An unexpected finding late after repair of coarctation of the aorta

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We describe a late complication in a 75-year-old man 50 years after repair of a coarctation of the aorta (CoA). Two years after an aortic valve replacement, mitral valve repair and radiofrequency MAZE the patient presented with dyspnoea and right-sided heart failure, based on a large pseudoaneurysm of the descending aorta, compressing the main bronchus and possibly temporarily the pulmonary arterial system. After sealing the aneurysm with an endovascular stent the patient recovered uneventfully. Recommendations are made for follow-up in patients after repair of CoA. (*Neth Heart J* 2008;16:260-3.)

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Coarctation of the aorta (CoA) is a narrowing of the aorta usually in the area of the ligamentum arteriosum.¹ A CoA is considered significant if an invasively determined pressure gradient of more than 20 mmHg is present. When left untreated CoA can cause left ventricular pressure overload leading to left ventricular hypertrophy, premature coronary artery disease and eventually heart failure.² Nowadays, CoA is usually diagnosed in childhood. However, after successful repair of a CoA, there are many associated

problems and long-term complications that can occur even decades later. Cardiologists should be aware of these complications. We would like to illustrate this with the following case report.

Case report

A 75-year-old man was admitted to our cardiology department because of progressive shortness of breath and peripheral oedema. His medical history revealed repair of a CoA at the age of 24 years via an end-to-end anastomosis of the aorta and ligation of the left subclavian artery. Two years before presentation, a

Figure 1. Left anterior oblique magnetic resonance angiogram before aortic valve replacement. Two small aneurysms at the coarctation repair site are shown (arrows).

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stenotic calcified bileaflet aortic valve (aortic valve area 0.8 cm^2) had been replaced using a bioprosthesis. During the same procedure mitral valve annuloplasty and radiofrequency MAZE were performed because of significant mitral valve insufficiency and atrial fibrillation, respectively. Two small aneurysms (14 and 20 mm) were seen on magnetic resonance images (MRI) of the aorta performed before this operation (figure 1).

After admission treatment was initiated with diuretics. Echocardiography showed right atrial and right ventricular dilatation along with severe tricuspid regurgitation. The estimated systolic pulmonary artery pressure was more than 60 mmHg. The function of the aortic valve bioprosthesis was within normal limits and mitral valve insufficiency was trivial. Images from the suprasternal window were suboptimal. Computed tomography (CT) showed narrowing at the site of coarctation repair and a large pseudoaneurysm with a diameter of more than 9 cm arising 3 cm distally, compressing the left main bronchus (figure 2). No clear compression of the pulmonary arterial system was seen.

Subsequently a thoracic endoprosthesis was inserted (figure 3). After stent placement a pressure gradient of 2 mmHg was measured across the site of the coarctation repair. The patient recovered uneventfully. A CT scan of the aorta six months later showed a well functioning endovascular stent without signs of leakage. On repeated echocardiography right ventricular diameters and pulmonary artery pressure normalised.

Discussion

CoA is a narrowing of the aorta preferentially just distal of the left subclavian artery and accounts for 6 to 8% of all congenital heart defects.^{3,4} This malformation occurs up to five times more frequently in males than females and is associated with ventricular septal defect (30 to 45%), bicuspid aortic valve (30 to 85%), patent ductus arteriosus, mitral stenosis and aortic stenosis (valvular, subvalvular, or supravalvular).^{5,6} Intracerebral aneurysms and Turner's syndrome are extracardiac defects associated with coarctation of the aorta.²

Significant CoA is defined as a gradient of more than 20 mmHg with or without proximal hypertension or the presence of (any degree of) coarctation associated with proximal hypertension.¹ The aetiology of CoA is usually congenital or less commonly acquired. The congenital form is thought to develop from ductal arteriosus tissue extending into the thoracic aorta or underdevelopment of the aortic arch by reduced antegrade blood flow.⁷ Occasionally inflammatory diseases of the aorta such as Takayasu arteritis or severe atherosclerosis can underlie acquired aortic narrowing, in which predominantly the midthoracic or abdominal aortic region is involved.⁸⁻¹⁰

Adults with previously undiagnosed CoA can present with severe hypertension and associated symptoms such as headache, heart failure, epistaxis or

Figure 2. CT angiogram of the patient before endovascular stenting of the descending aorta. A large pseudoaneurysm is present ($9 \times 5 \text{ cm}$) (white arrows), with calcification and signs of old and new bleeding, also compression of the left main bronchus is seen (black arrow).

Figure 3. CT reconstruction of the aorta after placement of a thoracic endoprosthesis.

aortic dissection.^{4,11} Physical examination in patients with an asymptomatic CoA may reveal continuous murmurs, as well as palpable pulsations from large collateral vessels. An arm-leg pressure gradient of at least 10 mmHg and diminished femoral pulses may be present. A systolic ejection murmur and ejection click may indicate the presence of a bicuspid aortic valve. Notching of ribs on chest X-ray by erosion from

Table 1. Complications and abnormalities that can occur (late) after repair of coarctation of the aorta (adapted from Pieper et al. and Swan et al.).^{2,3}

Residual or re-coarctation
Dilatation of the ascending or descending aorta
Aortic dissection
Formation of an aortic pseudoaneurysm
Degeneration of an associated bicuspid aortic valve
Hypertension
Premature coronary artery disease, atherosclerosis
Cerebral haemorrhage associated with intracranial aneurysms
Endarteritis

large collateral intercostal arteries may occur.¹² Also, a visible indentation at the site of coarctation can produce the '3-sign'.¹³ CoA can be diagnosed with echocardiography. CT, MRI or alternatively aortography are imaging modalities that are often diagnostic if echocardiography is not satisfactory.

After coarctation repair: healed but not cured

Even after successful repair of a CoA cardiologists should be aware of complications that can occur sometimes decades later (table 1).^{2,3,14} The prevalence of recoarctation reported in the literature varies widely and depends on the interventional technique (surgery or transcatheter) used.¹ Balloon angioplasty has been shown to be superior to repeated surgery with regard to safety, mortality and morbidity.^{15,16} True aneurysms but also pseudoaneurysms late after repair have been reported. Anastomotic pseudoaneurysms develop in 3 to 38% of patients.¹⁷ Endovascular stenting is a relatively new technique and is associated with lower mortality, relatively low morbidity, and good mid-term follow-up with regard to aneurysm-related death.¹⁸⁻²¹ Also aortic dissection at the repair site, both late after surgery and after balloon dilatation, have been documented.

Recommended follow-up after repair of coarctation of the aorta

All patients should be periodically followed up by a local cardiologist or at a centre for congenital cardiology and should preferably be registered at a central database (in the Netherlands CONCOR database).²² At least one CT, MRI or angiogram should be performed after the CoA repair. Once a year echocardiography, electrocardiography and an exercise test (in search for hypertension on exercise and inducible repolarisation abnormalities) should be performed. Annual MRI or CT imaging has been proposed.³ Attention should be paid to signs of heart failure, presence of hypertension, valve abnormalities such as a bicuspid aortic valve and dilatation of the ascending or descending aorta. Also (pseudo)aneurysms or late dissection proximal or distal to the repair site should be considered. Besides hyper-

tension other risk factors for coronary artery disease must be treated vigorously. Screening for intracerebral vascular anomalies should be performed in case of new or unusual headaches.^{22,23}

In the present case, the patient presented with dyspnoea and signs of right-sided heart failure, based on a large pseudoaneurysm of the descending aorta 50 years after surgical repair of the CoA and two years after replacement of a calcified bileaflet aortic valve, mitral valve annuloplasty and radiofrequency MAZE. Whether replacement of the aortic valve changed the haemodynamics in favour of growth of the pre-existent aneurysm is not clear, but enhancement of pulsatile forces could have caused a deterioration of the small aneurysm and increased the chance of pseudoaneurysm formation. Our patient presented with right-sided heart failure. The CT scan did not show compression of the main or peripheral pulmonary arteries nor stenosis of a pulmonary vein. Perhaps a certain mass effect of the large pseudoaneurysm on the pulmonary artery was at first present, which resolved after initiation of therapy with diuretics. ■

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