Anatomical variants in aortic atresia
Potential candidates for ventriculoaortic reconstitution

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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, angiocardiographic, and pathological findings in two patients are presented, with comments on palliation in the neonatal period, and the potential for ventriculoaortic reconstitution.

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).
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 angiocardiology were performed shortly after admission. The haemodynamic results are summarized in the Table. Left ventricular angiocardiology 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 aorta.

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

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

**FIG. 3a, b** Case 1: Frames from left ventricular cineangiocardigram 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.
thoracic aorta, and retrograde filling of a diminutive ascending aorta. The pulmonary artery laevoephase 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>
<thead>
<tr>
<th>Site</th>
<th>$O_2$ saturation (%)</th>
<th>Pressure (mmHg)</th>
</tr>
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<tbody>
<tr>
<td>Superior vena cava</td>
<td>67</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>79</td>
<td>Mean = 11</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>68</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>85</td>
<td>80/10</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>86</td>
<td>85/50 (65)</td>
</tr>
<tr>
<td>Left atrium</td>
<td>94</td>
<td>Mean = 13</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>92</td>
<td>90/12</td>
</tr>
<tr>
<td>Descending aorta</td>
<td>84</td>
<td>85/36 (50)</td>
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*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 laevoeardia, 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. Laevoeardia was present, and visceroaerial situs solitus, with the right-sided atrioventricular septum and inferior vena cava and coronary sinus. The morphological right ventricle

**Fig. 5** Case 2: The chest x-ray film shows laevoeardia, 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 ventriculooaortic reconstitution may be afforded some select patients with aortic atresia (Bernhard et al., 1975). They successfully bypassed extreme valve and supravalve 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 laeovphase of a postoperative biplane pulmonary angiogram 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 left-sided 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.
ventricle seems atresia with aortic atresia. There is an unusual departure from the usual 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; Bjork, Hultquist, and Lodin, 1961; Sellers, L'Heureux, 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 atriocentral 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 atriocentral 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 atriocentral 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 ducal restriction systemic and coronary perfusion will become inadequate, metabolic acidosis will become irreversible, 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-

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.
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 angiograms 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. Venticulaoaortic reconstitution might then be accomplished using a valved conduit and repair of the intracardiac defect when the patient is older.

References

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