Aortic atresia

Diagnostic cardiac catheterization in first week of life

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Four infants are described in whom cardiac catheterization and angiocardiology performed in the first week of life established a diagnosis of aortic atresia, confirmed at subsequent necropsy. Contrary to previous reports of non-specific haemodynamic findings in this condition, all four infants had an identical and diagnostic haemodynamic disturbance. A striking finding was that a severe bradycardia occurred when the catheter crossed the ductus arteriosus or entered the hypoplastic ascending aorta. This bradycardia, presumably due to obstruction to coronary flow, was abolished when the catheter was withdrawn. Unless the possibility of this occurrence is known its diagnostic significance may be missed; failure to withdraw the catheter could lead to an immediately fatal outcome.

Cardiac catheterization and angiocardiology form an essential part of the investigation of neonates and infants in severe distress from congenital cardiac defects. In general it is only by these means that adequate precision of diagnosis can be obtained to permit surgical treatment in those, admittedly few, conditions for which palliative or curative surgical procedures exist. Among infants with congenital cardiac anomalies causing death in the first week of life those with aortic atresia form the largest single group. Thus any centre adopting a policy of investigating such acutely ill infants will encounter a significant number with aortic atresia. It is therefore surprising that there are few, if any, reports of the findings at catheterization in patients with aortic atresia studied in the first week of life, and Watson and Rowe (1962) state that cardiac catheterization is not very helpful. While admittedly aortography is probably the single most informative investigation, it is our experience that the haemodynamic findings at catheterization can be diagnostic in themselves. It is the purpose of this communication to report the findings at catheterization in four infants studied in the first week of life, all of whom were shown to have aortic atresia.

Cardiac catheterization

Between October 1969 and April 1970 four patients with aortic atresia were studied by

cardiac catheterization and angiocardiology in the first week of life; two were male and two were female (age range 2-5 days), weighing between 3.0 and 3.7 kg. All were critically ill at the time of study and one (Case 3) had had an episode of cardiac arrest requiring external cardiac massage. The approach used was via the axillary artery and vein in three patients while in one (Case 2) the saphenous vein was employed. In Cases 1, 2, and 3 angiocardiology was performed with injection of contrast medium¹ into the right ventricle or pulmonary artery. In Case 4 the haemodynamic data were considered diagnostic of aortic atresia, and in view of this and the patient's critical condition no angiocardiology was performed. The average duration of the study for these four patients was 47 minutes.

Haemodynamic findings (Table)

In all patients there was a large left-to-right shunt detected at right atrial level. In all the saturation of blood in the right ventricle, pulmonary artery, and systemic artery was identical or nearly so. In all the pressure in the right ventricle and pulmonary artery was significantly higher than in the descending aorta. (While descending aortic pressure was obtained by catheter passage through the

¹ Sodium, calcium, and magnesium metrizoate (Triosil '75'). Glaxo Laboratories Limited, Greenford, Middlesex.

TABLE *Haemodynamic findings*

Site	Case 1		Case 2		Case 3		Case 4	
	O ₂ * satn %	Pressure (mmHg)	O ₂ † satn %	Pressure (mmHg)	O ₂ ‡ satn %	Pressure (mmHg)	O ₂ § satn %	Pressure (mmHg)
Superior vena cava	59	—	51	—	49	—	34	—
Inferior vena cava	70	—	72	—	57	—	35	—
Mixed venous	67	—	65	—	54	—	35	—
Right atrium	98	a = 11 x = 10 v = 19 y = 6 m = 12	96	a = 11 x = 5 v = 10 y = 9 m = 7	73	a = 16 x = 7 v = 12 y = 7 m = 12	86	v = 14 y = 6 m = 10
Right ventricle	97	47/5-7	96	60/6-10	75	73/11-13	91	38/6-8
Pulmonary artery	98	44/23 m = 33	98	54/23 m = 35	75	70/40 m = 49	90	38/11 m = 20
Wedge	—	v = 15 y = 10 m = 13	—	—	—	—	—	—
Subclavian artery	98	38/27	—	—	68	42/33	91	28/20
Descending aorta	98	33/25	—	29/20	79	30/24	91	22/15

* General anaesthesia, intubated, intermittent positive pressure respiration. Breathing 50% O₂.

† Breathing augmented oxygen (Gairdner box).

‡ Breathing room air.

§ General anaesthesia, intubated, intermittent positive pressure respiration. Breathing 25% O₂.

ductus arteriosus and might thus have been reduced by partial occlusion of the ductus, the subclavian artery pressure was recorded in 3 of the 4 patients and was also significantly lower than the simultaneously recorded pulmonary artery pressure at a time when no catheter was traversing the ductus arteriosus.) The most striking event during cardiac catheterization in all four patients was an episode of severe bradycardia occurring when the catheter crossed the ductus arteriosus (in Cases 2, 3, and 4) or when the arterial catheter was advanced down the hypoplastic ascending aorta (in Cases 1, 3, and 4 – ascending aorta not catheterized in Case 2). In each case recovery occurred when the catheter was withdrawn to the pulmonary artery or subclavian artery.

Findings at necropsy

All four patients died within a few hours of cardiac catheterization. At necropsy aortic atresia with a very small, blindly ending ascending aorta was confirmed in all four. In Cases 1, 3, and 4 the mitral valve was atretic (Case 1) or severely hypoplastic. In Case 2 a normal mitral valve communicated with a small left ventricle and there were multiple ventricular septal defects. There was a large atrial septal defect in Cases 1 and 2; in the remaining two patients the interatrial communication was a patent foramen ovale.

Discussion

Congenital cardiac anomalies are a major cause of death in the newborn. Probably the most common cardiac anomaly causing death in the first week of life is the hypoplastic left

heart syndrome (Rowe and Cleary, 1960; Lambert, Canent, and Hohn, 1966). Of infants with this anomaly, Lambert *et al.* (1966) found that three-quarters died during the first week of life. The 'hypoplastic left heart syndrome' is a term embracing a number of anatomical defects of which isolated aortic atresia is the commonest (15 of 37 cases reported by Lambert *et al.*, 1966). When heart failure occurs in the newborn and clinical evidence suggests that structural heart disease is responsible, it is unlikely that medical treatment alone will produce permanent improvement, nor, in this age group, are physical signs likely to provide a complete anatomical diagnosis on which surgical treatment can be based (Lees, 1969). Thus, despite the small salvage rate and the high risk of catheterization in this group of seriously ill infants full diagnostic studies including cardiac catheterization and angiocardiology are mandatory. In any centre adopting this policy a significant proportion of infants undergoing catheterization in the first week of life will have aortic atresia. Probably the most useful investigation in such cases is aortography, which can be diagnostic (Elliott, Best, and Schiebler, 1961; Neufeld *et al.*, 1962; Watson and Rowe, 1962). While this investigation only might be performed if the diagnosis is suspected clinically, it is likely that in many instances investigation will include cardiac catheterization. It is surprising, therefore, that few reports exist of the haemodynamic findings in infants with aortic atresia. Those reports that are available (Neill and Tuerk, 1968; Watson and Rowe, 1962; Elliott *et al.*, 1961; Nadas and Mody, 1966) refer to

patients of over 1 week of age. Reported findings include a high right atrial pressure and the detection of a left-to-right shunt at right atrial level as reported here; we also confirm the observation of Keith, Rowe, and Vlad (1967) that arterial desaturation may be slight – particularly when the infant is breathing oxygen. However, findings in older infants differ from those reported here in that right ventricular and systemic pressures have been identical; this is also our experience in two cases of aortic atresia studied at 1 month of age. It has been suggested that the haemodynamic findings in cases of aortic atresia are not helpful and that similar haemodynamics are observed in cases of total anomalous pulmonary venous return and of mitral atresia with normal aortic valve (Keith *et al.*, 1967; Watson and Rowe, 1962; Gasul, Arcilla, and Lev, 1966). While the findings may not be diagnostic in older infants, neither mitral atresia with normal aortic valve nor total anomalous pulmonary venous return is likely to be associated with a higher pulmonary than (often very low) systemic pressure; in particular, the occurrence of bradycardia when the catheter crossed the ductus arteriosus or entered the ascending aorta was very striking and, in our experience, unique to this condition when studied at less than 1 week of age. Presumably this bradycardia was due to sudden reduction in coronary blood flow which is dependent on flow through the ductus and thence in retrograde fashion down the very hypoplastic ascending aorta. It is important to appreciate the cause of this bradycardia, which is abolished when the catheter is withdrawn. If the catheter had been left in place it is probable that cardiac arrest would have ensued. It is of interest that, just as pulmonary and systemic pressures seem to be equal in older infants with aortic atresia, we have not observed this bradycardia in infants with aortic atresia studied at 1 month of age. It is possible that longer survival depends on there being an adequate sized communication between the pulmonary artery and aorta. Such observations suggest the possibility of surgi-

cal palliation by the creation of an aortopulmonary communication at the level of the descending aorta together with banding of the distal pulmonary arteries (Keith *et al.*, 1967). Additionally, enlargement of the interatrial communication might be necessary (Neill and Tuerk, 1968): in two of the four cases reported here a foramen ovale provided the only communication between left and right atrium.

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