Aortic valvotomy for critical aortic stenosis in neonates and infants aged less than one year

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SUMMARY Between April 1974 and December 1987, 20 infants (six under one month of age) (mean weight 4.9 kg) underwent surgical valvotomy for critical aortic stenosis. Three of the four patients treated before 1976 died. Since 1979, 16 infants (four neonates) have undergone valvotomy with no deaths. Mortality for the whole group was 15%; in the neonates it was 33%. There were no late deaths. The survivors were followed up for a mean of 3 years 9 months and two required re-operations.

The considerable improvement in surgical results in the past decade should be taken into account when newer techniques such as balloon dilatation are evaluated.

Critical aortic stenosis in infancy has been associated with high mortality and morbidity.¹⁻³ Medical treatment alone is unsuccessful. Surgical valvotomy is successful to varying degrees in relieving the stenosis but in infancy it is only palliative and all patients will later require replacement of the aortic valve or root. There has been recent interest in the use of balloon dilatation for this condition.⁴⁻⁶ This is why it is important to analyse the results of surgical valvotomy so that the risks to the patient of the various forms of treatment can be established.

This paper reviews our experience with surgical valvotomy in aortic stenosis.

Patients and methods

Between April 1974 and December 1987, 20 infants including six neonates underwent aortic valvotomy for critical or severe aortic stenosis at Southampton General Hospital. Age at surgery ranged from one day to 51 weeks (mean 14 weeks, median eight weeks). Weight at surgery ranged from 2.5 to 10.9 kg (mean 4.9 kg). Ten patients required emergency surgery and eight of them were moribund before operation with poor perfusion and acidosis and a mean pH of 7.25. Of the other 12, three were symptom free while all the rest had signs of heart

failure. Two patients with symptoms had already had an operation: one who had valvotomy at 11 months had had repair of coarctation at the age of two months and repair of ventricular septal defect at three months and the other had undergone repair of coarctation with ligation of the ductus.

The electrocardiogram showed left ventricular hypertrophy in 13 with strain pattern in three. Chest x ray showed a cardiothoracic ratio of 0.47-0.76 (mean 0.61). Before 1984 all patients underwent cardiac catheterisation, but since then, of ten patients treated, all but three had an operation on echocardiographic grounds alone. Retrospective analysis of the angiogram and/or the echocardiogram showed small left ventricles in three, dilated ventricles in six, and normal sized ventricles in the rest. The left ventricular ejection fraction measured in 15 patients (on the angiogram in five and the echocardiogram in 10) ranged from 10% to 80% (mean 45%). The pressure gradient across the aortic valve measured in 15 patients (by catheter in seven and Doppler in eight) ranged from 33 mm Hg to 120 mm Hg (mean 76 mm Hg).

All patients underwent aortic valvotomy under cardiopulmonary bypass and moderate hypothermia. Cardioplegia was used in all but the first four patients who were treated before 1979. At operation eight patients had tricuspid aortic valves and the rest were bicuspid. Associated lesions corrected at the same operation included ligation of the ductus arteriosus in three, closure of atrial septal defect in two, and repair of coarctation of aorta through a second (left

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Table Data on 20 infants who had valvotomy

Patient	Age	Associated lesion	Cardiac catheterisation before operation	AV gradient (mm Hg)		LVEF (%)		Pre-op state	Pre-op pH	Outcome	Re-op	AR	EFE
				Before	After	Before	After						
1	16 wk		Yes	37	—	—	—	M	7.38	Died		—	+
2	33 wk		Yes	—	43	66	70	CF	—	Alive		+	
3	3 days	ASD	Yes	56	—	—	—	M	7.01	Died		—	+
4	3 days	ASD	Yes	—	—	33	—	M	7.0	Died		—	+
5	19 wk		Yes	53	25	14	70	CF	7.41	Alive		++	
6	7 wk		Yes	60	40	—	89	M	7.26	Alive		++	
7	44 wk	COA, VSD	Yes	80	60	76	70	CF	—	Alive	Yes	+	
8	37 wk		Yes	—	35	36	50	CF	7.4	Alive		—	+
9	9 wk		Yes	33	40	23	82	M	7.2	Alive		++	
10	29 wk		Yes	80	40	71	54	SF	7.21	Alive		—	
11	12 wk		—	—	30	34	—	M	7.6	Alive		—	
12	5 days	DA	Yes	—	30	40	52	M	7.3	Alive		+	+
13	1 day	DA	—	100	50	80	73	M	7.23	Alive		—	
14	6 wk	COA	Yes	100	40	58	60	CF	7.43	Alive		—	
15	11 wk		—	40	60	—	50	SF	7.42	Alive		—	
16	5 wk	COA, DA	Yes	100	35	10	62	CF	7.55	Alive	Yes	+	+
17	5 wk		—	120	100*	45	89	CF	7.4	Alive		—	
18	51 wk		—	100	40	—	60	CF	7.52	Alive		+	
19	3 wk		—	36	35	53	97	CF	7.58	Alive		—	
20	5 days		—	40	25	36	67	SF	7.59	Alive		—	

*Dynamic left ventricular obstruction.

AR, aortic regurgitation; ASD, atrial septal defect; AV, aortic valve; CF, cardiac failure; COA, coarctation aorta; DA, ductus arteriosus; EFE, endocardial fibroelastosis; LVEF, left ventricular ejection fraction; M, moribund; SF, symptom free; VSD, ventricular septal defect; + = mild; ++ = moderate.

thoracotomy) incision in one patient (table). Two had previously undergone coarctation repair, one of whom also had ductus ligation. One of these had a ventricular septal defect closed at a further operation before aortic valvotomy.

Results

Three of the four patients treated before 1976 died and there have been no deaths since. All the infants who died did so within 24 hours of operation. At the time of operation, two of them (both three days old) were desperately ill; necropsy showed severe widespread endocardial fibroelastosis. The third, a girl aged four months, had had three cardiac arrests at cardiac catheterisation just before operation.

Mortality for the whole group was 15%, and for the neonates it was 33%. Since 1979, 16 infants including four neonates have been treated. None has died.

Patients spent 1–13 days (mean three) in intensive care after operation, and ten patients spent longer than 48 hours. Ten patients needed inotropic support for from one hour to six days, and five of them required inotropes for over 48 hours. Intubation and ventilation was continued for longer than 48 hours in five patients (maximum five days). Follow up of the 17 survivors ranged from two months to 12 years (mean of 3 years 8 months). Two have required re-operations—one for resection of fibrous subaortic stenosis and one aortic root replacement for severe residual aortic stenosis. Five children still have symptoms and effort intolerance and seven are on medication to counter heart failure. There were no late deaths.

The most recent electrocardiogram showed left ventricular hypertrophy in eight patients with strain pattern in one, and the chest x ray showed cardiothoracic ratios ranging from 0.47 to 0.73 (mean 0.57). Doppler estimation of residual aortic stenosis showed gradients ranging from 25 to 100 mm Hg (mean 43 mm Hg). The valvar gradient was less than 60 mm Hg in all but three: 60 mm Hg in two and another had a dynamic subvalve left ventricular outflow tract gradient of 100 mm Hg on Doppler which was not present at cardiac catheterisation under general anaesthesia. Eight had aortic regurgitation (mild in five and moderate in three). The left ventricular ejection fraction estimated by echocardiography in 16 survivors ranged from 50 to 97% (mean 68%).

None of the patients has had neurological complications caused by their primary cardiac problem or as a result of operation.

Discussion

Widely varying mortality ranging from 9% to 66% has been reported for aortic valvotomy under either conventional cardiopulmonary bypass or inflow occlusion.¹⁷⁸ The major determinants of mortality are the patient's preoperative clinical state, the size of the left ventricle and aortic annulus, the presence of associated anomalies and, in particular, the presence of mitral stenosis and endocardial fibroelastosis.⁸⁹ The lesion is a variant of the hypoplastic left heart syndrome and in borderline cases categorisation is subjective.

Alternatives to open valvotomy include closed transventricular valvotomy without bypass and

percutaneous balloon dilatation. Although there is less experience with transventricular valvotomy, the results are comparable to open valvotomy.⁹⁻¹¹ Balloon dilatation has produced good results in some patients,^{4,5} but has damaged the aortic valve and root in others.^{12,13} Wren *et al* reported a series of thirteen neonates and infants who underwent percutaneous balloon dilatation in which there were five early and two late deaths.⁶ Their children, however, are not strictly comparable with our series. They were smaller and more unwell, two had already undergone surgical valvotomy and two died at subsequent operation.

Clearly, operation is the only alternative when the aortic valve cannot be crossed at catheterisation. Whereas, in theory, open valvotomy might be expected to be more controlled and give better results, proponents of transventricular valvotomy argue that the gelatinous and dysplastic nature of the valve in infants with aortic stenosis makes it unlikely that direct visualisation of the valve would influence the results.⁹ We feel that open valvotomy allows the surgeon to perform a careful commissurotomy in all cases and that direct visualisation helps to decide where and how far the incision ought to run. We believe that our low rates of clinically significant restenosis and regurgitation show that a good balance can be achieved between too large or too small a commissurotomy.

In each series a small proportion of patients needs early re-operation for residual stenosis or regurgitation, and all patients are expected to need replacement of the aortic valve or root in due course. The main advantage of percutaneous balloon dilatation is expected to be the absence of adhesions in the thorax at definitive operation. Also, balloon dilatation can be repeated several times to achieve success. Repeated attempts risk damage to the aortic wall, valve or the femoral artery, however. Also, when balloon dilatation fails the condition of the infant might deteriorate during the procedure. On the other hand, if balloon dilatation is limited to children who are thought to be unsuitable or unfit for operation the risk associated with the procedure would increase, as shown by the results of Wren *et al*.⁶ Eight of our patients (of whom five survived) were moribund (mean pH 7.25) and might in some centres have been deemed unsuitable for operation. We showed that good results can be achieved even in these very sick infants.

The various methods available for the relief of infant aortic stenosis have their advantages and disadvantages, but which is the best method in children whose condition is stable and who have no adverse features? Newer techniques such as balloon dilatation will need further evaluation before the answer is known and such an assessment must take into account the improved results for valvotomy.

The results of valvotomy have improved significantly in the past decade as shown by our figures

and those of Messina *et al* who reported one operative and no late deaths in 11 neonates.⁷

Until we know more about the results of new techniques, valvotomy will remain the most suitable method for the relief of infant aortic stenosis. The fact that an infant is "too unwell" is no reason to attempt balloon dilatation rather than valvotomy.

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Addendum

We have operated on a further seven infants aged from five days to four months (mean 6 weeks) including three in the first week of life. One 36 week preterm infant aged six days, weighing 2.4 kg, who also had mitral stenosis and endocardial fibroelastosis died 56 days after the operation, having never left the intensive care unit. The rest survived and are well.