Development of subaortic stenosis and regurgitation several years after successful operation on a common atrium

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SUMMARY A patient who had had successful operation for a common atrium subsequently developed subaortic stenosis and regurgitation several years later. The diagnosis of subaortic lesion was confirmed by echocardiography. This case shows that the left ventricular outflow tract gradient should be measured before operation for common atrium and that after operation such cases should be examined by cross sectional echocardiography at follow up visits.

An Indian boy aged 5 was operated upon for common atrium at the National Institutes of Health, Bethesda, Maryland in 1980. A cleft in the anterior mitral leaflet causing only mild mitral regurgitation was left untouched. Preoperative cardiac catheterisation did not show a left ventricular outflow tract gradient while the angiogram showed a typical goose-neck deformity of the outflow tract. Catheterisation performed a year after operation again showed no left ventricular outflow tract gradient and a gradient of only 12 mm Hg between the right ventricular body and outflow tract. A grade 2/6 pansystolic murmur of mitral regurgitation caused by the cleft in the anterior leaflet was audible at the apex.

About two years after operation (1982) a systolic thrill developed at the upper left sternal border together with a prominent left ventricular impulse. Auscultation revealed a grade 4/6 systolic ejection murmur not preceded by click. The murmur was heard best at the site of the thrill, with poor conduction to carotid arteries. After another two years of follow up (1984) an early diastolic blowing murmur occupying about half of diastole also appeared. The murmur of mitral regurgitation mentioned earlier remained unaltered. Except for the development of these two new murmurs the child's symptoms were unchanged and he remains in New York Heart Association functional class I with blood pressure of 110/70 mm Hg and normal mental and physical development.

The electrocardiogram currently shows nodal rhythm at a rate of 60 beats per minute and increased right and left ventricular forces in addition to left axis deviation with a counterclockwise depolarisation sequence and right bundle branch block pattern.

The chest x ray shows a mild increase in the cardiothoracic ratio compared with that seen in the film taken two years after operation. The present M mode echocardiographic findings show mild dilatation of the left ventricular cavity with thickening of the interventricular septum and left ventricular free wall, fine diastolic fluttering of the anterior mitral leaflet with thickening, and systolic fluttering of the anterior (right coronary) cusp of the aortic valve with partial premature closure. The posterior (non-coronary) cusp could not be satisfactorily imaged (fig 1a and b). The cross sectional echocardiogram in the parasternal long axis view showed dense echoes in the left ventricular outflow tract just below the aortic valve. There was very little variation in these echoes with the phases of cardiac cycle (fig 2).

The above findings confirm the presence of subaortic stenosis and regurgitation. These were
not present before operation but have developed insidiously since.

Discussion

There are rare reports of an association between subaortic stenosis and uncorrected partial and complete forms of atrioventricular canal defects.1–3 The causes of left ventricular outflow tract obstruction are many and have been detailed by Ben-Shachar et al.4 The subaortic lesion is not usually symptomatic. In some instances the subaortic lesion has become apparent only after repair of the atrioventricular canal defects.4–7 How operation leads to the unmasking or development of a subaortic abnormality that was not present before intervention is a matter of conjecture. Several explanations have been suggested. In cases with interventricular connection, closure of the defect reduces the low impedance circuit where decompression may occur. After surgical closure all the left ventricular output goes into the aorta and this may unmask a latent gradient.4 Repair of a cleft mitral leaflet may also produce a gradient.6 The mechanisms operating include aboli-

Fig 1 M mode echocardiogram several years after operation for common atrium showing thickening and fine diastolic fluttering of the anterior mitral leaflet (a) and systolic fluttering and partial premature closure (arrows) of the anterior cusp of aortic valve (b).

tion of a low impedance circuit for left ventricular ejection, reduced mobility of the repaired cleft mitral leaflet that causes obstruction in systole, etc.6 Ebels and coworkers have shown by cross sectional echocardiography that malalignment of the aorta and interventricular septum and a narrow left ventricular outflow tract produced by a faulty contraction pattern are much more common than a fixed subaortic lesion, which is only occasionally encountered.7

Common atrium, which is a type of partial atrioventricular canal defect, however, has so far not been reported to be associated with subaortic lesions. The present report shows that this outcome may occur with this uncommon congenital malformation as well. We do not know why this child developed a subaortic lesion because none of the usual factors responsible for unmasking a latent gradient was present; there was no interventricular connection which was closed and the cleft of the mitral leaflet was not touched during operation. It seems most likely that narrowing of the left ventricular outflow tract with the development of a gradient and regurgitation occurred de novo after operation. The absence of left ventricular outflow gradient before operation and one year after operation was confirmed by cardiac catheterisation. Also, the clinical signs of stenosis and regurgitation developed two and four years respectively after operation. This thus suggests that the subaortic abnormality developed insidiously in the years after operation. Although we do not have preoperative echocardiograms for comparison, the present findings leave no doubt that an important subaortic lesion is now present. Cross sectional echocardiography is currently the non-invasive method of choice for investigating such lesions.7

Fig 2 Cross sectional echocardiogram several years after operation for common atrium in parasternal long axis view showing dense echoes below the aortic valve in the left ventricular outflow tract (arrows). LA, left atrium; LV, left ventricle; AO, aorta.
The observation is important and warrants an evaluation of the left ventricular outflow tract for obstruction in patients with common atrium in whom an operation is planned, so that not only the interatrial septum and the cleft mitral leaflet can be repaired but any abnormality of the left ventricular outflow tract can also be dealt with at the same time to avoid the possibility of a second operation. Because a subaortic lesion may develop years after successful operation for a common atrium, operated cases should be examined by echocardiography at follow up visits.

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References