

Management of critical valvular outflow obstruction in neonates

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Severe pulmonary or aortic valve stenosis may be lethal to the neonate if untreated. Two patients suffering from pulmonary stenosis and four with aortic stenosis were relieved surgically with the use of full cardiopulmonary bypass. Both neonates with pulmonary stenosis did well, making them the youngest recorded survivors of such treatment. Two of the four patients with aortic stenosis have done well, the other two having severe endocardial fibro-elastosis. The indications and technique for cardiopulmonary bypass in this age group are briefly described.

The potentially lethal nature of severe pulmonary or aortic valve stenosis in infancy has often been stressed in the past—for example, by Mustard, Jain, and Trusler (1968) and Hastreiter, Oshima, Miller, Lev, and Paul (1963). Not so well agreed is the surgical management. Operation in older children is usually conducted on cardiopulmonary bypass, but, to quote one textbook, 'The chief disadvantage of this procedure is the necessity of submitting the patient to extra-corporeal circulation; but this is no longer considered a serious handicap, except, perhaps, in very young critically ill infants' (Gasul, Arcilla, and Lev, 1966). We have recently operated on six neonates with critical outflow obstruction, and we consider the advantages of cardiopulmonary bypass outweigh any theoretical disadvantages, even at this age.

MATERIAL AND METHODS

The clinical presentation and investigations are detailed in the Table. There were two neonates with severe pulmonary valve stenosis, right-to-left shunting across a patent foramen ovale, and marked arterial desaturation. The other four neonates were in congestive heart failure with severe aortic valve stenosis, one having an additional patent foramen ovale with left-to-right shunting. Standard cardiac catheterization procedures were performed before operation in all cases, supplemented by selective biplane angiography in the two cases of pulmonary stenosis.

Operations were performed within 24 hours of cardiac catheterization. In each case total body perfusion was accomplished through a vertical median sternotomy with insertion of a thin-walled metal cannula into the ascending aorta through a small stab

incision. Excellent venous drainage was obtained by a single catheter inserted through the right atrial appendage. (The alternative technique of individual caval cannulation is potentially dangerous in these small patients, since kinking and occlusion of the catheter readily occur, resulting in rapid and dangerous rises in venous pressure. In addition venous pressure monitoring is technically difficult.) Decompression of the left heart was also obtained by this technique in those cases with a patent foramen ovale; in the remaining cases a left ventricular vent was used. Elective fibrillation was used for the pulmonary valvotomies, which were performed through a transverse arteriotomy. During this procedure the temperature was gradually reduced to permit elective circulatory arrest for closure of the foramen ovale. The neonates were rewarmed by restarting the circulation and defibrillation was not necessary. Operative pressure recordings in both cases confirmed the catheter finding of right ventricular systolic pressures in excess of systemic pressure. After valvotomy the right ventricular systolic pressures were 26 mm. Hg and 47 mm. Hg respectively, compared with systemic pressures of 85–90 mm. Hg.

A similar procedure was followed for aortic valvotomy in case 3, who had in addition a patent foramen ovale with left-to-right shunt. Cases 4, 5, and 6 had only an open aortic valvotomy, and for this the body temperature was not reduced, since the brief period of coronary ischaemia is well tolerated unless there is associated fibro-elastosis.

RESULTS

Case 3 died 10 hours after operation. His course after admission was uniformly downhill, and post-operatively increasing doses of isoprenaline were needed to maintain circulation. Necropsy

TABLE
CLINICAL AND INVESTIGATIONAL DATA

Case	Age at Onset of Symptoms (Days)	Cyanosis	Tachypnoea	Hepatomegaly	Oedema	Cardiac Enlargement (Radiological)	E.C.G.	Oxygen Saturation (%)	RV Pressure (mm. Hg)	PA Pressure (mm. Hg)	LV Pressure (mm. Hg)	Systemic Pressure (mm. Hg)	Principal Diagnosis	Other	Age at Operation (Days)	Weight at Operation (kg.)
1	3	++	+	+	0	+	RVH	40	102/5	—	76/5	78/50	PS	PFO (R-L) shunt	5	3.2
2	1	+++	+	0	0	++	RVH	20	113/8	—	60/3	—	PS	PFO (R-L) shunt	4	3.0
3	2	0	++	+	0	++	LVH RVH	97	84/8	84/44	115/14	73/60	AS	PFO (R-L) shunt PFO (L-R) shunt	3	2.7
4	7	0	++	++	++	+++	LVH with 'strain'	96	55/8	48/36	150/20	55/44	AS	—	10	3.9
5	6	+	++	+	0	++	LVH with 'strain'	86	40/2	40/20	87/6	43/35	AS	—	12	3.7
6	14	0	++	0	0	++	LVH with 'strain'	95	38/2	14/7	130/10	52/40	AS	PS (mild)	22	3.4

Cyanosis, tachypnoea, hepatomegaly, oedema and cardiac enlargement all graded 0+++.

RVH=right ventricular hypertrophy; LVH=left ventricular hypertrophy; PS=valvular pulmonic stenosis; AS=valvular aortic stenosis; PFO=patent foramen ovale.

confirmed that an adequate aortic valvotomy and closure of the patent foramen ovale had been achieved, but severe endocardial fibro-elastosis was present in the left ventricle. Case 5 died abruptly within one hour following operation, having appeared to have done well initially. Necropsy confirmed an adequate aortic valvotomy, but marked endocardial fibro-elastosis was present in the left ventricle.

The remaining four patients are all alive and asymptomatic. Case 4 is now 8 months old and has developed normally. He has an aortic ejection systolic murmur but no evidence of aortic regurgitation. A chest radiograph shows a normal-sized heart, but the electrocardiogram shows persistence of left ventricular hypertrophy. Case 6 is now 7 months old. He is normally grown and asymptomatic. Like Case 4 his intellectual progress is normal. On auscultation there is an aortic ejection systolic murmur but no diastolic murmur. Chest radiographs show considerable diminution in cardiomegaly, but the electrocardiogram also shows persistence of left ventricular hypertrophy.

The two patients with pulmonary stenosis and shunt reversal across a patent foramen ovale have done well. At 3 months of age both were progressing normally without evidence of cyanosis or heart failure.

DISCUSSION

PULMONARY VALVE STENOSIS It is generally agreed that at any age pulmonary valvotomy is

best performed under direct vision, though some surgeons still except the critically ill neonate. In infancy the usual technique consists of inflow stasis by caval occlusion with or without hypothermia, and good results have been reported (Cooley and Hallman, 1966; Mustard *et al.*, 1968). We preferred to use cardiopulmonary bypass in infants for several reasons. First, it affords the surgeon more time adequately to open what is often a small dysplastic valve, with less danger of producing pulmonary regurgitation, as occurred in 11 of Mustard's 26 cases. Although pulmonary insufficiency is probably benign in most cases if the pulmonary artery pressure is not elevated (Burnell, Woodson, Lees, and Starr, 1969), it is best avoided. Secondly, extracorporeal circulation allows closure of the foramen ovale at the same operation, preventing right to left shunting, which often persists after adequate pulmonary valvotomy, due to non-compliance of the right ventricle, and possibly necessitating a second operation (Mustard *et al.*, 1968). If this shunting is not great, as more often occurs in older patients, then the benefit is probably marginal. However, when it is pronounced, as may occur in very young patients, systemic and myocardial oxygenation in the post-operative phase may be grossly deficient, particularly if pulmonary complications occur following surgery in the infant. It was felt that these reasons were equally applicable in neonates when case 1 was seen. A third advantage became obvious as soon as bypass was established. This infant was severely anoxic and acidotic and

her heart was dilated and tense. Within moments of improved myocardial oxygenation the heart diminished in size and became much less tense. Similar findings were noted in case 2. It is our belief that the complete relief of hypoxia and acidosis possible with this technique contributed to the uneventful post-operative course in both patients. This contrasts with inflow occlusion, which temporarily must increase hypoxia and acidosis in an already depressed myocardium. Gersony, Bernhard, Nadas, and Gross (1967) pursued similar reasoning in advocating hyperbaric surgery for these infants, but this technique is not as widely available as cardiopulmonary bypass.

These two babies appear to be the youngest reported patients with severe pulmonary stenosis successfully corrected under cardiopulmonary bypass.

AORTIC VALVE STENOSIS The infant with severe aortic valve stenosis has a grim prognosis and justifies early operation, despite the frequency of associated lesions, in particular endocardial fibro-elastosis. Hastreiter *et al.* (1963), in an excellent review, consider that many such cases are intermediate forms between the hypoplastic left heart syndrome and isolated aortic valvular stenosis of older children, and considerable detail is given in their article of the various associated anomalies. While we realize that some of these anomalies can be defined by selective angiography, we have deliberately avoided this procedure because of the risk inherent in any patient with severe left ventricular outflow obstruction (two deaths in 15 patients of Hastreiter *et al.* being attributed to left ventricular angiography). Instead, we prefer the surgeon to diagnose and deal with anomalies present, recognizing that this will inevitably lead to increased difficulty in some patients. To follow this procedure the surgeon will require more time than that afforded by inflow occlusion, as for example described by Cooley and Hallman (1966), and we therefore advocate the use of cardiopulmonary bypass even in the neonate. Correcting the valve anomaly alone may require more than the two or three minutes afforded by inflow occlusion with normothermia if lethal aortic regurgitation is to be avoided.

Various reports of surgery in congenital aortic stenosis (Lees, Hauck, Starkey, Nadas, and Gross, 1962; Ellis, Ongley, and Kirklin, 1962) do not contain any neonates. Cooley and Hallman (1966) do not consider the neonates separately in their series of 20 infants, but mention that 'cardiopulmonary bypass was utilised in all but a few

newborn infants in whom a modified form of caval occlusion was employed for open valvotomy'. Hastreiter *et al.* (1963) reported two neonates who survived aortic valvotomy on bypass.

Similarly, several large series of operations with extracorporeal circulation in infants have been reported (Sloan, Mackenzie, Morris, Stern, and Sigmann, 1962; Baffes, Riker, and DeBoer, 1964) but there do not appear to be more than one or two neonates with aortic valve stenosis who survived. Therefore our cases 4 and 6 join a select group. It is admitted that the encouraging results are partly due to favourable anatomy (*i.e.*, relatively well-formed aortic valve), in contrast to cases 3 and 5, in whom severe left ventricular endocardial fibro-elastosis was present and the aortic valves consisted of irregular masses of myxomatous material. Our point in reporting these patients is to emphasize that surgery with cardiopulmonary bypass is feasible, and, we believe, essential if a better salvage rate is to be achieved in this group of neonates.

PERFUSION TECHNIQUE

The perfusion system had a total priming volume of 700 ml. and included an infant-size rotating disc oxygenator. A mean perfusion pressure of 50–60 mm. Hg was maintained and this required a perfusion rate of about 200 ml./min./kg. body weight. If circulatory arrest was necessary the cooling and rewarming rates were such as not to exceed a gradient of 5° C. between the blood in the arterial limb of the oxygenator and the pharynx. The total circulatory arrest periods varied between 7½ minutes at 30° and 29 minutes at 12° C.

We think the following points are important for perfusions in neonates:

1. The circuit shall be primed with blood drawn into heparinized bags within three hours of operation.
2. Bubble oxygenator systems are not used because we believe that the presence of silicone and/or anti-foam substances introduces additional hazards.
3. Before starting the perfusion the blood warmed to 37° C. by recirculation, to prevent excessive heat loss to which neonates are particularly prone. During this preliminary period the Pco₂ is adjusted to between 35 and 40 mm. Hg.
4. This group of patients requires a much higher flow rate than expected to maintain an adequate mean perfusion pressure.

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