Anatomical variants in aortic atresia *Potential candidates for ventriculoaortic reconstitution*

Robert M. Freedom, W. G. Williams, M. R. Dische, and Richard D. Rowe

From the division of Cardiology, Departments of Paediatrics, Cardiovascular Surgery and Pathology, The Hospital For Sick Children and Faculty of Medicine, The University of Toronto, Canada

Aortic valve atresia is usually associated with severe hypoplasia of the left ventricle and mitral apparatus. Palliative procedures, including atrial septectomy, pulmonary artery—aorta anastomosis, and pulmonary artery banding, have been performed in some patients. However, because of the distinct underdevelopment of the left heart, the long-term prognosis must be guarded even in the survivors of these procedures.

Rarely, aortic valve atresia is found with adequate left heart chambers. Because very few of these patients are described, the clinical, angiocardigraphic, and pathological findings in two patients are presented, with comments on palliation in the neonatal period, and the potential for ventriculoaortic reconstitution.

Aortic valve atresia is usually associated with severe hypoplasia of the left ventricle and mitral apparatus (Lev, 1952). Survival for even a short time depends on patency of the ductus arteriosus and lack of significant restriction at the atrial septum (Noonan and Nadas, 1958; Saied and Folger, 1972). With medical therapy alone, the majority of patients with aortic atresia and hypoplasia of the left heart chambers are dead by 6 weeks of life (Noonan and Nadas, 1958; Watson and Rowe, 1962; Saied and Folger, 1972). Surgical palliation of less severe forms of the hypoplastic left heart syndrome has included atrial septectomy and construction of a pulmonary artery—aortic anastomosis, but only a few survivors of these procedures have been reported (Sinha *et al.*, 1968; Cayler, Smeloff, and Miller, 1970; Van Praagh *et al.*, 1971), and even in the survivors of these palliative procedures the long-term prognosis must be guarded. To our knowledge, no patient with aortic atresia has survived such palliative surgery, though this approach was suggested in 1967 (Keith, Rowe, and Vlad, 1967).

Rately, aortic valve atresia is found with adequate left heart chambers. However, very few of these patients are discussed (Rosenquist, Taylor, and Stark, 1974). In order to define this clinically important entity, the clinical and pathological findings

in two such patients will be presented, with comments on palliation in the neonatal period, and the potential for ventriculoaortic reconstitution (Bernhard, Poirier, and LaFarge, 1975).

Case reports

Case 1

This 6-day-old female infant presented with signs of congestive heart failure and cyanosis.

Physical examination showed a mildly distressed, tachypnoeic (80/min) infant, with mild generalized cyanosis. All pulses were easily palpable. Systolic blood pressure in the arms was 90 mmHg (12.0 kPa) and in the legs 100 mmHg (13.3 kPa).

Cardiac examination revealed a biventricular impulse, single second sound, constant ejection click along the left sternal border, and a short ejection murmur. A gallop was audible at the left lower sternal border. The liver was palpable 3.0 cm below the right costal margin.

Chest x-ray film showed laevocardia, modest cardiac enlargement, and pulmonary overcirculation. Pulmonary oedema was not present. There was visceral situs solitus (Fig. 1).

The electrocardiogram showed sinus tachycardia, right axis deviation and right ventricular hypertrophy (Fig. 2).

Received 12 January 1976.

FIG. 1 Case 1: *Laevocardia and visceral situs solitus. The heart is modestly enlarged, and pulmonary overcirculation is evident.*

Echocardiography suggested a normal to large right ventricle, large left atrium and left ventricle, and one large anterior arterial root. These findings were interpreted as consistent with truncus arteriosus. Both tricuspid and mitral valves appeared normal, and there was a ventricular septum.

Diagnostic cardiac catheterization and angiography were performed shortly after ad-

FIG. 2 Case 1: *The electrocardiogram shows right axis deviation, right atrial and ventricular hypertrophy. Septal Q waves are not seen.*

mission. The haemodynamic results are summarized in the Table. Left ventricular angiography showed a normal-sized morphological left ventricle, high, possibly supracristal ventricular septal defect, and opacification of an anteriorly placed pulmonary artery (Fig. 3a, b). The ascending aorta did not opacify from this injection. An angiogram in the main pulmonary artery showed dilated pulmonary arteries, right-to-left shunting of the ductus arteriosus, with opacification of the descending

FIG. 3a, b Case 1: *Frames from left ventricular cineangiogram in anteroposterior and lateral projection. The venous catheter has been advanced from the inferior vena cava through an atrial communication into the left ventricle (LV). A normal-sized left ventricle (LV) is opacified, with visualization of an anterior pulmonary artery (PA). A ventricular septal defect (arrow) is seen with opacifications of the anterior right ventricle. The aortic arch and branchiocephalic vessels are not clearly seen.*

FIG. 4 Case 1: Frame from an aortogram in the anteroposterior projection. A catheter has been advanced from the right axillary artery to the aorta. A diminutive ascending aorta (arrow) is visualized, with retrograde opacification of the coronary arteries.

thoracic aorta, and retrograde filling of a diminutive ascending aorta. The pulmonary artery laevophase visualized pulmonary venous return, a normal left atrium, and normal left ventricle. Finally, using a right axillary artery approach, an ascending aortogram showed a diminutive ascending aorta with retrograde visualization of the coronary arteries (Fig. 4). Left ventricular volume measurements were calculated for two consecutive beats using the area-length method (Dodge *et al.*, 1960). The left ventricular end diastolic volume was 49 cm³/m² and the left ventricular end systolic volume was 18 cm³/m², giving an ejection fraction of 64 per cent.

TABLE Haemodynamic data (Case 1)

Site	O ₂ saturation (%)	Pressure (mmHg)
Superior vena cava	67	
Right atrium	79	Mean = 11
Inferior vena cava	68	
Right ventricle	85	80/10
Main pulmonary artery	86	85/50 (65)
Left atrium	94	Mean = 13
Left ventricle	92	90/12
Descending aorta	84	85/36 (50)

Conversion from traditional units to SI units: 1 mmHg ≈ 0.133 kPa.

After the catheter procedure, a 6 mm Potts anastomosis was constructed, and bilateral pulmonary artery banding was performed. The procedure was well tolerated, and the infant was discharged much improved, two weeks after operation. Six months after operation, her growth and development were satisfactory and she was not admitted to hospital again.

Case 2

This male infant presented at 3 days of age with mild congestive heart failure. Cardiac examination showed normal pulses, a single second heart sound, and a soft systolic murmur.

Chest x-ray film showed laevocardia, moderate cardiomegaly, and pulmonary over-circulation (Fig. 5).

The electrocardiogram was reported to show superior axis, right atrial and right ventricular hypertrophy (tracing is not available for reproduction).

Shortly after admission, the infant aspirated, sustained a cardiorespiratory arrest, and could not be resuscitated.

Pertinent necropsy findings were limited to the cardiovascular system. Laevocardia was present, and viscerotrial situs solitus, with the right-sided atrium receiving superior and inferior vena cava and coronary sinus. The morphological right ventricle

FIG. 5 Case 2: The chest x-ray film shows laevocardia, visceral situs solitus, and modest cardiomegaly. Pulmonary plethora is also present.

FIG. 6 Case 2. Internal view of morphological right ventricle. The dilated pulmonary artery (PA) originates above the crista supraventricularis (cs), and the pulmonary valve above is separated from the tricuspid valve component (tv) below. The right ventricular wall (RVW) is thickened (7.0 mm).

was right-sided and was hypertrophied and dilated (d-ventricular loop). A common atrioventricular valve was present, and there was considerable attachment of the common AV valve to the crest of the muscular ventricular septum. The subpulmonary conus was well developed, with semilunar atrioventricular valve discontinuity. The very large, dilated pulmonary artery originated above the subpulmonary conus (Fig. 6). The pulmonary valve was tricuspid. Pulmonary venous return to left atrium was normal. The left atrial chamber was of normal size. The mitral valve was grossly abnormal. The aortic leaflet of the mitral valve was cleft, and the distinct medial attachment of this leaflet of the mitral valve to the posterior edge of the grossly deficient ventricular septum was responsible for virtual subaortic atresia. The left ventricle measured 3.2 cm in vertical axis and appeared only slightly smaller than the grossly hypertrophied right ventricle (Fig. 7). The aortic valve was atretic and was formed of two, fused semilunar cusps. The diameter of the atretic aortic valve was 3.0 mm. The ascending aorta was diminutive (Fig. 8). The aortic arch was left-sided with normal brachiocephalic vessels. The ductus arteriosus was patent. In addition, there was evidence of aspiration, pneumonic, and hepatic congestion.

Final anatomical diagnoses: laevocardia, atrial situs solitus, d-ventricular loop, complete common

atrioventricular canal, subaortic and aortic valve atresia secondary to anomalous attachment of mitral valve, persistent ductus arteriosus, and normal left atrium and left ventricle.

Discussion

The recent reports of Bernhard and his colleagues lend impetus to the possibility that ventriculo-aortic reconstitution may be afforded some select patients with aortic atresia (Bernhard *et al.*, 1975). They successfully bypassed extreme valve and supra-aortic stenosis with associated hypoplasia of the aortic valve annulus in a 22-year-old patient with angina and syncope that had proved refractory to standard surgical intervention. In their patient, a valved conduit was positioned between the left ventricular apex and descending thoracic aorta. The patient was significantly improved by the operation and the laevophase of a postoperative biplane pulmonary arteriogram showed that the contrast was ejected through the prosthesis, with none passing through the patient's own aortic valve and hypoplastic outflow tract.

There is an alternative approach to the use of a

FIG. 7 Case 2: Internal view of the morphological left ventricle. The left ventricle (LV) is of normal size. The large ventricular component of the complete AV canal is seen, and is not restrictive (arrow). The mitral valve has a cleft anterior leaflet, and extreme subaortic obstruction results from the abnormal medial attachment of the anterior leaflet of mitral valve. The left ventricular free wall (LVW) is of normal calibre.

FIG. 8 Case 2: External view of heart showing relation between pulmonary artery (PA) and ascending aorta (ao). The main pulmonary artery is anterior and is considerably larger than the ascending aorta. The diminutive ascending aorta (ao) originates posteriorly, inferiorly, and slightly to the left of the pulmonary artery. The relative size of the great arteries suggests the typical features of the hypoplastic left heart syndrome, with significant hypoplasia of left ventricle and mitral valve apparatus. The large right atrial appendage (RA) is also seen.

ventriculo-aortic conduit. The ventricular septal defect could be patched into the pulmonary artery, providing left ventricular pulmonary artery continuity. The origins of the right and left branch pulmonary arteries could be divided from the main trunk, and right ventricular pulmonary artery integrity could be established by using a prosthetic bifurcation graft from right ventricle to branch pulmonary arteries. The ductus and Potts shunt would then provide continuity to the descending thoracic aorta. This hypothetical technique has the advantage that the patient's own semilunar (pulmonary) valve would then serve as the patient's systemic semilunar valve.

The presence of an adequate left ventricle, without associated endocardial fibroelastosis or mitral valve hypoplasia, is a most unusual departure from the usual pathological findings in the patient with aortic atresia (Lev, 1952; Noonan and Nadas, 1958; Saied and Folger, 1972; Sinha *et al.*, 1968; Kanjuh, Eliot, and Edwards, 1965). It is difficult

to estimate the frequency of this association, but undoubtedly it is rare. Rosenquist *et al.* recently described an infant with aortic atresia, aortopulmonary fenestration, interrupted aortic arch, ventricular septal defect, and persistent ductus arteriosus. The left ventricle in their patient was 2.7 cm in length, and their comments and diagrams suggest that the left ventricle was of reasonable size (Rosenquist *et al.*, 1974).

A complete atrioventricular canal and aortic atresia with a diminutive ascending aorta are the pertinent pathological findings in Case 2. The subaortic area in this patient was extremely narrowed by abnormal insertion of the aortic leaflet of the mitral valve and by deposition of accessory endocardial cushion tissue. There are at present many reports describing the anomalous attachment of the mitral valve causing subaortic stenosis (Ferencz, 1957; Björk, Hultquist, and Lodin, 1961; Sellers, Lillehei, and Edwards, 1964). As Edwards has previously stated: 'a variety of mitral valvular lesions may be responsible for subaortic stenosis. There is, however, a consistent association between the mitral valvular lesion of persistent common atrioventricular canal (so called endocardial cushion defect) and actual or potential subaortic stenosis' (Jue and Edwards, 1967). Coexistent aortic and mitral atresia have been associated with persistent common atrioventricular canal (Silberberg, 1965). But, unlike our second patient, there was apparently aplasia of the left ventricle in the patient described by Silberberg. Indeed, significant hypoplasia of the left ventricle seems to be the rule in patients with atrioventricular canal and subaortic atresia.

Differentiation of the patient with aortic atresia and adequate left ventricle from the patient with aortic atresia and diminutive left ventricle and mitral apparatus is probably not possible on clinical grounds alone. Survival in both depends on patency of the ductus arteriosus and with ductal restriction systemic and coronary perfusion will become inadequate, metabolic acidosis will become irrevocable, and shortly the patient will die. The electrocardiograms will probably not be helpful in distinguishing between the two. Even if good left ventricular voltage were present, this should not be interpreted as suggesting an adequate left ventricular cavity. Strong, Liebman, and Perrin (1970) described 4 patients with hypoplastic left heart syndrome and electrocardiographic evidence for left ventricular hypertrophy. Similarly, in the review by Noonan and Nadas (1958) of the hypoplastic left heart syndrome, 4 of 50 patients had left ventricular hypertrophy on electrocardiogram. In one of 43 patients with aortic atresia studied at necropsy by Watson and Rowe (1962), the electro-

cardiogram had shown pure left ventricular hypertrophy.

The echocardiogram will show a diminutive aortic root and normal left ventricle, but tetralogy of Fallot with pulmonary atresia and large bronchials or truncus arteriosus must be differentiated from aortic atresia and normal left ventricle. Selective aortic and left atrial and left ventricular biplane angiocardiograms are probably needed to define the anatomical features.

Palliation of these patients in infancy will necessitate construction of a pulmonary-aortic shunt, with banding of the pulmonary arteries to prevent intractable congestive heart failure and to prevent pulmonary vascular obstructive disease (Sinha *et al.*, 1968; Cayler *et al.*, 1970; Van Praagh *et al.*, 1971). Atrial decompression may be accomplished in some by balloon septostomy at the time of the diagnostic catheter study. Ventriculoaortic reconstitution might then be accomplished using a valved conduit and repair of the intracardiac defect when the patient is older.

References

- Bernhard, W. F., Poirier, V., and LaFarge, C. G. (1975). Relief of congenital obstruction to left ventricular outflow with a ventricular-aortic prosthesis. *Journal of Thoracic and Cardiovascular Surgery*, **69**, 223.
- Bjork, V. O., Hultquist, G., and Lodin, H. (1961). Subortic stenosis produced by an abnormally placed anterior mitral leaflet. *Journal of Thoracic and Cardiovascular Surgery*, **41**, 659.
- Cayler, G. G., Smeloff, E. A., and Miller, G. E. (1970). Surgical palliation of hypoplastic left side of heart. *New England Journal of Medicine*, **282**, 780.
- Dodge, H. T., Sandler, H., Ballew, D. W., and Lord, J. D., Jr. (1960). Use of biplane angiocardigraphy for measurement of left ventricular volume in man. *American Heart Journal*, **60**, 762.
- Ferencz, C. (1957). Atrio-ventricular defect of membranous septum: left ventricular right atrial communication with malformed mitral valve simulating aortic stenosis: report of a case. *Bulletin of the Johns Hopkins Hospital*, **100**, 209.
- Jue, K. L., and Edwards, J. E. (1967). Anomalous attachment of mitral valve causing subaortic atresia. Observations in a case with other cardiac anomalies and multiple spleens. *Circulation*, **35**, 928.
- Kanjuh, V. I., Eliot, R. S., and Edwards, J. E. (1965). Coexistent mitral and aortic valvular atresia. A pathologic study of 14 cases. *American Journal of Cardiology*, **15**, 611.
- Keith, J. D., Rowe, R. D., and Vlad, P. (1967). *Heart Disease in Infancy and Childhood*, 2nd ed., p. 474. Macmillan, New York.
- Lev, M. (1952). Pathologic anatomy and interrelationship of hypoplasia of the aortic tract complexes. *Laboratory Investigation*, **1**, 61.
- Noonan, J. A., and Nadas, A. S. (1958). The hypoplastic left heart syndrome: an analysis of 101 cases. *Pediatric Clinics of North America*, **5**, 1024.
- Rosenquist, G. C., Taylor, J. F. N., and Stark, J. (1974). Aortopulmonary fenestration and aortic atresia. Report of an infant with ventricular septal defect, persistent ductus arteriosus and interrupted aortic arch. *British Heart Journal*, **36**, 1146.
- Saied, A., and Folger, G. M. (1972). Hypoplastic left heart syndrome. *American Journal of Cardiology*, **29**, 190.
- Sellers, R. D., Lillehei, C. W., and Edwards, J. E. (1964). Subaortic stenosis caused by anomalies of the atrioventricular valves. *Journal of Thoracic and Cardiovascular Surgery*, **48**, 289.
- Silberberg, B. (1965). Coexistent aortic and mitral atresia associated with persistent common atrioventricular canal. *American Journal of Cardiology*, **16**, 754.
- Sinha, S. N., Rusnak, S. L., Sommers, H. M., Cole, R. B., Muster, A. J., and Paul, M. H. (1968). Hypoplastic left ventricle syndrome. Analysis of thirty autopsy cases in infants with surgical considerations. *American Journal of Cardiology*, **21**, 166.
- Strong, W. B., Liebman, J., and Perrin, E. (1970). Hypoplastic left ventricle syndrome. Electrocardiographic evidence of left ventricular hypertrophy. *American Journal of Diseases of Children*, **120**, 511.
- Van Praagh, R., Bernhard, W. F., Rosenthal, A., Parisi, L. F., and Fyler, D. C. (1971). Interrupted aortic arch: surgical treatment. *American Journal of Cardiology*, **27**, 200.
- Watson, D. G., and Rowe, R. D. (1962). Aortic-valve atresia. Report of 43 cases. *Journal of the American Medical Association*, **179**, 14.

Requests for reprints to Dr. Robert M. Freedom
The Hospital For Sick Children, 555 University
Avenue, Toronto, Ontario, Canada M5G 1X8.