Management of Congenital Aortic Stenosis

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Reid, John M., Coleman, Eric N., and Stevenson, John, G. (1970). *Archives of Disease in Childhood*, 45, 201. **Management of congenital aortic stenosis.** 72 patients with congenital aortic stenosis whose ages ranged from 1 month to 15 years were under supervision for periods ranging from several months to more than 9 years. The obstruction was subvalvar in 3. Additional malformations were aortic regurgitation, persistent ductus arteriosus, coarctation of the aorta, ventricular septal defect, partial anomalous pulmonary venous drainage, and mitral stenosis. Bacterial endocarditis complicated 3. Of 5 deaths unrelated to operation, 3 were sudden. Surgical treatment was carried out in 12 patients (commissurotomy, valve replacement with or without previous commissurotomy, subvalvar resection) with 2 deaths. 52 patients remained symptom-free. Detailed investigation to assess the patient for surgical treatment is indicated if symptoms are present or electrocardiographic evidence of severe left ventricular hypertrophy appears. Commissurotomy is, where possible, deferred until an age at which valve replacement can be undertaken if the stenosis cannot be adequately relieved without causing substantial regurgitation.

The introduction of cardio-pulmonary bypass surgery made relief of congenital aortic stenosis under direct vision possible (Ellis and Kirklin, 1962; Edwards and Jones, 1962), but uncertainty has persisted as to the clinical identification of critical obstruction and as to the optimum time for commissurotomy (Calatayud *et al.*, 1964; Putnam *et al.*, 1964; Morgan *et al.*, 1965). Though adults are satisfactorily treated with the insertion of a Starr-Edwards prosthetic valve (Starr *et al.*, 1963) or a valve homograft (Barratt-Boyes, 1964), children present technical difficulties in this respect, and the life-long anticoagulant therapy demanded by the prosthesis is an undoubted disadvantage. For these reasons, commissurotomy has remained the preferred operation for children with severe aortic stenosis. This communication based on a review of 72 consecutive patients reporting at a paediatric cardiac centre is concerned with indications for further investigation and surgery.

Material

Seventy-two patients whose ages at the time of this review ranged from 1 month to 15 years presented

between 1959 and 1968. In all, the aortic stenosis was believed to be congenital because the murmur had been heard during the first four years of life and there was no history of acute rheumatism. There were 45 boys and 27 girls, and the period of follow-up extended from 1 to 9 years. 52 were undoubtedly symptom-free, while in 9 a history of tiredness related to exercise was difficult to evaluate; definite symptoms were exercise-provoked and consisted of dyspnoea with syncopal attacks (1), dyspnoea with praecordial tightness (2), and dyspnoea alone (8). All were subjected to clinical, radiographic, and electrocardiographic examination. The principal electrocardiographic sign taken as evidence of left ventricular hypertrophy was a sum of the RV6 and SV1 deflections greater than 30 mm. in the first year of life, greater than 40 mm. in the next 9 years, and greater than 45 mm. thereafter. Depressions of the ST segment in the left chest leads, especially when accompanied by T wave inversion, were considered to be of particularly serious significance in this respect.

Cardiac catheterization was performed in 21 patients, left ventricular angiography in 5, and aortic root angiography in 8. These investigations were confined to patients with undue breathlessness on exercise or with symptoms suggesting myocardial or cerebral ischaemia, and to symptomless patients whose ECG revealed ST-T changes in the left chest leads, or steadily increasing left ventricular hypertrophy, or a substantial degree of left ventricular hypertrophy with neither ST-T changes nor steady deterioration.

Results

Clinical features. The clinical signs of aortic stenosis had initially drawn attention to the diagnosis but they were not found to supply any assistance with the decision to be made on further investigation and surgical treatment. Signs of aortic regurgitation were present in one. On radiographic examination prominence of the ascending aorta was often the only abnormality. Prominence of the left ventricular apex was common, but over-all cardiac enlargement was unusual except in 2 cases where aortic stenosis was accompanied by aortic regurgitation. The ECG was regarded as being within normal limits in 40, but in 32 there was evidence of left ventricular hypertrophy, with ST depression in V4-V6 in 5, and T wave inversion as an additional feature in a further 4, these characteristics being interpreted as reflecting severe outlet obstruction.

The peak systolic pressure gradient over the left ventricular outlet in the 21 patients who underwent cardiac catheterization is indicated in Table I. Table II relates this to the degree of left ventricular hypertrophy as reflected by the ECG. The correlation coefficient (r) between the sum of RV6 and SV1 and the systolic gradient over the left ventricular outlet was positive ($r = +0.7025$) and highly significant ($p > 0.001$).

TABLE I
Peak Systolic Gradient in 21 Patients

Systolic Gradient (mm.Hg)	10-29	30-49	50-69	70-89	90-109	110-129	>129
No. of patients	5	4	4	2	1	3	2

TABLE II
Systolic Pressure Gradient Related to Sum of RV6 and SV1 (21 Patients)

No. of Patients	RV6 + SV1 (mm.) Range	Systolic Pressure Gradient (mm.Hg)	
		Range	Mean Value
6	0-49	10-40	20
15	50-100	20-140	79

Cine-angiocardiology from the left ventricle showed the level of obstruction in the outflow tract, and in 3 it was subvalvar. Supra-avalvar stenosis was not encountered. Aortic root angiocardiology in 2 patients confirmed that stenosis was accompanied by regurgitation.

Complications and early death. 6 patients (8%) had an additional malformation. In 3 this was a persistent ductus arteriosus, ligated without incident. One had a ventricular septal defect. An infant who died in congestive cardiac failure at 1 month had preductal coarctation of the aorta as well as severe aortic stenosis. A boy of 11 years with subaortic stenosis, had mitral stenosis and partial anomalous pulmonary venous drainage in addition. He had latterly developed exertional dyspnoea, and his ECG showed the pattern of left ventricular hypertrophy with ST depression in the left chest leads. He died suddenly two weeks after catheterization while playing draughts. Bacterial endocarditis occurred in 3. 2 developed serious aortic regurgitation and 1 died. 2 deaths occurred in boys aged 8 and 15 years during moderately strenuous exercise, football and swimming, respectively. Neither had had premonitory symptoms. Both electrocardiograms indicated left ventricular hypertrophy but without ST depression in the left chest leads. One boy with mild aortic stenosis (peak systolic gradient 20 mm. Hg) developed mitral valve disease after acute rheumatism at the age of 9 years.

Surgical treatment. 12 patients had surgical treatment, the total number of operations being 14. 2 of 7 patients who underwent aortic commissurotomy subsequently required valve replacement because of a substantial degree of post-operative aortic regurgitation (Table III). One other aged 3 years died as a result of aortic regurgitation following commissurotomy on a severely stenosed bicuspid valve.

TABLE III
Surgical Procedures in 12 Patients

Procedure	No.	Deaths
Commissurotomy	7	1
Aortic valve replacement	6	1
Open excision of subvalvar obstruction	1	0

The ages of those who underwent commissurotomy ranged from 3 to 15 years, and the survivors have been under supervision for 1 to 7 years. Aortic valve replacement was confined to patients beyond the first decade of life, the youngest being 11 years; in 1 patient there was pre-existing regurgitation, in 2 the valve was bicuspid, and in the fourth, requiring valve replacement as the primary operation, the small valve ring made successful commissurotomy impossible.

Remaining patients. Of the survivors without operation, a further 2 patients who recently developed symptoms and increasing left ventricular hypertrophy were catheterized, and at the age of 9 and 12 years await aortic valve replacement. 52 patients remained well and symptom-free during the period of follow-up, and with no electrocardiographic sign of increasing left ventricular hypertrophy there has been no indication for further investigation or treatment. These patients appear to have relatively mild obstruction, but this might become more severe in the future were the valve to become calcified or distorted as a result of either infection or progressive sclerosis.

Discussion

This series showed the expected male preponderance (5 : 3) though not to the same extent as those of Nadas (1963) and of Putnam *et al.* (1964) in which males outnumbered females by 5 : 1 and 4 : 1, respectively.

It is essential first of all to identify patients with severe aortic stenosis because it is among these that sudden death is most likely and operative treatment must therefore be considered (Wood, 1960). Putnam *et al.* (1964) estimated that sudden death occurred in 5–10% of children with aortic stenosis and Campbell (1968a) that just over 25% of those alive at the age of 1 year die before the age of 20 years. During the period of this review 15 of our patients (21%) gave clear evidence of clinical deterioration. Of the 5 non-surgical deaths (7% of all patients) that of the 4-week-old infant who presented in congestive cardiac failure was attributable at least in part to preductal coarctation of the aorta. However, isolated aortic stenosis sufficiently severe to be complicated by congestive cardiac failure in infancy carries a notably bad prognosis (Nadas, 1963; Friedberg, 1966). In addition to one older child who died of bacterial endocarditis and one with a complicated malformation who died during sedentary activity, 2 died suddenly during moderate exertion. Neither of these had ST depression or T wave inversion in the left chest leads of the electrocardiogram; the child who died suddenly while sitting had ST depression without T wave inversion, and all 3 gave other electrocardiographic evidence of substantial left ventricular hypertrophy. Only one had experienced any symptoms. Campbell (1968b) found that 80% of deaths from aortic stenosis were sudden but that only 5% had a definite association with strenuous exercise. Even in the absence of symptoms, electrocardiographic evidence of left ventricular hypertrophy with any degree of ST-T depression

over the left praecordium is now taken by us to indicate severe outlet obstruction and the urgent need to measure the systolic gradient over the left ventricular outlet. In the absence of ST-T changes, electrocardiographic evidence of left ventricular hypertrophy of more than a mild degree justifies the same investigation but with a lesser degree of urgency.

A very significant positive correlation existed between the magnitude of the systolic gradient and the sum of RV6 and SV1, but accurate predictions from the ECG as to the extent of the gradient in individual cases could not be made because of the wide scatter of some observed values. In the age period of those catheterized (9–15 years), if the sum of RV6 and SV1 exceeded 50 mm. the systolic gradient was likely to exceed 50 mm. Hg. It is necessary, however, to record the disquieting observation by Nadas (1963) that severe aortic stenosis may be accompanied by a normal ECG. Hancock and Fleming (1960) in a study of 41 patients with aortic stenosis, but of whom only 6 were children, found a close correlation between the electrocardiographic evidence of left ventricular hypertrophy and the degree of obstruction. They found that T wave inversion occurred in all those with symptoms and that the gradient over the aortic valve was greater than 60 mm. Hg if the depth of the inverted T wave was more than 3 mm. Unfortunately, this useful correlation was not substantiated in the present series in which it was found that a gradient of this order might be accompanied by an upright T wave.

Other signs, such as the character of the arterial pulses or of the systolic murmur, or the radiographic appearances, were of little value in the assessment of the degree of obstruction. We cannot comment from our material on the positive correlation between delay in the maximum intensity of the murmur and the more severe grades of aortic stenosis, which has been pointed out by Oakley and Hallidie-Smith (1967).

Clinical evidence of aortic stenosis, productive of symptoms, or accompanied by electrocardiographic evidence of substantial left ventricular hypertrophy, requires that the site of the obstruction and the pressure gradient over the left ventricular outflow tract be determined. If severe valvar stenosis is found, the nature and the timing of surgical treatment must be considered. Cardiovascular symptoms which had constituted an indication for cardiac catheterization and angiography must also be an indication for surgical treatment, provided that the subsequent investigations have given reason for believing that the symptoms are a true reflection

of organic disease. Similarly ST-T depression in the left chest leads has been regarded as an indication for both investigation and operation. Even in the absence of these features a systolic pressure gradient across the left ventricular outlet of at least 50 mm. Hg, with evidence of substantial left ventricular hypertrophy has been taken as an indication for surgical treatment. The timing of the operation is dependent not only upon the clinical urgency but also upon the suitability of the patient for valve replacement if commissurotomy should produce aortic regurgitation. This complication is particularly likely if the valve is bicuspid, and those affected can be expected to derive little benefit from surgical treatment which has replaced one anomaly by another at least as serious (Morgan *et al.*, 1965). Apart from this immediate trouble commissurotomy may prove to be of no more than temporary benefit, and valve replacement may be required later because of progressive sclerosis, increasing regurgitation, or calcification of the aortic valve during adult life. Ellis and Kirklin (1962), reporting short-term results of commissurotomy on 47 patients (ages not given), did not detect significant aortic regurgitation in any of their 41 survivors, and this they attributed to meticulous conservation of cusps and commissures at operation. By contrast, 3 of the 6 survivors of our 7 patients who underwent an initial commissurotomy had a degree of post-operative regurgitation (significant in 2), attributable to the effect of operation and narrow valve ring with thickened distorted cusps. These 2 patients with significant post-operative regurgitation and 4 others (1 with pre-existing regurgitation and 3 with an irreparable valve) were subjected to aortic valve replacement with a Starr-Edwards prosthesis.

When deciding on the timing of operative treatment, it has to be borne in mind that clinical deterioration may occur with increasing age, and that during the period of delay and deterioration sudden death may occur related or unrelated to strenuous exercise. This event is practically always preceded by electrocardiographic evidence of left ventricular hypertrophy, but not necessarily by features implying imminent disaster. Having weighed the hazards of operation against those of refraining from operation, we have adopted the policy of proceeding to commissurotomy or to subvalvar resection where detailed investigations, performed in accordance with the indications already discussed, have confirmed the existence of a substantial degree of stenosis and localized its site. If the valve is shown to be bicuspid a distinctly limited double commissurotomy may have to be

accepted in order that severe post-operative regurgitation may be avoided. Whenever possible, we now make every effort to postpone operative intervention until the child is of an age (not less than 10 years) and of a size at which the valve ring is likely to accept a Starr-Edwards aortic prosthesis. In these circumstances, the surgical team can proceed to valve replacement if the commissurotomy leads either to inadequate relief of the obstruction or to an unacceptable degree of aortic regurgitation. Where detailed investigations reveal the coexistence of stenosis and regurgitation, the problem is regarded as one for medical management until an age is reached at which valve replacement becomes practicable. Patients accepted for operation and all whose preliminary investigations suggest the presence of more than mild aortic stenosis should be warned to avoid competitive sports and games. This seems the correct course to adopt, though we know that death may occur suddenly even when patients are at rest or engaged in quite gentle exercise. Not unexpectedly, however, the recommendation is not necessarily observed in practice; the 2 children who died during moderately strenuous exercise had been engaging in unsupervised activities. We are also particularly careful to underline the importance of penicillin cover for operations on the mouth, nose, or throat, including dental extraction, though such procedures seem seldom to be the causal event in cases of bacterial endocarditis during childhood.

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