Aortic valve disease

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Abstract

Aortic valve disease encompasses aortic stenosis, aortic regurgitation, and congenital malformations. Aortic valve stenosis is a common problem among the elderly, and valve replacement is still the only treatment option for severe symptomatic disease. Advances in our understanding of the pathophysiology of calcific aortic valve disease support the presence of an active disease process. Congenital bicuspid aortic valves account for half of all valve replacements and are associated with aortic dilatation. Echocardiography remains an important diagnostic tool in management of aortic valve disease and is recommended in all patients with suspected disease.

Keywords Aortic valve disease; aortic valve insufficiency; aortic valve stenosis

What's new?

- The 2014 American and 2012 European valvular guidelines both recommend earlier aortic valve interventions for asymptomatic low-surgical risk patients with severe disease and high-risk factors
- A large randomized clinical trial of an oral direct thrombin inhibitor showed excess harm for anticoagulation in mechanical heart valves
- Trans-catheter aortic valve implantation for symptomatic severe calcific aortic stenosis is currently recommended in inoperable patients and is a non-inferior choice to surgery in high-risk patients

Introduction

Aortic valve disease refers to any damage or dysfunction of the aortic valve and encompasses a wide range of aetiologies including congenital and acquired disorders. Although there may be various pathological processes, the haemodynamic consequences result from either blood flow obstruction (stenosis), regurgitation, or both. This chapter reviews the evaluation and management of aortic valve stenosis and regurgitation.

Bicuspid aortic valve

A bicuspid aortic valve is the most common congenital cardiac malformation, occurring in 0.5–2% of the population. Nearly all patients with a bicuspid valve develop calcific aortic stenosis by the seventh to eighth decade of life, requiring valve replacement once severe obstruction causes symptoms. A small subset of bicuspid valve patients has significant aortic regurgitation, which requires valve replacement in young adulthood.[1,2] The presence and morphology of a bicuspid valve is associated with aortic dilatation and an increased risk of dissection (Figure 1).[3,4] Computed tomographic (CT) or magnetic resonance aortography is recommended when the ascending aorta is dilated or not well seen on echocardiography. Periodic follow-up imaging is indicated when the aortic diameter reaches 4.0 cm, with annual surveillance if the aorta exceeds 4.5 cm. Bicuspid valves may be familial in up to 30% of patients with variable inheritance patterns. Imaging of first-degree relatives is advisable when the patient has an associated aortic aneurysm or family history of aortic or valvular disease.

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Aortic stenosis

Definition and epidemiology

Valvular aortic stenosis, defined as obstruction to left ventricular (LV) outflow, is most commonly caused by calcific changes of a trileaflet or congenitally bicuspid valve. Aortic stenosis is present in 2–4% of adults over the age of 65 years, but earlier disease without haemodynamic obstruction (called aortic sclerosis) is present in about 25% of older adults.[5] Rheumatic aortic stenosis is common in developing countries but less common in Europe and the USA, and is invariably associated with mitral valve disease. Rare causes of aortic stenosis include systemic lupus erythematous, chest radiation, Fabry's disease and alkaptonuria; these aetiologies should be sought in middle-aged and younger individuals without congenital valvular abnormalities. Approximately 300,000 aortic valve replacements were reported to the American Society of Thoracic Surgeons between 1999 and 2009; half involved concomitant coronary bypass surgery.

Pathophysiology

Calcific aortic stenosis is the end stage of an inflammatory disease process, characterized by the presence of lipid, macrophages and T lymphocytes, and calcium in the valve leaflets. These inflammatory changes are associated with an osteoblast-like cellular phenotype that results in calcification of the valve.[6] Clinical factors associated with the presence and progression of calcific aortic valve disease include hypertension, older age, male gender, hyperlipidaemia, smoking and diabetes mellitus.[7] Evidence for the role of potential genetic risk factors is emerging: a large genome-wide association study discovered that specific variants in genetically determined lipoprotein (a) concentration were associated with incident aortic stenosis.[8]

As valve area decreases, blood flow velocity across the valve increases and the pressure in the left ventricle exceeds the aortic pressure (Figure 2). The average rate of decrease in valve area is 0.1 cm^2/year, with an average increase in aortic velocity of 0.3 m/s/year, but there is wide individual variability (Figure 3). The pressure overload in the left ventricle most often leads to concentric hypertrophy to maintain normal wall stress but also to diastolic dysfunction. Symptoms occur when the flow across the

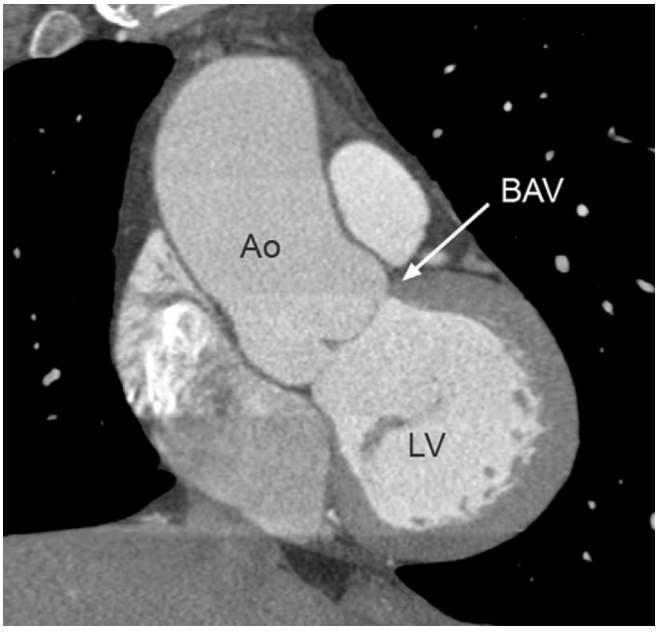


Figure 1 A contrast-enhanced computed tomographic (CT) view of the ascending aorta (Ao) and left ventricle (LV) shows typical aortic root dilatation with a maximum diameter of 56 mm in the 31-year-old man with a bicuspid valve (BAV). Based on current guidelines, he underwent aortic root replacement for aortic dilatation.

narrowed valve can no longer meet the metabolic demands of exercise or ventricular filling pressures rise due to diastolic dysfunction. Symptoms occur later in life in patients with a tri-leaflet valve (age 70–80 years) than in patients with a bicuspid valve (age 50–60 years).

Clinical features

Symptoms: the natural history of aortic stenosis is marked by a long asymptomatic period during which valve changes are evident on echocardiography and a murmur may be noted on auscultation. The onset of symptoms is associated with an ominous prognosis – mortality is more than 50% at 2 years. It is important to note that:

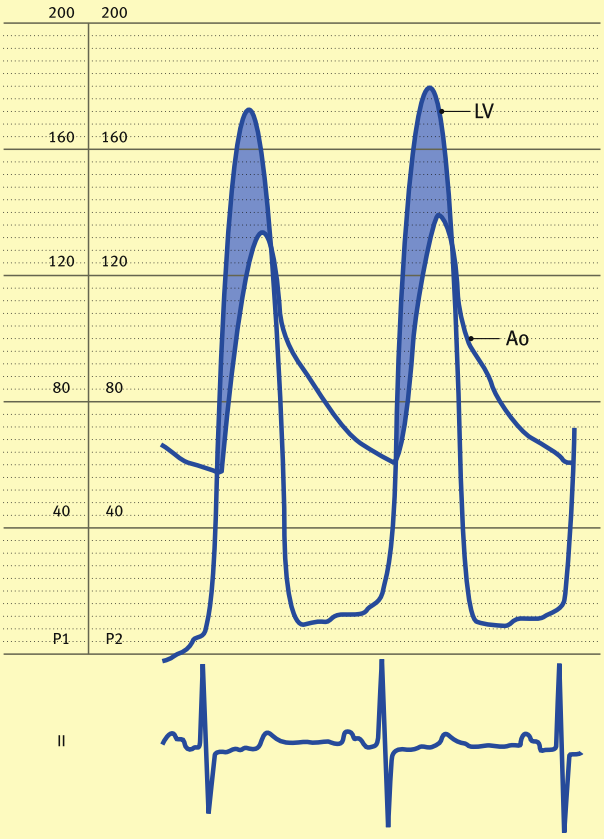
- the classical symptoms of aortic stenosis are angina, dyspnoea and syncope
- most patients present initially with a gradual decrease in exercise tolerance.

Because symptom onset is insidious, careful questioning and comparison of current exercise tolerance with specific past time points are needed to avoid missing significant disease. The rate of symptom onset in asymptomatic patients is predictable, based on the severity of the disease; 80% of those with severe obstruction develop symptoms within 2 years. Sudden death is rare in asymptomatic patients.

Clinical examination: the classical findings on physical examination include:

- a harsh, crescendo–decrescendo systolic murmur, maximal at the base of the heart with radiation to the carotid arteries
- a single second heart sound (S2)
- a delayed and diminished carotid upstroke.

Simultaneous left ventricular (LV) and aortic pressure (Ao) tracings from a 64-year-old male with chest pains and shortness of breath



Shaded area corresponds to the mean pressure gradient which was 36 mmHg. Cardiac output was slightly reduced at 4.6 L/min and calculated aortic valve area by the Gorlin equation came to 0.8 cm^2, consistent with severe aortic stenosis. Ventricular tracings can be identified by the increase in pressure during diastole whereas aortic pressures decrease in diastole. Patient was noted to have multi-vessel coronary artery disease on angiography and was referred for aortic valve replacement and coronary bypass surgery.

Figure 2

These findings are specific, but not sensitive, for severe stenosis. Most patients with severe stenosis have only a murmur and many have an apparently normal carotid upstroke as a result of concurrent atherosclerosis. The only findings on physical examination that exclude severe stenosis are the absence of a murmur or the presence of a physiologically split S2.

Investigations

Echocardiography is essential in patients with a murmur and symptoms that might be caused by aortic stenosis, to visualize valve anatomy, quantify the severity of stenosis, evaluate LV systolic and diastolic function, and assess associated valve abnormalities. Standard measures of stenosis severity include the peak velocity across the valve, the mean pressure gradient and the valve area. Mild stenosis is characterized by a peak velocity of 2.5–3.0 m/s and a valve area of more than 1.5 cm^2, and severe stenosis by peak velocity more than 4.0 m/s and valve area less