Ventricular Septal Defect Repair in Childhood

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The criteria to be satisfied before the repair of a ventricular septal defect is advised in childhood cannot yet be stated with certainty. Even while knowledge of the natural history remains incomplete, however, little difficulty should be found in selecting for repair the large but uncomplicated defect, or in rejecting as not in need of repair the very small defect. There are more complex situations where the decision to advise operation remains largely dependent upon the personal experience of the risks and immediate benefit of operation possessed by the physician called upon to make that decision.

In describing this series of ventricular septal defect repairs, we are attempting an objective medical assessment of the results of operation. While we are unable to define precise criteria for operation, we feel it is possible and desirable to indicate those situations where the prospects are most promising or least promising.

SUBJECTS AND METHODS

Ninety-one children (44 male, 47 female) aged 12 months to 12 years were operated upon for the closure of ventricular septal defects from April 1960 until December 1966 in the Cardio-Thoracic Unit at Mearns Kirk Hospital. Supervision and investigation before and after operation took place at the Royal Hospital for Sick Children, Glasgow. In 41, the defect was accompanied by pulmonary hypertension (33–110 mm. Hg systolic) and in 17 it was complicated by pulmonary infundibular stenosis (30–112 mm. Hg systolic gradient); 5 patients had both infundibular stenosis and pulmonary hypertension. Twenty-nine patients had a systolic gradient of 10–28 mm. Hg across the right ventricular outflow tract, not caused by organic stenosis. Three had undergone previous operation: one for closure of a ductus arteriosus, the second for pulmonary artery banding,

both in the first year of life, because of congestive cardiac failure; the third had undergone pulmonary valvotomy 4 years before. Excluded from consideration were patients undergoing at the same operation, or to undergo later, treatment for any other cardiovascular malformation.

Repair was effected by direct suture in 74 patients and by the insertion of a patch in 17. Three patients had two defects in the ventricular septum. Until March 1963, a vertical ventriculotomy incision was used (43 patients), and thereafter a transverse incision (48 patients). Operations were performed with normothermic or hypothermic cardiopulmonary bypass. Resection of the right ventricular outflow obstruction was undertaken where appropriate. The main pulmonary artery was reconstructed once where there had been a previous banding operation.

Data other than mortality figures were derived from 78 patients (36 male, 42 female) operated upon before January 1966; the period of post-operative review was from 1 to 5½ years. Features investigated were symptoms (lack of energy, inactivity, tiredness, exertional dyspnoea), clinical phenomena (weight, height, blood pressure, heart size, heart sounds and murmurs), x-ray findings (cardiothoracic ratio, cardiac contour, pulmonary vascularity), and electrocardiographic evidence (chamber hypertrophy and conduction defects). Right ventricular and pulmonary artery pressures, the pulmonary:systemic blood flow ratio (Qp/Qs), and the pulmonary vascular resistance were derived from the pre-operative cardiac catheterization performed under minimal sedation. Forty-five patients were accepted as having a sufficient number of weight and height measurements before and after operation to permit an assessment of the effect of operation on growth; percentile ratings were used for this purpose. Post-operative assessment was aided in 10 patients by cardiac catheterization and in 6 by angiocardiography.

An over-all assessment of the result of treatment was made, and survivors were classified in 4 groups: (a) *cured* (defect repaired, infundibular stenosis relieved, innocent murmur frequent, minor deviation of cardiac contour or of electrocardiogram in a minority); (b) *improved* (usually substantial improvement in major features, i.e. heart

Received March 20, 1968.

size, contour, pulmonary vasculature, murmur, electrocardiogram); (c) *unchanged* (no substantial change in major features mentioned); and (d) *worse* (deterioration in a major feature or appearance of a complication, e.g. aortic regurgitation).

RESULTS

Deaths. There were 13 deaths in 91 patients (14.5%); all occurred within 8 days of operation (9 male, 4 female). Pulmonary hypertension had been present in 10, in 8 of whom the pulmonary artery pressure was between 70 and 110 mm. Hg systolic; in 3 of 6 where histology was available, proliferative medial changes of pulmonary hypertension were found in pulmonary arterioles. Two had infundibular stenosis (76, 80 mm. Hg systolic gradients); operation caused aortic incompetence in one and third-degree atrioventricular block in the other. There were no deaths in 1966 (13 patients).

Survivors. The over-all survival rate (1960–66) of 85.5 per cent represented 78 patients, 43 recorded as cured, and 30 provisionally as improved.

A final assessment of the period 1960–65 revealed complete recovery or improvement in 77 per cent, representing 34 patients *cured* (44%) and 26 *improved* (33%); one was *unchanged* (1.5%) and 4 *worse* (5%).

Two recorded as improved, whose septal defect had been accompanied by severe infundibular stenosis, still have substantial anomalies; in one a second operation is required to repair the residual defect and to relieve the stenosis, while in the other a second operation on the septal defect is under consideration, though relief of the stenosis has been complete. Eight others of the 26 had evidence of small and probably negligible residual defects.

Two of those recorded as worse had severe pulmonary hypertension, the post-operative right ventricular systolic pressure of one child rising to 150 mm. Hg. Infundibular stenosis had been present in the other 2 patients (30, 50 mm. Hg systolic gradient); aortic incompetence was caused at operation, persisting in one, but in the other relieved by partial resuture of the defect which, however, remained incompletely closed. The patient whose operation did not lead to any substantial change had a large incompletely repaired defect accompanied by severe pulmonary hypertension (70 mm. Hg systolic).

Symptomatology. Symptoms that suggested a possible cardiovascular origin were recorded in 9 patients (26%) whose operation brought about cure; they ceased in 4, diminished in 4, and continued unchanged in 1. Symptoms were present in 10 (38%) of those whose operation resulted in improve-

ment; they ceased in 4 and diminished in 6. By contrast, only 3 (23%) of the 13 patients who died were considered to have had any symptoms.

Weight and Height. Most patients were slender and of short stature, with both measurements before operation at the 10th percentile or lower. One-third of patients cured by operation and rather more of those who had undergone improvement showed a change in their percentile rating after operation, but the direction of the change was not uniform (Table I).

Pulmonary Hypertension and Vascular Resistance. The over-all incidence of pulmonary hypertension, including five instances where it accompanied pulmonary stenosis, was 45 per cent (41 patients), with a mortality rate of 24.5 per cent (10 patients). Its effect on prognosis was calculated from the 30 instances in 1960–1965 where it was unaccompanied by pulmonary stenosis. As a rule, the more severe the hypertension, the worse was the prognosis (Table II); the mortality rate where pressures were 70 mm. Hg systolic or above (54%) was three times greater than the rate where these were below 70 mm. Hg (18%).

When pulmonary vascular resistance (PVR units/m.² body surface area (BSA)) was related to the result of operation (Table III), it was shown that as pulmonary vascular resistance levels increased above 3 units, results of operation deteriorated. The highest value to be associated with post-operative cure was 10 units. Though post-operative improvement was once recorded where the pulmonary vascular resistance was 12 units, no patient with a higher pulmonary vascular resistance survived.

A raised pulmonary vascular resistance implied pulmonary hypertension, but the reverse relation was not constant. One patient aged 9 years with severe pulmonary hypertension (94 mm. Hg systolic; 63 mm. Hg mean) had a Qp/Qs ratio of 4:1 and a pulmonary vascular resistance of only 3 units per m.² body surface area. He successfully survived operation, whereas the only patients with comparable main pulmonary artery mean pressures (66, 68 mm. Hg) died; by contrast their pulmonary vascular resistance values were 16.5 units and 16 units, respectively, with correspondingly low Qp/Qs ratios of 1.35:1 and 1.4:1.

Defect and Shunt Size. The Qp/Qs ratio was used as a measure of shunt volume. There was nothing to suggest that the volume of the shunt might adversely affect the result of operation (Table IV). Nevertheless, those patients (17) with the

TABLE I

POST-OPERATIVE CHANGE IN WEIGHT AND HEIGHT PERCENTILE RATING RELATED TO RESULT OF OPERATION

Result of operation (No. of patients)		Percentile rating		
		Increased	Unchanged	Decreased
Cured (21)	Weight Height	5 4	14 14	2 3
Improved (24)	Weight Height	5 4	13 16	6 4

largest defects, which had been repaired by patching, had in comparison with other patients an unfavourable outlook. This was attributed to the effect of a raised pulmonary vascular resistance. Six survivors with pulmonary hypertension had pulmonary vascular resistance values ranging from 1.4 to 12 units/m.² body surface area (mean 4.3 units), whereas among five who died the range was 6 to 30 units (mean 16.2 units). Six with pulmonary stenosis had values between 0.8 to 3.5 units (mean 1.98 units).

In 10 out of 17 patients with a Qp/Qs ratio less than 1.5:1, this small flow, which had not been suspected from the accompanying signs, was attributable to right ventricular hypertension.

Pulmonary Stenosis. The over-all incidence of pulmonary infundibular stenosis was 18.5 per cent (17 cases), with a mortality of 11.5 per cent (2 cases). The outcome for the 13 patients in 1960-1965 showed no obvious connexion with the degree of right ventricular hypertension before operation.

The systolic pressure gradient of 10-28 mm. Hg over the right ventricular outlet in 29 patients was apparently not caused by organic obstruction; it was

TABLE II

PULMONARY ARTERIAL SYSTOLIC PRESSURE (MM. HG) BEFORE OPERATION RELATED TO OVER-ALL POST-OPERATIVE ASSESSMENT

Result of operation (No. of patients)	35-69	70-104	> 104
Cured or improved (17)	14	3	0
Unchanged or worse (3)	0	3	0
Dead (10)	3	5	2

TABLE III

PULMONARY VASCULAR RESISTANCE (PVR UNITS/M.² BSA) BEFORE OPERATION RELATED TO OVER-ALL POST-OPERATIVE ASSESSMENT

Result of operation (No. of patients)	0-1.5	1.6-3	3.1-6	6.1-12	> 12
Cured or improved (60)	15	28	15	2	0
Unchanged or worse (5)	0	2	2	1	0
Dead (13)	2	2	3	1	5

regarded as a "flow gradient" and did not influence the outcome of treatment.

Effect of Age on Prognosis. Repair was not attempted during the first year of life. Table V, which relates the result of treatment to the age at operation, shows that repair between the fifth and seventh birthdays was attended by the best results. Further analysis revealed that the best results for single years of age were obtained at 2 years (5 patients) and 5 years (9 patients); all patients were either cured (3 and 7 respectively) or improved (2 and 2 respectively); one aged 2 years and four aged 5 years had pulmonary hypertension. Worst results were encountered among those aged 1 year (4): three with pulmonary hypertension died, and the single survivor who had pulmonary stenosis was merely improved by operation.

TABLE IV

SHUNT VOLUME (Qp/Qs RATIO) RELATED TO RESULT OF OPERATION IN PATIENTS WITH A LOW PULMONARY VASCULAR RESISTANCE (PVR < 2.5 UNITS/M.² BSA)

Result of operation (No. of patients)	Qp/Qs ratio				
	1.0-1.4:1	1.5-1.9:1	2.0-2.4:1	2.5-2.9:1	> 2.9:1
Cured or improved (36)	10	18	3	3	2
Unchanged or worse (1)	0	0	0	1	0
Dead (3)	1	2	0	0	0

TABLE V

AGE AT OPERATION (YEARS) RELATED TO RESULT

Result of operation (No. of patients)	1-2	3-4	5-6	7-8	9-10	11-12
Cured or improved	6	21	16	11	4	4
Unchanged or worse	0	0	0	1	2	2
Dead	3	6	1	1	2	0

DISCUSSION

When patients were being selected for surgical treatment in 1960, there was scanty information on the natural history of ventricular septal defect, uncertainty about spontaneous closure, and ignorance of the effect of operation on pulmonary hypertension. The diagnosis was confirmed by cardiac catheterization, and from 1963 to 1966 in every case by selective angiocardiology in two planes. Repair was recommended if cardiomegaly was persistent, and if the blood oxygen saturation increased in the right heart by at least 10 per cent, or if there was substantial pulmonary hypertension without shunt reversal at rest, or moderate to severe pulmonary infundibular stenosis (gradient over 50 mm. Hg). The age at operation was determined by several factors; for psychological and social reasons, the age selected was 4 to 6 years. There were, however, some patients who did not come under observation until later, and others operated upon much earlier because of increasing cardiomegaly or pulmonary hypertension. Our criteria for operation modified by experience could now be formulated as persisting cardiomegaly with the pulmonary vascular resistance not exceeding 12 units/m.² We have become aware that successful operations are not necessarily precluded by pulmonary hypertension close to systemic arterial level, and that the large defect with a torrential pulmonary blood flow promises a good operative result. Only by relating pulmonary artery pressure to pulmonary blood flow, by the standard but arbitrary measurement of pulmonary vascular resistance, can the state of the pulmonary vasculature be assessed and the benign form of reversible pulmonary hypertension (hyperkinetic) associated with a copious pulmonary blood flow be recognized. In contradistinction to our practice with atrial septal defects, the repair of ventricular septal defects was often recommended and performed in the presence of a Qp/Qs ratio of less than 2:1. Often (i.e. 10 of 17 of those with a ratio below 1.5:1) these smaller shunt volumes were attributable to right ventricular hypertension associated with infundibular stenosis or a raised pulmonary vascular resistance rather than to the defect being of small area. It is arguable that some did not require repair, being likely to undergo spontaneous reduction in size (Arcilla *et al.*, 1963; Simmons, Moller, and Edwards, 1966). Nevertheless, a strict rule fulfilled before operation was that there should have been no substantial improvement in the abnormal cardiovascular features during the period of observation; the validity of this rule was demonstrated by the early and substantial post-operative improvement observed in most of these features. Perhaps

in retrospect where the Qp/Qs ratio was less than 1.5:1 and the right ventricle normotensive, a longer period of observation would have been justified, but experience of the more numerous defects accompanied by pulmonary:systemic ratios between 1.5:1 and 1.9:1 was that, though sometimes operation was postponed for as long as two or three years, the extent and persistence of the abnormal findings was such that further delay began to seem unreasonable.

In none of our patients was infundibular pulmonary stenosis the principal malformation, but in certain instances it added a major complication to the surgical procedure and influenced its outcome. Its effect on prognosis, however, was not quantitatively comparable in terms of the level of right ventricular hypertension with the more serious effect of a high pulmonary vascular resistance. The size of a defect had an unfavourable influence on the operative prognosis only in so far as large defects and high pulmonary vascular resistance were often found in association.

Symptoms were found to bear no constant relation to the haemodynamic situation, and their absence cannot, therefore, be used in assessing the need for, or the effect of, operation. The children whom we describe were with few exceptions small for their age, both in weight and height. Most of those for whom sufficient measurements were available showed no substantial improvement in this respect after operation, and these findings seemed independent of shunt volume and the presence of pulmonary hypertension. This experience appears to be at variance with that of Cartmill *et al.* (1966), and it is therefore possible that further observation over a longer post-operative period might yield different results. In the light of our present knowledge, however, a suggestion to parents that a poorly nourished child over the age of 1 year will be immediately benefited in this respect by operation could not be justified, nor are we able to adopt the suggestion that poor weight gain should be regarded as one of the indications for repair (*Brit. med. J.*, 1967).

Repair during the second year of life was hazardous and soon abandoned as unnecessary. Patients who had previously seemed to justify repair at this age because of large defects, pronounced cardiomegaly, and pulmonary hypertension were later managed more effectively by pulmonary artery banding in the first or second year, with repair to follow at leisure. Best results in relation both to mortality and to the extent of post-operative improvement followed repair at the age of 5 years. No advantage seemed to result from delaying repair

until later childhood. In order that it may not interfere with schooling, we now make arrangements for surgical treatment to take place during the few months that immediately precede school entry. Moreover, since in the opinion of Arcilla and his colleagues (1963) most ventricular septal defects which are going to close have done so by the age of 6 years, there should be little chance at the age of 5 years of recommending unnecessary surgical repairs.

The post-operative category described here as *cured* admits of no dubiety as regards long-term prognosis, but the category designated *improved* implies some prognostic uncertainty. Some patients so classified have signs of a residual small defect regarded as of little haemodynamic significance for the future. In 2 instances where the residual defect is substantial, a second operative approach has become unavoidable. We have, however, witnessed in connexion with ventricular septal defect, as after closure of an atrial septal defect or of a ductus arteriosus, a gradual regression of abnormal features, especially radiological and electrocardiographic, over a period of two or three post-operative years. One patient, five years after the technically successful repair of a very large defect, has the clinical features of a negligible residual defect, with a normal x-ray and electrocardiogram, though even 18 months after operation distinct cardiomegaly, pulmonary plethora, and evidence of left ventricular hypertrophy were manifest. Within the category of patients here described as *improved*, there are still several believed to be continuing to improve, and who as yet have not reached a stable post-operative haemodynamic state.

Surgical technique has improved during the review period. Nevertheless, the results quoted remain relevant, we believe, in relation to haemodynamic situations which are more favourable or less favourable to a satisfactory outcome of attempted surgical repair. Whether repair through a right atriotomy, now adopted by some surgeons in preference to the traditional ventriculotomy (Cartmill *et al.*, 1966), will justify its early promise where the pulmonary vascular resistance is high cannot yet be judged; this method has the advantage of preserving intact in such cases the right ventricular myocardium, though it must be admitted that it provides a less satisfactory exposure of the defects which have to be repaired.

SUMMARY

A survival rate of 85 per cent was experienced among 91 children from 1–12 years of age operated upon for the repair of ventricular septal defect from 1960–66 and followed for up to 5½ years. Pulmonary hypertension was a feature in 45 per cent, and infundibular stenosis in 18 per cent. Complete cure was achieved in approximately 45 per cent, a figure that may underestimate the final position. Deaths were attributable largely to a high pulmonary vascular resistance, less often to complications of the repair of infundibular stenosis. Large defects without pulmonary hypertension were repaired safely. It was concluded that a ventricular septal defect would merit repair when accompanied by persisting cardiomegaly, a resting pulmonary: systemic blood flow ratio of 1.5:1 or greater (or less if the right ventricle were hypertensive), and a pulmonary vascular resistance not exceeding 12 units per square metre body surface area, the repair being undertaken just before school entry (5 years). Repair in the second year of life undertaken because of pronounced cardiomegaly and pulmonary hypertension was abandoned in favour of pulmonary artery banding. Symptoms before operation were infrequent; when present they were not always abolished by operation. Successful operation was not always found to have a beneficial influence on the rate of growth.

We are particularly indebted to Mr R. S. Barclay, as head of the surgical team, and to Mr T. M. Welsh, Mr N. McSwan, and all other members of the Cardio-Thoracic Unit, Mearns Kirk Hospital, Glasgow, for the surgical treatment of these patients reviewed. We are grateful to the physicians of the Royal Hospital for Sick Children, and of other hospitals, who referred patients to us, and to the radiologists for their diagnostic assistance.

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