Left ventricular outflow obstruction

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Left ventricular outflow tract obstruction occurs in six per 10 000 live births, or 7% of patients with congenital heart disease. 1,2 Its importance is disproportionate to the incidence because obstruction is either critical, giving rise to circulatory failure in the neonatal period or progressive, necessitating surveillance throughout childhood and into adult life. Furthermore aortic stenosis may cause severe obstruction without the symptoms of diminished cardiac reserve frequently encountered in other congenital heart defects. Aortic valve stenosis accounts for 70% of patients, 14% have sub-aortic obstruction, 8% have supravalve stenosis, with a similar percentage having more than one level of obstruction. 2,3

The majority of patients present with a murmur detected on routine examination, which is usually first heard at about 2 years of age. 2 A small but important group with critical stenosis present with heart failure in the first few months of life. In a further group, the clinical presentation is determined by associated defects such as a ventricular septal defect or coarctation of the aorta.

Nearly all patients have a harsh ejection systolic murmur in the aortic area with a thrill palpable over the right carotid artery. 4 However, the murmur of subaortic stenosis may be more obvious at the lower left sternal edge and may be difficult to distinguish from the pansystolic murmur of a ventricular septal defect. 5 An ejection click is often heard with valve stenosis, and an early diastolic murmur due to aortic regurgitation is present in 50% of patients with subvalve stenosis. Although it is not always possible to determine the level of obstruction by auscultation, it can be confirmed by cross sectional and Doppler echocardiography. 6,7

The severity of aortic stenosis correlates reasonably well with the clinical features (table). 8 The more severe the stenosis, the later the accentuation of the systolic murmur. An anacrotic pulse may also be present in older patients with severe stenosis. However, no single clinical observation has been found to be diagnostic of severe or mild stenosis. 9 Syncope, angina, and the presence of left ventricular strain on the electrocardiogram indicate that aortic stenosis is severe, but the absence of these features does not preclude severe stenosis. 10 Doppler echocardiography provides the best guide to the severity of aortic stenosis (table). The peak systolic gradient across the left ventricular outflow tract measured at cardiac catheterisation, particularly under general anaesthesia, is often substantially less than Doppler measured gradients under more physiological conditions.

Critical aortic stenosis has been defined as stenosis of severe degree, resulting in symptoms and requiring intervention within the first few months of life. 11 These infants usually present with cardiac failure and signs of poor cardiac output with weak peripheral pulses, hypotension, and metabolic acidosis. The aortic ejection systolic murmur is often soft or even absent. Inotropic support may be required and, as infants presenting in the first few days of life may be dependent on the arterial duct to adequately supply the systemic circulation, a prostaglandin infusion is indicated. In almost all cases, the stenosis is valvar, as other levels of obstruction are rarely severe in the first few months of life. 12

Hazard analysis confirms that the age of the patient and severity of stenosis at presentation, together with the level of obstruction, have a significant effect on survival and the need for treatment. 2,13 The risk of premature death in a patient presenting with moderate valve stenosis is small, but increases markedly in a similar patient with subvalve, supravalve, and particularly multilevel obstruction. 2 Even when the stenosis is mild at presentation, a patient with subvalve or multilevel obstruction is more likely to undergo intervention or develop endocarditis than a similar patient with valve or supravalve stenosis. 2 Aortic regurgitation is more common at presentation in patients with subvalve stenosis, but is most likely to develop with time in those with valve stenosis. 2

Aortic valve stenosis

Aortic valve stenosis is the most common type of obstruction and has the best prognosis. 2 The majority of patients (78%) present with mild
steno-
sis, 13 and they have a significantly better prognosis than those who present with moder-
ate stenosis. A recent study showed that the 20 year survival for a patient who presents with mild stenosis was 94%, but only 46% for a similar patient presenting with moderate stenosis. 13 A patient with moderate stenosis at presentation was also more likely to experience an important clinical event (surgery, balloon dilatation, or endocarditis) than one who presented with mild stenosis. After 20 years of follow up, the chance of being free of an important clinical event was 69% for a patient presenting with mild stenosis and only 5% in a similar patient presenting with moderate stenosis. 13

Nevertheless mild aortic valve stenosis does progress, with less than 20% of patients still having mild stenosis 30 years after presenta-
tion, 14 emphasising the need for follow up into adult life. Hazard analysis did not identify any significant risk factors associated with progres-
sion, which appears to be entirely related to the duration of follow up. 14

Critical stenosis occurs in 8% of patients with aortic valve stenosis. 13 In most of these infants the aortic valve is unicuspid, and examination of the aortic aspect of the valve reveals a single asymmetrical commissure and two raphe where the other commissures have failed to develop. 15 The circumference of the valve and the ascending aorta may be markedly reduced. A spectrum of underdevelopment of the left ventricle is also encountered, from those where the chamber is a tiny vestige to those with a normal or enlarged left ventricle. More than 60% of patients will survive initial inter-
vention. 17 18 This is related to the condi-
tion of the infant at the time of presentation and intervention, the size of left ventricle, aortic and mitral valves, 19 20 and the presence of a duct dependent systemic circulation, acidoses, 21 and other cardiac lesions. The type of intervention (surgery or balloon dilatation) may also influence initial survival. 16 18 23

In infants with critical aortic stenosis who survive initial intervention, the subsequent 10 year survival is 70%, 11 24 A small aortic valve diameter at presentation (<6 mm) and residual stenosis after initial intervention are important determinants of late prognosis. 11

Non-stenotic bicuspid aortic valves occur in up to 1% of unselected necropsies, but the inci-
dence is lower in clinical studies. 13 25 This may be because no murmur was detected or, when present, was thought to be innocent. Cross sectional and Doppler echocardiography have made the diagnosis of a bicuspid aortic valve easier. 26 The natural history of the con-
genitally bicuspid aortic valve is unclear, but data are slowly accumulating which may permit a further insight into its long term prognosis. In some patients it is associated with significant stenosis in childhood, whereas in other cases, calcific aortic stenosis develops in adult life. 27 28 The rate of calcification is much greater in a bicusp than in a tricuspid aortic valve and occurs at an earlier age, 29 but in some patients a bicuspid aortic valve may still be functioning normally in the seventh and eighth decades of life. 30 31 Recent studies that utilised cross sectional echocardiography to identify patients with a bicuspid aortic valve and Doppler echocardiography to demonstrate progressive stenosis 32 33 indicated that a bicuspid valve without initial stenosis will often function normally for the first 50 years, but that progression will gradually occur, usually as a result of valve calcification.

Surgery for aortic valve stenosis is undertaken in 25% of patients during childhood and adolescence usually because the pressure gradient has exceeded 60 mm Hg, a level beyond which there is increasing risk of sudden death. 13 The median age at first operation is in the region of 8 years. The reoperation rate is 17%, and 70% of reoperations will result in aortic valve replacement. 13 34 The median duration from first operation to aortic valve replacement is 12 years. 2 The overall operative mortality is approximately 10%, but death is rare after simple valvotomy.

Transcatheter balloon valvuloplasty, which was introduced in 1984, provides an attractive alternative to open heart surgery. 35 Reduction of valve gradient is comparable but the incidence of valve regurgitation is higher than with operation. Many surgeons believe that the precise valvotomy achieved by open operation also reduces the risk of restenosis and inci-
dence of valve replacement. Only time will tell. Balloon valvuloplasty is often undertaken for aortic valve gradients of 45 to 60 mm Hg, 35 36 which is less than that usually accepted for surgical intervention and is an alternative to early valve replacement with restenosis after surgery.

Subvalve stenosis

Subaortic stenosis constitutes between 8 and 20% of left ventricular outflow obstruction, 37 39 and comprises a diverse group of conditions. More than 60% of patients have short segment (fibromuscular) obstruction just below the aortic valve. 40 Other types of subaortic stenosis include long segment muscular obstruction, atrioventricular excrescence and malalignment of structures in the left ventricular outflow tract in association with a ventricular septal defect. 40

Subaortic stenosis is rarely detected during the first year of life 2 but subsequent progression is often inexorable, so that 50% require surgery during childhood or adoles-
cence. 2 Complete relief of the obstruction may be difficult, particularly with long segment obstruction. The reoperation rate increases with the length of follow up and is in the region of 17% at 10 years. 2 Balloon dilata-
tion has a limited application, 42 43 and may be of most value in the treatment of recurrent stenosis.

Thirty per cent of patients with subaortic stenosis have a ventricular septal defect and half of these have malalignment of muscular structures into the left ventricular outflow trac. 44 These patients undergo surgery for subaortic stenosis at a younger age than those with short segment, fibromuscular
obstruction.\textsuperscript{44} Two types of malalignment occur. The type of malalignment determines the level of obstruction in relation to the ventricular septal defect. In patients with posterior deviation of structures into the left ventricular outflow tract, the obstruction occurs above the ventricular septal defect and in those with anterior deviation of structures into the right ventricular outflow tract, the obstruction occurs below the ventricular septal defect. All patients with a ventricular septal defect should have echocardiography to detect malalignment of septal structures which may subsequently cause subaortic stenosis particularly if there is aortic coarctation or interruption.

**Supravalve aortic stenosis**
Supravalve aortic stenosis is the least common type of left ventricular outflow obstruction. The majority of patients have Williams syndrome or a positive family history,\textsuperscript{45, 46} but approximately 25% of cases are sporadic.\textsuperscript{47} Diffuse supravalve stenosis occurs in 15 to 24% of patients,\textsuperscript{45, 48} 49 and these patients also have a worse prognosis than those with localised obstruction.\textsuperscript{45, 50, 51} Coronary artery abnormalities have been reported in 20–33% of patients\textsuperscript{49} and there is a positive correlation between the severity of supravalve aortic stenosis and coronary artery dilatation. The prognosis of supravalve aortic stenosis related to coronary artery abnormalities is difficult to define. Older patients with supravalve aortic stenosis had symptoms of angina which were disproportional to the degree of left ventricular outflow obstruction.\textsuperscript{52} Sudden death has also been described in association with mild stenosis in a patient with obstruction of the origin of the right coronary artery.\textsuperscript{52} The most important factors in relation to prognosis are the age of the patient and the severity of the stenosis at presentation (D Kitchener \textit{et al}, unpublished data).

**Multilevel obstruction**
Patients with more than one level of left ventricular outflow tract obstruction have more severe stenosis at presentation and a worse prognosis than those with a single level of stenosis.\textsuperscript{53} Multilevel obstruction has been reported in 20 to 45% of patients with supravalve aortic stenosis.\textsuperscript{47–49} This incidence was significantly higher than in patients with aortic valve stenosis, where less than 10% of patients had other levels of obstruction.\textsuperscript{13, 54} Patients with multilevel obstruction present with more severe stenosis, are more likely to undergo operation, and have a higher incidence of reoperation. The operative and late mortality is also higher, emphasising the difficult and expensive problems confronting the surgeons.

Inf ective endocarditis occurs in approximately 2% of patients with left ventricular outflow tract obstruction\textsuperscript{55} and the incidence is a little higher in patients with subvalve or multilevel obstruction.\textsuperscript{2}

**Sudden death in patients with aortic stenosis**
The risk of sudden death in patients with all levels of left ventricular outflow tract obstruction is less than previously reported.\textsuperscript{28, 56} This approach is related to careful follow up and intervention before the development of severe stenosis. The advent of cross sectional and Doppler echocardiography has facilitated the detection of significant stenosis and this ability, together with regular clinical and echocardiographic assessment, should ensure that the risk of sudden death today is very small. In the large study by Hossack \textit{et al}, the only two patients who died suddenly had been lost to follow up for many years.\textsuperscript{8} The North American multicentre natural history study reported by Keane \textit{et al} recorded an incidence of sudden death of 6%, but 26% of patients had not been seen for at least 10 years.\textsuperscript{55} The incidence of sudden death in the Liverpool study was only 0.3%,\textsuperscript{2} indicating that with better surveillance the risk of sudden death is very low.

As sudden death has been reported exclusively with severe aortic stenosis,\textsuperscript{57} it is unnecessary to restrict sporting activities in patients with mild or moderate aortic stenosis, conversely a stenosis of sufficient degree to merit restriction of activity would constitute a strong indication of urgent surgery or balloon dilatation. With few exceptions all patients unoperated or after surgery require follow up into adulthood.

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18 Balaji S, Keeton BR, Sutherland GR, Shore DF, Monro JL. Aortic valvotomy for critical aortic stenosis in neonates and infants aged less than one year. Br Heart J 1989; 61: 358-60.


