Congenital discrete subvalvar aortic stenosis

Surgical experience and long-term follow-up in 20 paediatric patients

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Twenty patients aged from 4 to 14 years have been operated upon during the past 10 years at The Hospital for Sick Children, Toronto, for relief of congenital discrete subaortic stenosis.

The preoperative peak systolic gradients across the left ventricular outflow tract ranged between 42 and 156 mmHg, with an average of 84 mmHg. There was no operative death, but one late sudden death after an apparent good relief of the obstruction. Two children had to be operated upon again 4 and 6 years later for a recurrence of the lesion

It is suggested that the discrete diaphragm should be removed with a thin rim of muscle in the safe area.

Congenital discrete subvalvar aortic stenosis is one of the left ventricular outflow obstructions that can be successfully treated by operation with a very low risk. It remains a rare anomaly, accounting for 8 to 10 per cent of all cases of congenital aortic stenosis, which form 5.5 per cent of the various congenital cardiac defects (Keith, Rowe, and Vlad, 1967).

The purpose of this study is to review experience with the surgical treatment of this lesion at The Hospital for Sick Children in Toronto between 1960 and 1971 in 20 patients, and to present a long-term clinical follow-up in these children. This study is confined to discrete subvalvar aortic stenosis represented by a fibrous diaphragm seen and resected at the time of the operation. The other types of subvalvar stenosis, i.e. the muscular form and the so-called tunnel lesion, with fixed, fibromuscular obstruction of the left ventricular outflow tract, usually with involvement of the aortic leaflets (Reis *et al.*, 1971), are excluded.

Clinical material

Age and sex

The 20 patients ranged in age from 4 to 14 years with an average of 9 years. There were 14 male and 6 female patients, a sex ratio of 2.3:1, comparable to that noted

by others (Braunwald *et al.*, 1963). The age at which a murmur was first recorded was within the first month of life in 5 cases, within the first 6 months in 9 cases, and within the first year of life in 12 cases.

Associated malformation

Other malformations were noted in 3 patients; hydro-nephrosis in one, having necessitated a nephrectomy in early infancy, Turner's syndrome in one, and a rubella syndrome in one (with bilateral inguinal hernia and bilateral cataract). Associated cardiovascular malformations were noted in 5 other patients: persistent ductus arteriosus in 2, coarctation of the aorta in one, a small ventricular septal defect in one, and pulmonary stenosis in one.

Symptoms

Dyspnoea, fatigability, and chest pain on exercise occurred in 5 cases, and syncopal attacks in 2. Angina and exertional dyspnoea were both recorded in 5 patients. All had a peak systolic gradient of 70 mmHg or more across the left ventricular outflow tract and a peak systolic left ventricular pressure of 170 mmHg or more. One patient was in heart failure preoperatively, requiring digitalis.

Physical signs

On physical examination 5 patients were considered to have retarded growth. A praecordial systolic thrill was palpable in all patients but 2, and a systolic ejection murmur was recorded in each patient and was noted to be Grade 3 in 10 cases and Grade 4/6 in 9 cases. An early diastolic murmur was heard along the left sternal

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border in 7 cases. A pansystolic murmur best heard at the apex and thought to be related to mitral regurgitation was found in 4 cases and graded in each case 2/6. The pulse pressure ranged between 15 and 45, and averaged 30 mmHg.

Electrocardiography

The mean electrical axis of QRS ranged in the frontal plane between 30 and $+135^\circ$, with an average value of 57° . One patient had left axis deviation and 4 had right axis deviation. The sums $SV_1 + RV_6$ ranged between 24 and 62, with an average of 45, but could not be correlated with the severity of the stenosis. A strain pattern was found in 4 patients, all of them having a peak systolic gradient across the left ventricular outflow tract of 65 mmHg or more; left ventricular hypertrophy was present in 12 patients.

Radiology

On the plain chest x-ray the cardiothoracic ratio ranged between 40 and 63 per cent with an average of 49.7 per cent. The ascending aorta was dilated in 4 cases and left ventricular enlargement was reported in 6 other cases. No calcification was noted in the aortic area. Every patient had a left aortic arch.

Haemodynamics

Left heart catheterization data were available in each patient except for one who had only a right-sided angiogram for a technical reason. Retrograde catheterization was performed via the femoral artery in 6 and the brachial artery in 13 instances. The left ventricular cavity was entered in all cases but one, which necessitated transcutaneous left ventricular puncture. The mean pulmonary artery pressure ranged between 9 and 26 mmHg, with an average of 13.6. The left ventricular systolic pressure ranged between 150 and 310 mmHg, with an average of 206 mmHg. The peak systolic pressure gradient between the left ventricular cavity and the aorta ranged from 42 to 156 mmHg, with an average of 84 mmHg. Using our gradient classification (Mustard, Trusler, and Yao, 1963): *Grade 0*, < 30 mmHg; *Grade 1*, 30–49 mmHg; *Grade 2*, 50 to 100 mmHg; *Grade 3*, > 100 mmHg; no patient was *Grade 0*, 2 patients were *Grade 1*, 14 patients were *Grade 2*, and 3 patients were *Grade 3* (Fig.). The pull-back tracing revealed a typical subvalvar chamber in 11 instances. The left ventricular end-diastolic pressure ranged between 0 and 28 mmHg, being more than 12 mmHg in 6 cases. A contrast study was performed in the left ventricular cavity and the root of the aorta in 11 cases, and in the right side of the heart in 3 others. The left-sided study revealed a subvalvar obstruction in all cases, but also mild aortic regurgitation in 4 patients and an additional obstruction at the valve level verified at the operation in 2 other patients.

Operation was carried out under cardiopulmonary bypass using mainly a disposable bubble oxygenator. After a midline sternotomy and the usual caval and femoral artery cannulations, the lesion was visualized through a transverse incision in the cross-clamped aorta.

FIG. Peak systolic left ventricular-aortic gradient before operation in 19 patients.

Operative findings

At operation the valve appeared normal in 16 patients. In the remaining 4, all having aortic regurgitation, the leaflets were thickened with some degree of commissural fusion. In 2, the lesions were obvious enough to justify a valvotomy as well at the time of operation. There was one instance of bicuspid valve.

In all of the 20 cases the subvalvar stenosis appeared as a crescent-shaped fibrous diaphragm, 0.5 to 1 cm below the level of the aortic valve and adherent to the ventricular aspect of the anterior mitral leaflet. In 2 of the patients, however, a muscular bulging of the septum could be found as well as a fibrous diaphragm. The diameter of the subvalvar area, which ranged between 4 and 14 mm, with an average of 7 mm before the procedure, ranged between 10 and 19 mm postoperatively, with an average of 13 mm, as assessed by the size of the sound that could be put through the left ventricular outflow tract. At the same time a persistent ductus arteriosus was ligated once and a pulmonary valvotomy was performed in one case. The postoperative gradient recorded on the table was measured by simultaneous recording of the left ventricular and aortic or the brachial artery pressures. The gradient was between 0 and 35 mmHg, with an average of 20 mmHg.

Results

There was no operative death. Two patients had to be taken back to the operating room on the evening of the operation because of bleeding from the aortotomy site. A postpericardiotomy syndrome with fever and articular pain occurred in 3 patients, one of them having had a reoperation for bleeding.

Arrhythmias occurred in 3 patients postoperatively. One patient developed a complete heart block on the table when the aortic clamp was released. After a temporary epicardial pacing, a transvenous pacemaker was implanted. The second child developed

a first-degree heart block ($PR = 0.24$), well tolerated so far. The last child has an incomplete left bundle-branch block.

Cardiac failure requiring digitalis was present in 2 patients, each of them having preoperative gradients higher than 85 mmHg across the left ventricular outflow tract.

Follow-up

One patient was lost to follow-up. The remaining 19 were followed up clinically at the outpatient department over a period ranging from 6 months to 9 years and averaging 3.2 years. One patient died suddenly with an epileptic-like attack one year after an uneventful postoperative course. He was subject to fainting before the operation and had a gradient of 67 mmHg across the left ventricular obstruction and a left ventricular end-diastolic pressure of 17 mmHg without any clinical failure.

Two patients had to be reoperated upon 4 and 6 years later. The first child was complaining of headaches and fainted once. She also had electrocardiographic changes with appearance of left ventricular hypertrophy and strain patterns which she did not have preoperatively, and she was found to have a gradient of 110 mmHg when recatheterized (70 mmHg and 25 mmHg, respectively, before and after the first operation). At the second operation the fibrous diaphragm looked incompletely resected. The left ventricular outflow tract also appeared diffusely narrowed, requiring a vertical myotomy as well as a resection of the diaphragm. The postoperative gradient recorded on the operative table was 35 mmHg. The patient is now doing well 18 months later without any medication. The second child had been asymptomatic since the first operation (gradient of 10 mmHg measured on the operating table). He had a grade 2 ejection systolic murmur over the right second interspace, without aortic regurgitation. He was recently restudied because of inverted T waves on a routine electrocardiogram, and found to have a gradient of 85 mmHg across the left ventricular outflow tract. At operation he presented with the same type of fibrous diaphragm, which was resected with a piece of septal muscle. The postoperative pressures have not been taken.

The remaining 17 patients have no symptoms, 6 of them practising various sports with an excellent exercise tolerance. On physical examination all of them have an ejection systolic murmur over the aortic area, rated grade 4 in 2 patients thought to have some restenosis but who were asymptomatic and without electrocardiographic abnormalities.

Three patients now have a blowing ejection systolic murmur best heard at the apex and radiating to the axilla thought to be due to some mitral regurgitation. One of these patients had this murmur preoperatively but it appeared at the follow-up study in the 2 others. On the other hand, the murmur disappeared after the operation in 3 out of the 4 patients who had it preoperatively. A basal diastolic murmur could be heard in 11 cases. It was graded 2/6 in 7 cases, but in only 3 patients was the aortic pulse pressure greater than 60 mmHg, the usual clinical criterion for significant aortic regurgitation. The heart size was almost unchanged, the cardiothoracic ratio averaging 49.1 against 49.7 per cent, preoperatively. The postoperative electrocardiographic findings were not very different from the preoperative ones. The left ventricular hypertrophy was still present at the time of the follow-up in 7 patients. At the same time this pattern had disappeared in 5 other patients who had it preoperatively. There had been no instance of bacterial endocarditis.

Discussion

It is well known that the surgical relief of the pure membranous type of subvalvar aortic stenosis can be achieved with a very low risk and without operative mortality as in this series (Friedman and Pappelbaum, 1971; Shariatzadeh *et al.*, 1972).

The lesion can be associated with muscular bulging of the septum resembling idiopathic hypertrophic subaortic stenosis. This may be the result of the stenosis, but it is not correlated, at least in this study, with the degree of the stenosis or its duration. This overlapping of the two lesions has already been stressed by some authors (Roberts, 1971). The important technical point is that the concomitant muscular stenosis can be overlooked at the time of the operation if the subvalvar area is not carefully inspected.

The possibility of the recurrence of this lesion has to be raised. In two children with apparently good relief of the stenosis over a 4- and 6-year period, respectively, without any symptoms, a reoperation was indicated for fainting and electrocardiographic abnormalities. The usual appearance of the membranous diaphragm could be seen again. However, a fibromuscular type of stenosis in the subvalvar area, requiring myotomy, was present in 1 case. We now feel that the discrete diaphragm should be removed with a thin rim of muscle in the safe area.

The fate of the aortic and mitral valves is of importance because of the close relation to the subvalvar diaphragm. Thickening of the aortic leaflets possibly caused by the jet through the subvalvar ob-

struction may be responsible for the 7 preoperative aortic diastolic murmurs, as well as for the 11 postoperative ones. Only 2 patients had significant aortic regurgitation. Interference of the fibrous membrane with the motion of the mitral valve is usually thought to explain the preoperative mitral regurgitation. This occurred in 4 cases. The murmur disappeared in 2 out of 3 patients after the operation. On the other hand, care should be taken at operation not to injure the anterior mitral leaflet which is best avoided by leaving a layer of fibrous tissue over it.

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