Aortic Stenosis

The normal aortic valve is composed of three thin cusps that project from the wall at the origin of the aorta. Aortic stenosis (AS) refers to a tight valve. Aortic sclerosis is usually considered to be the precursor of calcified, degenerative AS but sclerosis is rather more common than stenosis.

Epidemiology

- AS has become the most frequent type of valvular heart disease in Europe and North America. It most often presents as calcific AS in adults of advanced age (2-7% of the population aged over 65 years).
- AS occurs in almost 10% of adults over the age of 80 years, with a mortality rate about 50% at two years unless outflow obstruction is relieved.
- The second most frequent cause, which dominates in the younger age group, is congenital.
- Rheumatic AS has become rare in developed countries.

Risk factors

A congenital bicuspid valve predisposes to both AS and regurgitation.

Presentation

See also the separate Heart Auscultation and Heart Murmurs in Children articles. Aortic valve stenosis is the most common form of valvular heart disease in the elderly population and occurs frequently in conjunction with coronary artery disease.

- Symptoms include shortness of breath on exertion, angina, dizziness or syncope.
- The characteristic murmur may occasionally be faint and so the patient may present as heart failure of unknown cause.
- The disappearance of the second aortic sound is specific to severe AS, although not a sensitive sign.

Symptoms

- Even with a quite marked pressure gradient across the valve, patients may be asymptomatic for many years but they may still be susceptible to sudden events. However, only 4% of sudden cardiac deaths in severe AS occur in asymptomatic patients.
- AS predisposes to angina and is a reason to auscultate the chest when a patient presents with angina. Angina occurs in approximately two thirds of patients with critical AS, of whom 50% have significant coronary artery disease.
- It can also cause syncope on exertion and even sudden death and so violent exertion should be avoided.
- Symptoms of congenital disease often appear between ages of 10 and 20 years.
- The classic triad of chest pain, heart failure and syncope are found in only 30-40% and usually after the age of 50 years.
- Fatigue may be the presentation in children.
- Syncope, especially on exertion, may be caused by arrhythmia or the sudden onset of heart failure with decline in cardiac output.
Signs

- Examination of the pulse in significant AS reveals a slow-rising, flat character called pulsus parvus et tardus. Blood pressure will show a narrow pulse pressure (difference between systolic and diastolic pressures). In the elderly, a rigid aorta may make this sign less obvious.
- Examination of the cardiovascular system includes palpation of the cardiac apex. There may be a thrill. In regurgitation the left ventricle is enlarged as each beat has to pump the required cardiac output plus that which regurgitates. In AS the left ventricle hypertrophies as more force is required to eject the blood past a tight valve. In laminar flow, the resistance to flow is proportional to the 4th power of the radius so that a small reduction in calibre has a marked effect on resistance.

- **AS murmur:**
  - A2 is soft in AS. In aortic sclerosis, A2 is normal or loud.
  - Both conditions are associated with an early, harsh systolic murmur that is transmitted to the carotids. A similar murmur may occur without stenosis if turbulence is due to aortic aneurysm causing dilation of the proximal aorta. Such pathology may cause leakage from the valve too and with it an early diastolic murmur.
  - The typical murmur of AS is a crescendo-decrescendo systolic ejection murmur shortly after the first heart sound that ends just before the second heart sound. It is a rough, low-pitched sound that is loudest at the base of the heart and most commonly heard in the second right intercostal space.
  - If congestive heart failure leads to a fall in cardiac output, the murmur will be quieter.
  - An ejection click may be present, especially with bicuspid valves. Ejection sounds are more obvious in children in whom the valves are more mobile than in older people.
  - A fourth heart sound indicates left ventricular hypertrophy (LVH) in severe AS. If the left ventricle dilates and fails, a third heart sound may be heard.

Differential diagnosis

Other causes of murmurs that are, or appear to be, from the aortic valve include:

- Aortic sclerosis.
- Aortic regurgitation.
- Subacute bacterial endocarditis.
- Dilatation of the root of the aorta (may also lead to a leaking valve).
- Flow murmurs (turbulence from high cardiac output in anaemia, thyrotoxicosis and marked aortic regurgitation).
- Murmurs originating from the pulmonary valve with disease of that valve or atrial septal defect (the pulmonary and aortic areas are very close).

NB: flow murmurs can be normal in children and in neonates. Interpretation of cardiac sounds in children can be very difficult. Aortic valve disease is often not detected until the child is about 2 years old.

Investigations

- ECG may show evidence of LVH or left ventricular strain.
- CXR may show cardiac enlargement, calcification of the aortic ring or evidence of other disease. It is often normal except in advanced disease.
- Echocardiography:
  - Is the key diagnostic tool. It confirms the presence of AS, assesses the degree of valve calcification, left ventricular function and wall thickness, detects the presence of other associated valve disease or aortic pathology and provides prognostic information.
  - Trans-oesophageal echocardiography should be considered when transthoracic echocardiography is of insufficient quality.
  - Doppler echocardiography is the preferred technique for assessing AS severity.
- Exercise testing:
  - Is contra-indicated in symptomatic patients with AS but it is recommended in physically active patients for unmasking symptoms and in the risk stratification of asymptomatic patients with severe AS.
  - Exercise testing is safe in asymptomatic patients, provided it is performed under the supervision of an experienced physician while monitoring for the presence of symptoms, changes in blood pressure, and/or ECG changes.
- Multi-slice computerised tomography (MSCT) and cardiac magnetic resonance:
  - Provide additional information on the assessment of the ascending aorta when it is enlarged.
  - MSCT may be useful in quantifying the valve area and coronary calcification, which aids in assessing prognosis.
  - MSCT has become an important diagnostic tool for evaluation of the aortic root, the distribution of calcium, the number of leaflets, the ascending aorta, and peripheral artery pathology and dimensions before undertaking transcatheter aortic valve implantation (TAVI) - see ‘Management’, below.
- Natriuretic peptides have been shown to predict symptom-free survival and outcome in normal and low-flow severe AS and may be useful in asymptomatic patients.
- It may still be necessary to undertake cardiac catheterisation to measure pressures across the valve to assess the severity of disease and the need for intervention.
- Coronary angiography may be indicated as part of the assessment of coronary artery disease.
The risk factors for aortic sclerosis are similar to the risk factors for coronary heart disease and so similar investigations should be undertaken. [2]

Management[6]

See also separate Prevention of Infective Endocarditis article.

Patients with AS should avoid heavy exertion. Symptomatic patients require early surgical intervention because no medical therapy for AS is able to improve outcome. Treatment of high surgical risk patients has been modified with the introduction of TAVI.

Medical therapy

- The progression of degenerative AS is an active process, in many ways similar to atherosclerosis.
- Statin therapy should not be used in AS patients if the only purpose is to slow progression, as this has not been shown to be of any benefit.
- However, modification of atherosclerotic risk factors is strongly recommended. Aortic sclerosis and stenosis in the older age group, should be seen as a strong risk for coronary heart disease and appropriate steps should be taken.
- Patients who are unsuitable candidates for surgery or TAVI, or who are currently awaiting a surgical or TAVI procedure, may be treated with digoxin, diuretics, angiotensin-converting enzyme (ACE) inhibitors (or angiotensin-II receptor antagonists) if they experience heart failure symptoms.
- Co-existing hypertension should be treated. However, treatment should be carefully titrated to avoid hypotension and patients should be re-evaluated frequently.
- Maintenance of sinus rhythm is important, using anti-arrhythmic drugs as indicated.

Monitoring

- Stress tests should determine the recommended level of physical activity.
- Follow-up visits should include echocardiography.
- Asymptomatic severe AS should be re-evaluated at least every six months for the occurrence of symptoms, change in exercise tolerance (ideally using exercise testing if symptoms are doubtful) and change in echocardiogram.
- Measurement of natriuretic peptides may be considered.
- In the presence of significant calcification, mild and moderate AS should be re-evaluated yearly. In younger patients with mild AS and no significant calcification, intervals may be extended to 2 to 3 years.
Aortic valve replacement (AVR)

- AVR is the definitive therapy for severe AS. Operative mortality of isolated AVR for AS is 1-3% in patients younger than 70 years and 4-8% in selected older adults.
- Early valve replacement is strongly recommended for all symptomatic patients with severe AS who are otherwise candidates for surgery.
- There is limited evidence supporting the efficacy of sutureless AVR for AS in the short term. The evidence on safety raises no major concerns in the short term apart from the risk of paravalvular leak. There is concern about the risks of paravalvular and central leaks in the longer term.
- Management of asymptomatic severe AS is controversial. The decision to operate on asymptomatic patients requires careful weighing of the benefits against the risks.
- Pulmonary autograft replacement of the aortic valve (the Ross procedure) is a popular choice for AVR in infants and children but its use in adults remains controversial.

Balloon valvuloplasty

- The National Institute for Health and Care Excellence (NICE) guidelines state that current evidence supports the safety and efficacy of balloon valvuloplasty for aortic valve stenosis in adults and children.
- However, re-stenosis and clinical deterioration occur within 6-12 months in most patients.
- In adults, the procedure should only be used to treat patients who are unsuitable for surgery, as the efficacy is usually short-lived.
- In infants and children, critical AS is very rare and balloon valvuloplasty is usually used palliatively until the child is old enough to have valve replacement.

Transcatheter aortic valve implantation (TAVI)

TAVI is a recent development and provides a method of AVR which does not carry the same risks as surgical AVR. At present it is only recommended for AS.

Procedure

- TAVI can be performed under a general anaesthetic or under local anaesthetic with sedation, making it a consideration in patients who are unsuitable for surgical AVR.
- Balloon valvuloplasty of the aortic valve is performed, followed up by insertion of the specialised valve device. The whole procedure occurs under fluoroscopy and echocardiography guidance.
- The procedure is made easier by concomitant rapid pacing of the heart to 200 beats per minute.

Efficacy of TAVI

- The procedure is of equal efficacy as surgical AVR in patients who are unsuitable for surgery.
- Patients considered not suitable for AVR after surgical consultation clearly benefit from TAVI, compared with conservative treatment including balloon valvuloplasty.
- Reported 30-day mortality rates range from 5-15%. Approximately 1-2% of TAVI patients require immediate cardiac surgery for life-threatening complications. Reported one-year survival for TAVI ranges from 60-80%. Most survivors experience significant improvement of health status and quality of life.

Complications of TAVI

- Major bleeding from entry sites.
- Stroke or transient ischaemic attacks - there is a higher rate of these compared with surgical AVR.
- Ventricular tachyarrhythmias.
- Myocardial infarction.
- Aortic dissection.
- Cardiac tamponade.
- Postoperative aortic regurgitation and paravalvular leak.
Complications

- Aortic valve disease will eventually lead to decompensation with raised end-diastolic pressure, increased pressure in the pulmonary system and congestive heart failure.
- Damaged valves are susceptible to infective endocarditis.
- Calcified AS may produce small systemic emboli. The effect will depend upon where they lodge.
- Sudden death occurs in less than 0.2% of patients per year.

Prognosis

- Calcific AS is a chronic, progressive disease. The duration of the asymptomatic phase varies widely between individuals.
- Sudden cardiac death is a frequent cause of death in symptomatic patients but appears to be rare in truly asymptomatic patients, even in very severe AS.
- In asymptomatic patients with severe AS, reported average event-free survival at two years ranges from 20% to more than 50%.
- Predictors of symptom development and adverse outcomes in asymptomatic patients are:
  - Clinical: older age, presence of atherosclerotic risk factors.
  - Echocardiography: valve calcification, peak aortic jet velocity, left ventricular ejection fraction (LVEF), rate of haemodynamic progression, increase in gradient with exercise, excessive LVH, and abnormal tissue Doppler parameters of systolic and diastolic LV function.
  - Exercise testing: discovery of symptoms during exercise testing in physically active patients, particularly those younger than 70 years, predicts a very high likelihood of symptom development within 12 months.
  - Elevated plasma levels of natriuretic peptides.

- As soon as symptoms occur, the prognosis of severe AS is very poor, with survival rates of only 15-50% at five years.
- After successful AVR, symptoms and quality of life are in general greatly improved.
- Long-term survival may be close to the age-matched general population in older patients.
- In younger patients, there is substantial improvement compared to conservative medical therapy but there is a lower survival compared to age-matched controls.
- Risk factors for late death include age, comorbidities, severe symptoms, LV dysfunction, ventricular arrhythmias and untreated co-existing coronary artery disease.

Further reading & references

- British Heart Foundation

6. Guidelines on the management of valvular heart disease; European Society of Cardiology (2012)

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