Chronic aortic disease

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Abstract

Chronic aortic disease encompasses thoracic and abdominal aortic aneurysms, including chronic dissection, genetic aortopathies and post-intervention states after surgical or endovascular repair. Most aortic aneurysms are degenerative and histopathologically involve cystic medial degeneration and atherosclerosis. Surgery is currently indicated where the maximum aortic diameter is >55 mm, but lower thresholds are appropriate if there are additional risk factors. A variety of techniques – surgical, endovascular and hybrid – can be used depending on the aneurysm site and patient factors. The mortality rate for elective proximal aortic surgery is <1.5%.

Keywords Aneurysm; aortopathy; Dacron graft; frozen elephant trunk procedure; thoracic endovascular aneurysm repair (TEVAR)

Key points

- Most aortic aneurysms are degenerative and histopathologically involve cystic medial degeneration and atherosclerosis
- Surgical repair is indicated where the maximum aortic diameter is >55 mm, but lower thresholds are appropriate if there are additional risk factors
- The type of operation performed depends on the aneurysm location, aortic valve morphology and patient's age
- Advances in endovascular technology have resulted in an increasing number of thoracic endovascular aneurysm repair and hybrid procedures
- Postoperatively, blood pressure control is key, using a combination of β-blockers and angiotensin-converting enzyme inhibitors/angiotensin receptor blockers

Introduction

Chronic aortic disease represents a diverse spectrum encompassing thoracic and abdominal aortic aneurysm, chronic aortic dissection, genetic aortopathies and post-intervention states after surgical or endovascular repair. The natural history is heterogeneous, from indolent slow-growing degenerative aneurysms to rapidly progressive genetic disorders with a high risk of rupture.

The 2024 European Association for Cardio-Thoracic Surgery/Society of Thoracic Surgeons guidelines[1] have recognized the aorta as an organ and provide a framework to mark a shift towards a life-long, systematic approach to aortic pathology, integrating comprehensive risk assessment, refined imaging surveillance and lower thresholds for intervention.

Anatomy

The thoracic aorta can be divided into four different components (Figure 1), each posing different challenges and treatment strategies.

Aortic root – forms a bridge between the left ventricle and the ascending aorta and includes the aortic valve annulus, aortic valve cusps and sinuses of Valsalva.

Ascending aorta – the tubular portion of ascending aorta, running from the sinotubular junction to the origin of the brachiocephalic (innominate) artery.

Aortic arch – originates just proximal to the origin of the brachiocephalic artery and terminates just beyond the left subclavian artery.

Descending aorta – begins at the isthmus between the origin of the left subclavian artery and the ligamentum arteriosum and passes through the diaphragm into the abdomen.

Epidemiology

Thoracic aortic aneurysms are defined as the enlargement of the thoracic aorta to >1.5 times normal diameter. In the UK the incidence is about 6 per 100,000/year and there is a male preponderance.

Classifications of aortic aneurysms are based on the anatomy, i.e. aortic root, ascending, arch or thoraco-abdominal aneurysm. Aneurysms can involve one segment in isolation or several different segments. Approximately half of aneurysms involve the aortic root or ascending aorta.

Most aortic aneurysms are degenerative and histopathologically involve cystic medial degeneration and atherosclerosis. Risk factors for the development of thoracic aortic aneurysms include hypertension, smoking, atherosclerosis and chronic obstructive pulmonary disease.

Other causes include hereditary aortic aneurysms associated with connective tissue disorders such as Marfan syndrome, Ehlers–Danlos syndrome and aortopathies occurring with bicuspid aortic valves. Inflammatory aneurysms include Takayasu arteritis and Kawasaki disease. Infective causes such as mycotic and syphilitic aneurysm are not seen commonly in the UK.

Although hereditary aneurysms often show aggressive progression, contemporary studies demonstrate that many degenerative aneurysms grow much more slowly, often at <0.2 mm/year.[2] Overall, the average rate of expansion of thoracic aortic aneurysms is estimated to be 0.10–0.42 cm/year. As an aneurysm expands, the rate of expansion per year also increases, as does the risk of rupture.

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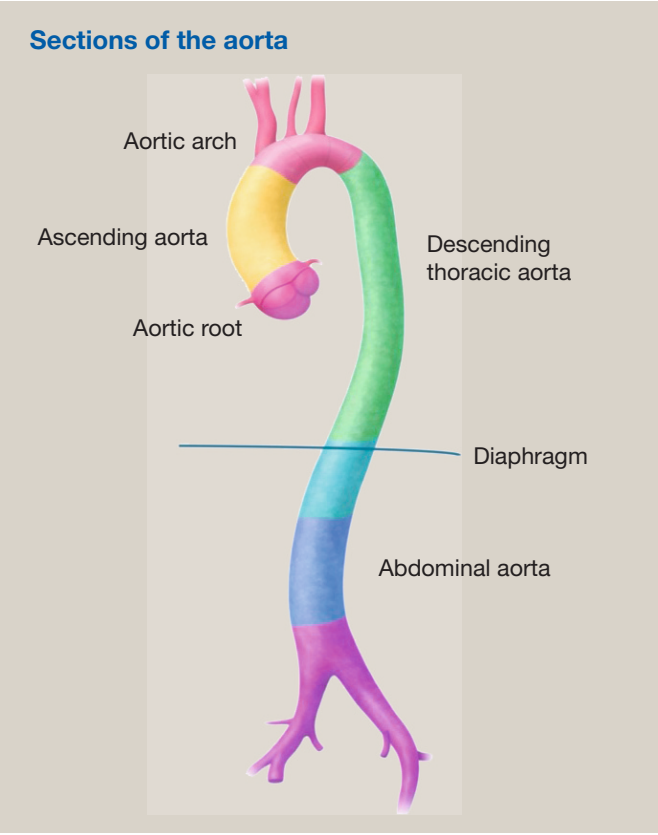

Figure 1 Sections of the Aorta. Adapted from reference 1.

Clinical presentation

There has been an increasing trend of thoracic aneurysms being diagnosed incidentally, commonly in the context of screening for lung cancer. These patients are referred to aortic surveillance clinics.

Other presentations include symptoms from a mass effect or direct compression of the aneurysm on adjacent structures (e.g. hoarseness from stretching of the recurrent laryngeal nerve, stridor from tracheal compression). There can be embolization and ischaemia from haematoma or debris. Symptoms can also arise from associated aortic valve regurgitation. In addition, patients can present with acute aortic syndromes such as acute aortic dissections, intramural haematomas and penetrating aortic ulcers.

Diagnosis

Contrast-enhanced computed tomography (CT) is the main imaging modality for thoracic aortic aneurysms. Magnetic resonance imaging (MRI) is preferred in younger patients with hereditary aortopathies, who often need multiple scans, to minimize the radiation risk.

Transthoracic echocardiography is useful in detecting aneurysms in the aortic root and proximal ascending aorta. It is less useful for aneurysms of the distal ascending aorta and aortic arch.

Management

The primary goal in the management of thoracic aortic aneurysms is to prevent the fatal complications of aortic rupture and aortic dissection. Medical management involves blood pressure control and reduction of atherosclerotic risk factors, including smoking cessation.

The aortic diameter is the primary aortic dimension used to guide the timing for surgery. To define the optimal threshold for surgery the natural history with regard to complications must be weighed against the operative risks. In view of recent evidence showing that the aorta may dissect or rupture at a lower diameter (i.e lower hinge point), there has been a shift towards intervening at lower thresholds. Furthermore, the mortality rate for elective proximal aortic surgery has decreased to $<1.5\%$ over recent decades.

Surgery is currently indicated for maximum aortic diameters of $\geq$55 mm in all patients with an acceptable surgical risk. Lower thresholds of 50–52 mm are considered in individuals with an aortic root phenotype dilatation (i.e. aortic root dimension larger than that of the ascending aorta), bicuspid aortic valves and certain risk factors; these include age $<$50 years, short stature $<$1.69 m, ascending aorta length $>$11 cm, aortic diameter growth rate $>$3 mm/year, a strong family history or refractory hypertension.

A diameter of $\leq$50 mm is an indication for surgery in hereditary aortopathies that have a more aggressive natural history and in patients undergoing concomitant non-aortic surgery. Figure 2 summarizes the thresholds for intervention in different aortic phenotypes and associated risk factors.

Surgical options

The type of operation depends on the location and extent of the aneurysm, the morphology of the aortic valve and the patient's age.

In isolated ascending aortic aneurysms, the aorta can be replaced by a Dacron graft. With aortic root aneurysms, an aortic root replacement with a composite of a valve and conduit (modified Bentall procedure) can be performed. The choice of the valve prosthesis depends on the patient's age for biological valves in older patients and mechanical valves in younger patients. If the proximal arch is dilated, a hemi-arch graft can be performed using an extended Dacron graft.

A valve-sparing root replacement can also be performed, especially in younger people. Procedures include the David reimplantation technique and Yacoub remodelling technique.[3,4] Concomitant procedures on the mitral valve (mitral valve pathologies are seen more often in individuals with Marfan disease) and coronary artery bypass grafting can be performed in the same setting.

If the entire arch needs to be replaced, a 'frozen elephant trunk' (FET) procedure can be performed.[5] This is a composite of a stented graft (guided into the descending thoracic aorta) and a Dacron graft (for the ascending aorta) with side branches for reimplantation of the head and neck vessels.

Endovascular and hybrid options

Advances in endovascular technology have resulted in an increasing number of thoracic endovascular aneurysm repair (TEVAR) and hybrid procedures, especially in the context of thoraco-abdominal aneurysms. This avoids the need for a thoracotomy incision, cardiopulmonary bypass or clamping of a diseased and atherosclerotic aorta. Endovascular options are especially beneficial in older individuals with multiple

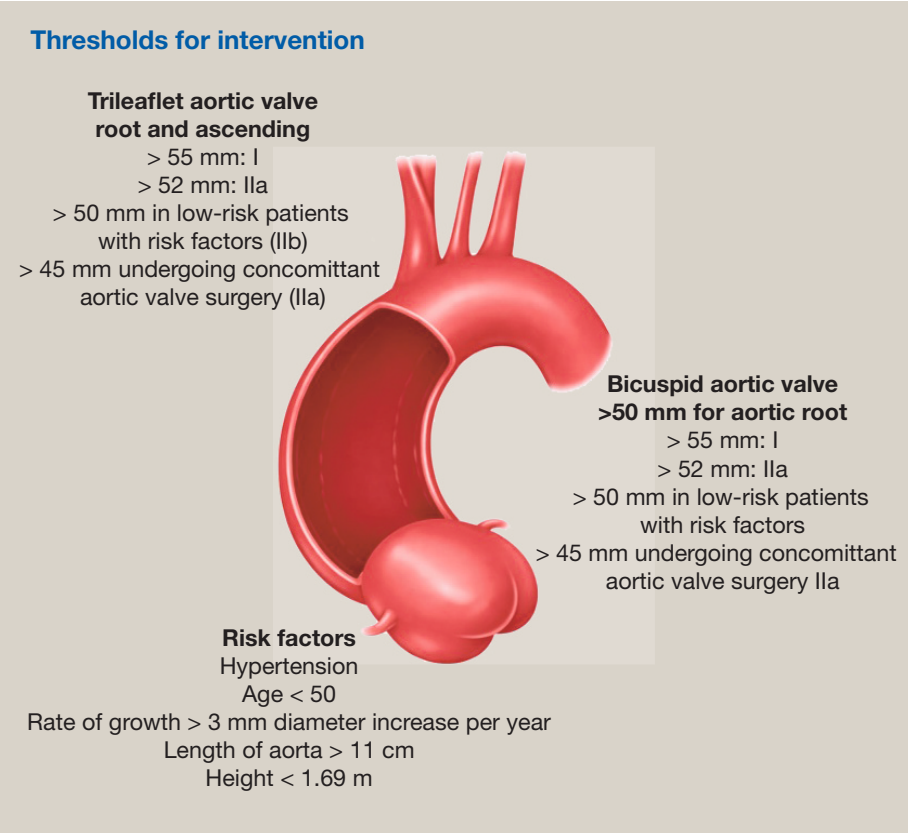

Figure 2 Thresholds for Intervention. Adapted from reference 1.

co-morbidities and a heavy calcific/atherosclerotic burden in the aorta.

Hybrid options are also available to manage an aneurysmal arch and thoraco-abdominal aorta. For example, an initial FET procedure can be followed by a completion TEVAR approach to treat the descending thoracic and abdominal aorta.

Results and postoperative management

The results of aortic surgery have improved significantly over the last decade, with mortality rates approaching 0.25% in large-volume centres. Postoperatively, blood pressure control is key, with a combination of β-adrenoceptor blockers and angiotensin-converting enzyme inhibitors/angiotensin receptor blockers. Continued surveillance of the aorta with CT/MRI scans at regular intervals is an essential part of postoperative management.

Relatives of patients with hereditary aortopathies should also be screened for aortic aneurysms. ◆

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TEST YOURSELF

To test your knowledge based on the article you have just read, please complete the questions below. The answers can be found at the end of the issue or online here.

Question 1

A 68-year-old man was reviewed for monitoring of a thoracic aortic aneurysm and bicuspid aortic valve. He was taking ramipril 5 mg od for hypertension. His mother had died of a myocardial infarct aged 88. On examination his blood pressure was 155/92 mmHg. Echocardiographic and CT imaging revealed a dilated ascending aorta with a maximum diameter of 46 mm with normal arch and thoracoabdominal aortic dimensions. Imaging 1 year previously had revealed a maximum aortic dimension of 44 mm.

What aspect of this case would most strongly support elective aortic surgery over continued monitoring?

A. Bicuspid aortic valve
B. Family history
C. Hypertension
D. Rate of aortic dilation
E. Site of maximum aortic dilation

Question 2

A 58-year-old man was due to undergo elective surgery for a thoracic aortic aneurysm involving the ascending aorta, aortic arch and proximal descending thoracic aorta. Cardiovascular physical examination was normal.

What is the most appropriate surgical technique for his procedure?

A. David reimplantation technique
B. Dacron graft with mechanical aortic valve replacement
C. Frozen elephant trunk procedure
D. Thoracic endovascular aneurysm repair (TEVAR)
E. Yacoub remodelling technique

Question 3

A 34-year-old woman was being monitored in the clinic for an aneurysm of the proximal descending thoracic aortic.

What is the most appropriate imaging technique for sequential monitoring in this situation?

A. CT scanning
B. Transthoracic echocardiography
C. Transoesophageal echocardiography
D. MRI scanning
E. Roentgenography