HEART

Editorial

Subaortic stenosis: still more questions than answers

One of the most striking features about subaortic stenosis is the diversity of lesions that it encompasses and the inconsistency of terms used in its classification. Authors use the same terminology to denote different conditions and a variety of names for identical lesions. Some terms are descriptive, others histological or anatomical. Choi and Sullivan[1] suggested a classification based on morphological features that can be determined on cross sectional echocardiography (table 1).

Short segment obstruction is defined as subaortic stenosis with a length of less than one third of the aortic valve diameter, and consists of types previously termed membranous, diaphragm, discrete, fixed, fibrous or fibromuscular. The advantage of the term "short segment" is that it avoids descriptions that are inaccurate, and does not imply a histological diagnosis for that seen on echocardiography. Long segment subaortic obstruction is defined as stenosis that has a length of more than one third of the aortic valve diameter. This is sometimes called "tunnel" obstruction, but this term is confusing as it is applied to a number of different conditions. Subaortic stenosis can also be caused by deviation or malalignment of structures in the left ventricular outflow tract in association with a ventricular septal defect[2] or atrioventricular valve tissue in the subaortic area.

Short segment obstruction is caused by a complex fibromuscular structure a short but variable distance below the aortic valve with extension onto the anterior leaflet of the mitral valve and occasionally to the right coronary cusp of the aortic valve. In addition to being the most common type of subaortic stenosis, controversy surrounds both its cause and management.

A genetic predisposition has been suggested as there are reports of a familial incidence and a similar condition in Newfoundland dogs. Coarctation of the aorta, bicuspid aortic valve, mitral valve abnormalities or ventricular septal defect occur in more than 50% of patients[1 3 4] indicating that a congenital factor is involved in its pathogenesis. It is rare, however, in infancy and the characteristic changes seen on echocardiography develop gradually.[1 5] This suggests an acquired lesion based on a congenital or genetic predisposition. Gewillig and colleagues[6] and others have suggested that abnormal flow patterns could result from a septal ridge, malalignment of the interventricular septum, a small or elongated outflow tract, or an apical muscle band. They postulated that turbulence could damage the endothelium, resulting in deposition of fibrin and the subsequent development of the typical fibromuscular obstruction.

Table 1 Classification of subaortic stenosis

Short segment subaortic obstruction (length < 1/3 of aortic valve diameter)
Long segment subaortic obstruction (length > 1/3 of aortic valve diameter)
Subaortic stenosis due to malalignment of septal structures in the presence of a VSD
posterior malalignment with obstruction above the VSD
anterior malalignment with obstruction below the VSD
Subaortic stenosis due to atrioventricular valve tissue in LVOT
accessory mitral valve tissue
anomalous attachment of mitral valve chordae
tricuspid valve tissue prolapsing through VSD
abnormal left atrioventricular valve in AVSD

AVSD, atrioventricular septal defect; LVOT, left ventricular outflow tract; VSD, ventricular septal defect.

Controversy regarding the timing of surgery for patients with short segment subaortic stenosis revolves around a number of important considerations. These relate to the rate of progression of subaortic stenosis, the development and progression of aortic regurgitation, the risk of endocarditis, and the incidence of recurrence following resection. Cross sectional echocardiography can detect minor changes[5] in the subaortic area of patients presenting with an asymptomatic murmur or another cardiac lesion. Early diagnosis, coupled with the low morbidity and mortality from surgery, has led to a number of authors[4 5 7] advocating early surgery to prevent increasing stenosis and possibly the development and progression of aortic regurgitation. It was also hoped that this would reduce the risk of endocarditis and the incidence of recurrence. Some authors have advised surgery at a systolic gradient of 25–30 mm Hg across the left ventricular outflow tract,[4 8] while other institutions developed a policy of operation at diagnosis regardless of the degree of obstruction or aortic valve involvement.[7 9] Results of this move to early surgery have only recently been evaluated. Two studies[8 9] specifically compared the outcome of patients who had early surgery with those who did not. These provided further information on the factors that need to be considered when deciding on the timing of surgery.

Progression of subaortic stenosis

Progression of subaortic stenosis occurs but the rate is variable[1 3 4] and the factors influencing it are unknown. Most patients have surgery in childhood but, in a small number of patients, low gradients may remain for many years.[5 10] The article by Rohlicek *et al* in this issue,[11] indicates that patients who present with mild stenosis are likely to progress less rapidly than those who initially have a higher gradient. Patients with an increasing gradient need early surgery,[10 12] but surgery in mild cases may be delayed if follow up is meticulous.

Development and progression of aortic regurgitation

Aortic regurgitation occurs in more than 50% of patients with short segment subaortic stenosis.[3 4 8] It appears to be due to damage to the valve by the systolic jet produced by the subaortic obstruction.[1 4] Extensions of fibrous tissue onto the valve may also result in thickening and distortion of the leaflets.[1] A bicuspid aortic valve may contribute to regurgitation in some patients.

A compelling reason for early operation could be to prevent the development or the progression of aortic regurgitation. This is seldom severe in childhood[4 10] but age at operation was found to be a risk factor in some studies[9 13]

with a higher incidence and more severe regurgitation found in older children and adults.[14] An independent but related risk factor is the severity of stenosis. Previous reports, where most patients had a gradient of > 50 mm Hg,[10 13] show a clear relation between the severity of stenosis and aortic regurgitation. This correlation has been confirmed for patients with lower gradients.[8 9] The incidence and severity of aortic regurgitation[8 9] does not decrease significantly following surgery. In most cases the degree of regurgitation was unchanged following surgery but may progress.[8 9 11] A significantly lower incidence of late progression of aortic regurgitation has also been found in patients with a lower gradient at surgery.[8 9] Coleman *et al* found that the incidence and severity of aortic regurgitation after operation related significantly to the duration of follow up.[8]

Risk of endocarditis

Endocarditis is now a rare complication of congenital heart disease due to the widespread use of antibiotics. It is therefore difficult to assess the impact of early surgery. Wright *et al* found a reduced incidence after surgery[4] but it can still occur, and antibiotic prophylaxis remains essential.

Recurrence of subaortic stenosis after resection

Subaortic stenosis can recur after adequate surgical resection[3 12] and early surgery does not entirely prevent it.[8] Reoperation rates vary between 4% and 35% and the incidence of reoperation increases progressively with duration of follow up.[8 9 12] Reoperation may be necessary for residual stenosis or regrowth of fibromuscular tissue. Lupinetti *et al* showed that resection of a muscle wedge in addition to excision of the fibrous tissue significantly lowered the incidence of recurrence requiring surgery.[15] However, this has not been confirmed in other studies.[8 9] Gewillig *et al* suggested that surgery must be aimed at the removal of structures causing turbulence.[6] Despite increasing sophistication of echocardiography, the cause of altered flow patterns is seldom clear or readily treatable. Thus, even if this theory is correct, early operation is unlikely to prevent recurrence.

The higher the preoperative gradient, the higher the recurrence rate.[9] Brauner *et al* also found a higher recurrence rate in younger patients.[9] This may relate to technical difficulties associated with adequate resection through a smaller aortic valve. Brauner *et al* suggest that early intervention decreases the incidence of recurrence, although these findings are not borne out by Coleman *et al*.[8]

Any recommendations for operation must be tempered with the recognition that surgery for subaortic stenosis carries a risk. Mortality is low but complications such as damage to the mitral or aortic valves do occur,[4 8 10] and there is an incidence of complete heart block and perforation of the interventricular septum.[8 11]

Despite all these studies, the indications for surgery for subaortic stenosis are still not entirely clear. Regular follow up of even mild cases is essential and progression of the stenosis is an indication for surgery. There is little doubt that patients with a subaortic gradient of 40 mm Hg should undergo surgery to reduce the risk of significant aortic regurgitation and possibly the incidence of recurrence. Some authorities advocate surgery at lower gradients. Coleman *et al* recommend surgery at a gradient of 25–30 mm Hg[8] and Brauner *et al* advise surgery at diagnosis irrespective of gradient to reduce the damage to the aortic valve.[9]

The conclusions of the studies advocating early surgery are based on relatively small numbers and short follow up. The benefits become less attractive if late recurrence and progression of aortic regurgitation are not reduced. With longer follow up from the advocates of early surgery, this will become clearer. Freedom[16] emphasises the complexity of this disorder and reminds us that an intrinsically abnormal left ventricular outflow tract provides its own ultimate prognosis.

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