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Surgical treatment of congenital valvular aortic stenosis

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Chiariello, L., Vlad, P., and Subramanian, S. (1976). *Thorax*, **31**, 398–404. **Surgical treatment of congenital valvular aortic stenosis.** Twenty-two patients with congenital valvular aortic stenosis were surgically treated between 1967 and July 1975. Five (23%) were under 1 year of age (group I) and 17 (77%) were between 2 and 24 years (group II). All infants exhibited severe congestive heart failure and electrocardiographic (ECG) evidence of left ventricular hypertrophy (LVH) with strain pattern. In group II, angina was present in three cases, syncope and fatigue in two; the ECG indicated LVH in 10 cases (59%) with strain pattern in five (29%). A bicuspid aortic valve was present in 77% (17/22) of the cases; 32% had other cardiac anomalies. Aortic valvotomy was performed on cardiopulmonary bypass in 20 cases, and with deep hypothermia and circulatory arrest in two.

Three infants under 1 month of age with associated anomalies died (hospital mortality 14%). Intraoperative average peak left ventricular-aortic systolic pressure gradient decreased from 86 to 21 mmHg ($P<0.001$). Late clinical (in all cases) and haemodynamic (26%) follow-up showed severe restenosis in two patients of group II; one of them had a second operation, the other one died three and a half years postoperatively. Results assessed on the basis of symptoms, ECG changes, aortic valve function, and/or haemodynamic findings were fair in the two surviving infants. Results in group II were excellent in three, satisfactory in seven, fair in four, and poor in two cases. In infants, aortic valvotomy is a palliative procedure which carries a high risk. In the older age group, early and latc results are more gratifying.

Valvular aortic stenosis comprises approximately 50% of all forms of congenital left ventricular outflow tract obstruction reported in several large series (Morrow, Sharp, and Braunwald, 1958; Braunwald *et al.*, 1963; Keith, Rowe, and Vlad, 1967; Bertranou *et al.*, 1971; Lambert *et al.*, 1971). The malformation is usually discovered in the first five years of life. Auscultation discloses a systolic ejection murmur, commonly maximal at the second right interspace and frequently associated with an early systolic click. When indicated by deteriorating clinical and haemodynamic conditions, a commissurotomy is performed for relief of the obstruction. In the favourable cases of simple fusion of relatively normal cusps, surgical treatment would appear curative. However, in many instances, the valve apparatus is grossly abnormal because of rudimentary, thick, and deformed cusps. A commissurotomy in these cases would result in a palliative relief of obstruction, control of symptoms, and prevention of sudden death.

In infants, isolated valvular stenosis is rarely seen. Usually it is associated with other congenital cardiovascular defects. Symptomatic patients in this age group have an operative risk that is disturbingly high, although lower than non-operative treatment (Coran and Bernhard, 1969; Vlad *et al.*, 1971; Bernhard *et al.*, 1973).

These problems indicate that surgical treatment of valvular aortic stenosis still presents less than optimal results. In this report we present our series of patients, the concepts of management we have followed, and the results obtained in a seven-year period at the Buffalo Children's Hospital.

PATIENTS AND METHODS

Between September 1967 and July 1975, 22 patients underwent intracardiac repair of valvular aortic stenosis at the Buffalo Children's Hospital. At operation five infants (23%) ranged in age from 2 days to 6 months (group I) and 17 (77%) from 2 years to 24 years (group II). Age distribution is reported in Table I. In none of the cases was there a previous history of rheumatic heart disease. All but two patients underwent preoperative cardiac catheterization and selective angiocardiography. Indications for haemodynamic study were a history of heart failure, syncope or angina, and/or evidence of electrocardiographic (ECG) changes in patients with clinical findings consistent with aortic stenosis. Left ventricular hypertrophy on ECG was evaluated according to the method of Keith *et al.* (1967).

TABLE I

AGE DISTRIBUTION AND MORTALITY IN 22 PATIENTS WITH VALVULAR AORTIC STENOSIS

Age	No. of Patients	Percent	Hospital Deaths	Percent
< 3 mth	4	8	3	75
3 mth–6 mth	1	5	—	—
2 yr–5 yr	2	9	—	—
> 5 yr	15	68	—	—
Total	22	100	3	14

Indications for surgery consisted of any of the symptoms listed, even with normal electrocardiographic findings and irrespective of the catheterization data. Haemodynamic parameters only were used in the asymptomatic patients with normal electrocardiograms. A left ventricular-aortic peak systolic pressure gradient of 50 mmHg or more was considered to be an indication for surgery. We adopted the 50 mmHg pressure gradient as reported by Lambert *et al.* (1971) to separate mild from moderate aortic stenosis and 75 mmHg to separate moderate from severe stenosis. Aortic regurgitation was considered haemodynamically significant when the pulse pressure was over 60 mmHg (Lambert *et al.*, 1971). Follow-up data by clinical and/or haemodynamic assessment in our cardiology department have been obtained in all cases from two months to seven years after operation (mean follow-up time 35 months). Late results were arbitrarily graded on the basis of the following criteria:

Excellent result—patient asymptomatic, normal electrocardiogram, left ventricular-aortic peak systolic pressure gradient equal to or less than 25 mmHg, no aortic insufficiency

Satisfactory result—patient asymptomatic, stable electrocardiogram with no strain pattern, mild left ventricular-aortic pressure gradient (26–50 mm Hg), mild aortic insufficiency (pulse pressure less than 60 mmHg)

Fair result—patient slightly symptomatic (FC II, NYHA classification), electrocardiogram stable with or without strain pattern, moderate left ventricular-aortic pressure gradient (51–75 mmHg), significant aortic insufficiency (pulse pressure equal to or greater than 60 mmHg)

Poor result—patient symptomatic (FC III–IV, NYHA classification), further operation necessary, impaired electrocardiogram with appearance of a strain pattern, severe left ventricular-aortic pressure gradient (greater than 75 mmHg), significant aortic insufficiency (pulse pressure greater than 60 mmHg).

In evaluating the surgical results, each patient was assigned to the poorest category in which his clinical data could be located.

GROUP I Five infants were included in this group. Three were males and two females. They ranged in age from 2 days to 6 months, four being less than 3 months old (Table I). All the infants presented with congestive heart failure and one of them was cyanotic (Table II).

Electrocardiograms indicated left ventricular hypertrophy with a strain pattern in all instances (Table III). Preoperative cardiac catheterization

TABLE II

PREOPERATIVE SYMPTOMS IN 22 PATIENTS WITH VALVULAR AORTIC STENOSIS

Group	No. of Patients	Symptomatic		Symptom				
		No.	%	Angina	Syncope	Fatigue	Heart Failure	Cyanosis
I	5	5	100	—	—	—	5	1
II	17	7	41	3	2	2	—	—
Total	22	12	55	3	2	2	5	1

TABLE III

PREOPERATIVE AND POSTOPERATIVE ELECTROCARDIOGRAM IN 22 PATIENTS WITH VALVULAR AORTIC STENOSIS

Group	Preoperative ECG							Postoperative ECG							
	No. of Patients	Normal		LVH		LVH and Strain		No. of Patients	Normal		LVH		LVH and Strain		Strain	
		No.	%	No.	%	No.	%		No.	%	No.	%	No.	%	No.	%
I	5	—	—	—	—	5	100	2	—	—	1	50	1	50	—	—
II	17	7	41	5	29	5	29	16	8	50	3	19	3	19	2	12
Total	22	7	32	5	23	10	45	18	8	44	4	22	4	22	2	11

ECG = electrocardiogram; LVH = left ventricular hypertrophy.

and selective angiocardiography were carried out in three cases.

Two infants were operated upon as emergency operations after clinical assessment only. The left ventricular-aortic peak systolic pressure gradient was 70, 45, and 30 mmHg in the three infants studied.

Four patients had a bicuspid aortic valve and one a tricuspid valve. All bicuspid valves presented with rudimentary myxomatous cusps and an eccentric orifice. A coronary artery arose from behind each cusp. The raphe representing a vestigial commissure was located in two instances in the anterior cusp and in two in the posterior one (Fig. 1). In the only infants with a tricuspid valve the cusps appeared relatively normal with complete fusion of the right and non-coronary cusps and partial fusion of the other two commissures.

Valvular stenosis presented as an isolated cardiac anomaly in two cases; the remaining three infants had associated defects, that is, patent ductus arteriosus, endocardial fibroelastosis, and mitral insufficiency in two cases each (Table IV).

GROUP II Seventeen patients ranging in age from 2 to 24 years are included in this group. Two patients were below 5 years of age (Table I). Ten were males and seven females. One patient had had a previous operation eight years earlier. Seven patients were symptomatic. Angina was present in three cases and episodes of syncope and easy fatiguability in two cases each (Table II). Electrocardiograms showed a normal pattern in seven patients and left ventricular hypertrophy in 10, five of which were associated with a strain pattern (Table III). Haemodynamic studies were performed in all instances. The left ventricular-aortic peak systolic pressure gradient ranged from 50 to 145 mmHg with an average of 86 mmHg.

A bicuspid aortic valve was observed in 13 patients of this group. In nine, the two cusps were located anteriorly and posteriorly, one coronary artery arising from behind each cusp. The raphe in seven cases was observed in the anterior cusp and in one in the posterior. One patient presented two false commissures, one in the anterior and the other in the posterior cusp. In four cases, both

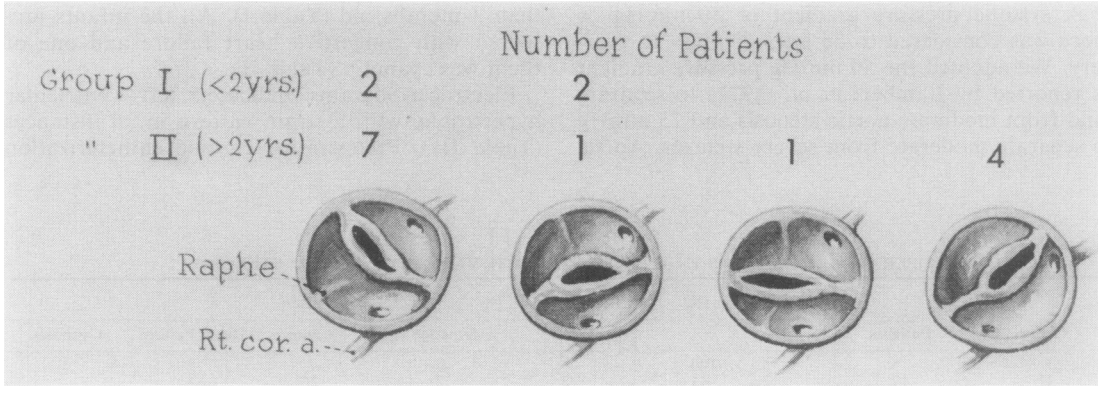

FIG. 1. *Anatomical variations observed in 17 patients with bicuspid aortic valve.*

TABLE IV

INCIDENCE OF BICUSPID AORTIC VALVE AND ASSOCIATED CARDIAC ANOMALIES IN 22 PATIENTS WITH VALVULAR AORTIC STENOSIS

Group	No. of Patients	Bicuspid Valve		PDA	EFE	AI	MI
		No.	%				
I	5	4	80	2	2	0	2
II	17	13	76	0	0	3	0
Total	22	17	77	2	2	3	2

AI = aortic insufficiency; EFE = endocardial fibroelastosis; MI = mitral insufficiency; PDA = patent ductus arteriosus.

coronary arteries arose from the left anterior cusp and the raphe was visible between the two orifices (Fig. 1). In four instances, the aortic valve was tricuspid with thick and nodular margins (Fig. 2). Moderate calcification was observed in two bicuspid and two tricuspid valves.

Three patients presented with mild aortic insufficiency (Table IV).

OPERATIVE TECHNIQUES

Twenty patients have been operated upon using temporary cardiopulmonary bypass under normothermia or moderate hypothermia at 30°C. Since 1969 two infants have been operated on using surface-induced deep hypothermia and total circulatory arrest.

A patent ductus arteriosus, when present, was ligated. An oblique incision was made anteriorly in the ascending aorta and extended into the noncoronary sinus of Valsalva. With a knife, one to three incisions were used to open the stenotic valvular orifice (Fig. 3). The extent and number of incisions were related to the anatomical findings. In those cases in which relatively normal, although fused, cusps could be identified, a precise division of the cusps was made to one millimetre from the aortic annulus. However, in four instances, marked deformity of the valve demanded a more cautious attitude, and a less extensive commissurotomy was performed. Thus a residual pressure gradient was preferred to significant valvular incompetence. In one of the cases with a tricuspid aortic valve an incision was performed in only one of the two fused commissures, creating a functional bicuspid valve. In one patient, residual aortic insufficiency was managed and a valvuloplasty performed.

RESULTS

GROUP I Three of the five infants with valvular aortic stenosis died in the postoperative period (Table V). These three were operated on when they had severe congestive heart failure and one was cyanotic. Two had been operated upon using deep hypothermia and circulatory arrest. In the third case, cardiopulmonary bypass had been used. They all had other associated cardiac anomalies, confirmed at necropsy, consisting of a ligated ductus arteriosus in two instances, endocardial fibroelastosis in two, and an insufficient mitral valve in two. The immediate cause of death was considered to be acute heart failure in all three patients.

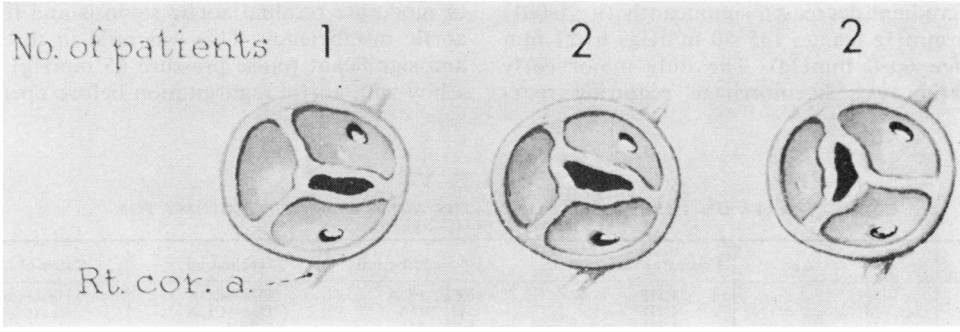

FIG. 2. *Anatomical variations observed in five patients with tricuspid aortic valve, one from group I and four from group II.*

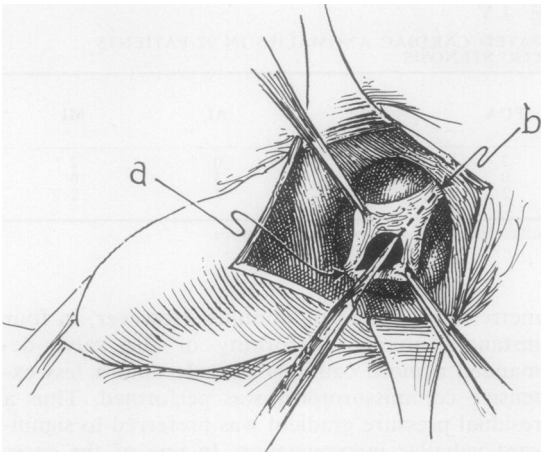

FIG. 3. *Aortic valvotomy performed with a knife, separating in the bicuspid valve only the larger unicusp from the other cusp. The incision is extended out to 1 mm (a and b) from the aortic annulus, thus accepting some degree of residual aortic stenosis.*

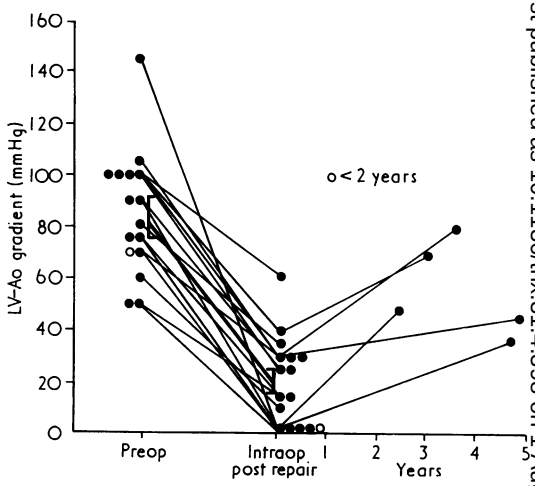

FIG. 4. *Peak systolic left ventricular-aortic pressure gradients obtained before and after repair in 15 patients with congenital valvular aortic stenosis.*

In only one of the two surviving infants are preoperative and intraoperative post-repair pressure measurements available. The peak systolic left ventricular-aortic pressure gradient dropped from 70 mmHg to 0 (Fig. 4). The two surviving patients are alive 30 months and two months after operation. The first has a fair result because mild symptoms and clinical evidence of aortic stenosis and insufficiency persist. The follow-up time is too short for evaluation in the second child.

GROUP II All 17 patients in this group survived operation and were discharged from hospital in good condition.

Pressure measurements obtained during the operation after repair showed relief of the obstruction (Fig. 4). The mean left ventricular peak systolic gradient decreased significantly ($P<0.001$) from 86 mmHg (range 145–50 mmHg) to 21 mm Hg (range 60–0 mmHg). The only major early complication was haemorrhage requiring reexploration in one case. One late death occurred suddenly in a 20-year-old man three years after a second operation for correction of residual valvular stenosis. The first operation had been performed elsewhere at the age of 9 years. The second operation, at the age of 17 years, was undertaken because of a left ventricular-aortic systolic pressure gradient of 100 mmHg and electrocardiographic evidence of left ventricular hypertrophy with a strain pattern. The gradient was reduced to 40 mmHg but at cardiac catheterization, performed two months before his death, it had increased to 70 mmHg. At the follow-up examination, 11 (69%) of the 16 survivors were asymptomatic and three (19%) presented with mild fatiguability. A basal ejection murmur was detected in all patients. Clinically, nine had mild to moderate residual aortic stenosis and four had aortic insufficiency. This was mild in three cases and significant (pulse pressure 65 mmHg) in one, a boy with aortic regurgitation before operation.

TABLE V

CLINICAL DATA ON THREE HOSPITAL DEATHS AFTER COMMISSUROTOMY FOR VALVULAR AORTIC STENOSIS

Patient	Sex	Age	Preoperative Symptom	Associated Lesion	Technique	Cause of Death
1	F	2 d	CHF	EFE, PDA	DH—CCA	Heart failure
2	M	2 d	CHF	MI, PDA	DH—CCA	Heart failure
3	F	1 mth	CHF/cyanosis	EFE, MI	CPBP	Heart failure

CHF=congestive heart failure; EFE=endocardial fibroelastosis; PDA=patent ductus arteriosus; DH—CCA=deep hypothermia, cardiac circulatory arrest; MI=mitral insufficiency; CPBP=cardiopulmonary bypass.

TABLE VI
POSTOPERATIVE RESULTS IN 22 PATIENTS WITH VALVULAR AORTIC STENOSIS

Group	No. of Patients	Deaths		Result							
				Excellent		Satisfactory		Fair		Poor	
		No.	%	No.	%	No.	%	No.	%	No.	%
I	5	3	60	—	—	—	—	2	40	—	—
II	17	1	6	3	18	7	41	4	23	2	12
Total	22	4	18	3	14	7	32	6	27	2	9

The postoperative electrocardiogram (Table III) was normal in eight patients, showed left ventricular hypertrophy in three, left ventricular hypertrophy with strain pattern in three, and an isolated strain pattern in two. Late haemodynamic studies were performed in five patients who had symptoms or persistent left ventricular strain in the ECG (Fig. 4). An increase of the left ventricular-aortic pressure gradient was evident in all of them and was severe in two. The death of one of these two has already been described. The other one had a second commissurotomy three and a half years after the first with early complete disappearance of the pressure gradient. Thirty months after the reoperation the patient was asymptomatic.

According to the criteria adopted, results were considered excellent in three cases, satisfactory in seven, fair in four, and poor in two (Table VI).

DISCUSSION

The results of the present series indicate that the surgical treatment of congenital valvular aortic stenosis has a different prognosis for infants than for older patients. In infants, even with the benefit of improved techniques allowing adequate exposure (that is, cardiopulmonary bypass, deep hypothermia with circulatory arrest), the surgical risk is high.

Three out of five of our infant patients died at operation. The two infants who survived had a successful early result, but the future for them is uncertain. Recently, Lakier *et al.* (1974) postulated that aortic stenosis in infants represents a different spectrum of disease as compared with that occurring in older children. They considered these patients as an intermediate group between infants with aortic atresia and hypoplastic heart syndrome on the one hand and aortic stenosis occurring later in life on the other. Our series confirms that these patients have anatomical and clinical features which mitigate against successful surgical treatment. Before operation all were in severe congestive heart failure with electrocardiographic evidence of left ventricular hypertrophy and strain. Operation was performed as an emergency procedure. In four instances (all the infants with a bicuspid aortic valve) the cusps of the valve were rudimentary, myxomatous, and rigid. Surgical repair, although providing a more adequate valvular orifice, cannot change the poor quality and mobility of the cusps. Associated cardiac anomalies are other factors which may affect the prognosis. The three infants who died all had additional defects, two of them endocardial fibroelastosis confirmed at necropsy.

Although the operative risk is high and the late results are poor, we feel that surgical palliation still has to be attempted in selected cases. Vlad *et al.* (1971) report that in isolated valvular stenosis, when surgery could be expected to help, only a minority of patients can survive through infancy when treated medically. With endocardial fibroelastosis, prolonged survival is exceptional (Vlad *et al.*, 1971). Since the presence of this anomaly is not predictable, all babies with aortic stenosis who fail to respond promptly to medical treatment should be considered for surgical treatment.

Results of surgery in the older age group are in sharp contrast with those in infants. Although less than curative, surgical treatment is a safe and effective procedure. In our series, no hospital deaths or major complications occurred except for one reexploration for bleeding. Approximately 80% of the patients of group II became asymptomatic or slightly symptomatic and were able to engage in normal activities. Postoperative electrocardiograms showed some early improvement in the asymptomatic patients and support the view of Vlad *et al.* (1971) that the ECG may return to normal or near normal over a period of years and may indicate a good result. Residual aortic insufficiency is a possible postoperative complication which may require a valve replacement later. Special care was taken to stop commissurotomy at least at one millimetre from the aortic annulus; an

incomplete relief of the obstruction was preferred to an incompetent valve. In our series only one boy with preoperative aortic insufficiency showed significant regurgitation after surgery.

Relief of the obstruction at the time of surgery is effective, as demonstrated by early post-repair pressure measurements. These measurements obtained under abnormal physiological conditions should be accepted with caution. However, the high significance ($P<0.001$) of the pressure gradient relief at operation justifies the intraoperative measurements at least as a practical guide. Indeed, intraoperative pressure measurements had demonstrated inadequate or incomplete relief of the obstruction in the two patients who presented important late complications (sudden death and severe aortic stenosis requiring reoperation).

In conclusion, in patients beyond infancy, aortic commissurotomy provides better results than in infants. Although surgery cannot be considered curative in all instances, approximately 60% of the cases demonstrated excellent or good results. In the patients with fair or poor results, reoperation may be anticipated to prevent further deterioration and sudden death. In these patients aortic valve replacements may become necessary.

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