctation of the aorta had been diagnosed, and an end to end resection of coarctation of the aorta had been done. On routine follow-up visits his femoral pulses were weak and his blood pressure was raised. His raised blood pressure was treated with propranolol 80 mg twice a day. Further investigation with cardiac catheterisation showed a gradient of 20 mm Hg across the coarctation, but clinically the coarctation had seemed more severe than catheterisation findings; hence he had been referred for surgery at the age of 10 years. He was lost to follow-up after age 21, when he understood he did not require continuing surveillance. He worked as a builder until this episode of haemoptysis.

His bronchoscopy and lungs on computed tomography pulmonary angiography on this admission were essentially normal. However the imaging showed a small 10 mm aneurysm of the aortic arch, close to the anastomosis. A 3D reconstruction of his images showed that the pseudoaneurysm involved the

Fig 1 | Aneurysm (arrow) of the descending aorta

Fig 2 | Pseudoaneurysm (arrow) of the aorta close to the subclavian artery and the left lung

left subclavian artery (fig 2). This case was discussed at the multidisciplinary team case conference. Because the pseudoaneurysm was very close to the subclavian artery, and given his employment as a builder, it was decided to treat him surgically rather than by percutaneous approach. Resection of the pseudoaneurysm and repair using a Gore-tex patch was successful, and he continues to work as a builder and is followed up routinely by the congenital heart disease team.

Discussion

Several studies have shown the importance of long term follow-up after repair of coarctation of the aorta.³⁻⁸ Late cardiovascular complications after operative repair of coarctation of aorta include systemic hypertension, premature coronary artery disease, aortic valve abnormalities, aortic aneurysm, and re-coarctation. As these may not produce symptoms until a major complication occurs, it is essential to monitor blood pressure in these patients. Aortic aneurysms are a particular concern and are an important cause of morbidity and death long after the operation in patients with repaired aortic coarctation.

Recent studies show that postoperative aortic aneurysm is found in only 5-9% of patients.⁹ Some anastomotic aneurysms have been found more than 30 years after operation. In many patients aneurysms are detected as an incidental finding, because they seldom produce any symptoms. They can develop after balloon angioplasty for coarctation of the aorta. The reported frequency of aneurysms after angioplasty in adult coarctation varies from 4% to 12%. Aneurysms may develop immediately after angioplasty or several months later, but even major tears caused by angioplasty may diminish and disappear without aneurysm forming.

The patients described here represent a failure of surveillance, and others with a similar history may not receive routine surveillance. Since 1990, children operated on for coarctation of the aorta would be moved on to adult services for follow-up. The number of patients operated on before the 1980s and not undergoing regular check-ups is unknown. The patients, their families, general practitioners, and the general medical and nursing community need to be aware that such patients, although probably clinically well, need regular expert follow-up to prevent complications.

Conclusion

As the incidence of aneurysm after surgery or balloon dilatation seems to increase with longer periods of follow-up, all patients need careful follow-up after angioplasty and surgical procedures. Local expert care by a cardiologist specialising in congenital heart disease allows earlier and more aggressive treatment of complications and may also improve compliance. Where a local specialist is not available, general cardiologists need protocols and easy access to specialist support.

KV and AG cared for the patients on the ward and in the outpatients' department; JO'S was the consultant in charge of both the patients; AH operated on both patients. KV prepared the manuscript, which was then reviewed and revised by all the authors. KV is guarantor.

Funding: No special funding.

Competing interests: None declared.

Provenance and peer review: Not commissioned; externally peer reviewed.

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Accepted: 10 June 2007

Endpiece Learning how to die

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Seneca. *On the Shortness of Life*. Chapter VII