Case reports

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Interrupted aortic arch type A associated with congenitally corrected transposition of great arteries and ventricular septal defect

Successful direct aortic anastomosis and pulmonary artery banding in an infant

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SUMMARY A case is described of congenitally corrected transposition of the great arteries (atrioventricular discordance, ventriculoarterial discordance) with interrupted aortic arch, a previously unreported association. Aortic continuity was achieved by direct surgical anastomosis. Pulmonary artery banding was subsequently necessary, because of heart failure associated with a large ventricular septal defect. A possible pathogenetic mechanism is suggested.

Interruption of the aortic arch is always associated with other cardiovascular abnormalities, such as persistent ductus arteriosus, ventricular septal defect, truncus arteriosus, or transposition of the great arteries.¹⁻³ The association of interrupted aortic arch with corrected transposition is referred to by van Praagh *et al.*,¹ but the only published case is included in the necropsy series of Sherman,⁴ and from his description could be interpreted as a form of univentricular heart rather than corrected transposition (now defined as atrioventricular discordance, ventriculoarterial discordance).⁵ Furthermore, Bliddal,⁶ reviewing previous publications, and Bjarke and Kidd,⁷ reporting a large series, found no case with the association of interrupted aortic arch and corrected transposition.

Interrupted aortic arch still carries a high mortality,² but many successful repairs have been reported. These have, however, usually involved the use of a graft, with the consequent likelihood of reoperation. Only four successful repairs by direct anastomosis in infancy have been reported.^{2 8-10}

We therefore report a case of interrupted aortic arch type A, associated with corrected transposition of the great arteries and ventricular septal defect, in which there was successful surgical restoration of aortic continuity by direct anastomosis.

CASE REPORT

A boy, born at full term weighing 4.13 kg, was referred to the Regional Cardiothoracic Centre at the age of 6 days because of progressive heart failure since birth. On examination he was in heart failure with slight central cyanosis and small volume peripheral pulses. The only auscultatory abnormality was a mid-systolic murmur, grade 1/16, at the left sternal edge. The chest x-ray film showed severe cardiomegaly and pulmonary plethora. The electrocardiogram showed sinus rhythm, a normal PR interval, a QRS axis of 180°, and right atrial and right ventricular hypertrophy, with a striking absence of left ventricular voltages and a lack of Q waves in the left chest leads. M-mode echocardiography showed two ventricular chambers, each containing an atrioventricular valve, with the larger ventricle lying posteriorly. Two semilunar valves were seen, the anterior valve lying to the patient's right and the posterior one to the left. It was not possible to show normal echocardiographic continuity between the posterior atrioventricular valve and a great vessel. The extreme difficulty experienced in consistently profiling the interventricular septum from the parasternal approach raised the possibility that it was abnormally orientated. Two-dimensional echocardiography was not available at that time in our unit.

Treatment was started with digoxin, diuretics, and oxygen. Cardiac catheterisation with cineangiography was performed at the age of 7 days, but because of technical problems only a lateral angiogram was obtained. The results suggested a diagnosis of transposition of the great arteries, ventricular septal defect, and interrupted aortic arch, with the descending aorta supplied by a large persistent ductus arteriosus.

Despite maximal medical treatment, he remained in severe cardiac failure. In view of this and the unsatisfactory initial investigation, a further catheterisation was performed at the age of 10 weeks. Aortography showed an interruption of the aortic arch distal to the origin of the left subclavian artery (type A, Celoria and Patton).¹¹ Pressures and oximetry showed two atrial and two ventricular chambers. Angiography showed a small anterior ventricle of right ventricular morphology, which was entered via an atrioventricular valve from the atrium of higher oxygen saturation (Fig. 1). This ventricle gave rise to an anteriorly placed aorta. Contrast passed from this chamber through a large ventricular septal defect into a larger posterior ventricle which was crescentic in shape and extended to the apex. A further angiogram was carried out in the larger ventricle, which had some morphological features of a left ventricle, and was entered via an atrioventricular valve from the atrium of lower oxygen saturation (Fig. 2). The large main pulmonary artery arose from this ventricle and contrast passed from it into the descending aorta via a

persistent ductus arteriosus. The diagnosis was, therefore, atrial situs solitus, atrioventricular and ventriculoarterial discordance (congenitally corrected transposition), and ventricular septal defect associated with interrupted aortic arch type A.

Four days after this investigation, a successful restoration of the aortic arch was carried out via a left thoracotomy. The descending aorta was disconnected from the ductus which was closed. The under side of the aortic arch was then directly anastomosed end-to-end to the descending aorta.

The operation resulted in good volume peripheral pulses but the baby continued to require digoxin and large doses of diuretics to control cardiac failure. His progress was however reasonably satisfactory, with steady weight gain, until the age of 4½ months when cardiac failure recurred despite medication. He now had signs of a high flow ventricular septal defect with an apical mid-diastolic murmur and an accentuated pulmonary component of the second sound.

It was felt that an attempt to close the ventricular septal defect would be extremely hazardous, and therefore at the age of 5½ months pulmonary artery banding was performed. Cardiac failure was immediately much improved and has not been a problem since then.

The child is now 15 months old, thriving, developing normally and on no drugs.

When real time echocardiography became available subsequently, it was confirmed that there was ventricular inversion associated with a ventricular

Fig. 1 (a) Posteroanterior and (b) lateral views of injection into the small, anterior ventricle, which was heavily trabeculated and gave rise to the interrupted aorta.

Fig. 2 (a) Posteroanterior and (b) lateral views of injection into the large, posterior ventricle, which was finely trabeculated and gave rise to the pulmonary artery.

septal defect. Using a parasternal "5 chamber view" (tomographic section 15, Tajik *et al.*),¹² two ventricular chambers and an infundibular ventricular septal defect were easily visualised. With transducer angulation to a parasternal "4 chamber view" (Fig. 3) (tomographic section 16, Tajik *et al.*),¹² the inlet ventricular septum was seen to be intact, but its relation to the atrioventricular valves was abnormal. The left atrioventricular valve ring was attached to the interventricular septum in a more apical position than was the right (that is the reverse of the normal situation). In addition, a single large papillary muscle was seen to take origin from the free wall of the right-sided ventricular chamber, whereas no such structure could be visualised within the left. These echocardiographic features are fully compatible with the diagnosis of congenitally corrected transposition.

Discussion

Interrupted aortic arch is widely believed to be the result of malformations producing reduced aortic blood flow *in utero*. Lesions implicated have included subpulmonary ventricular septal defect with pulmonary override of the left ventricle, mitral valve obstruction, and truncus arteriosus with preferential flow to the pulmonary circulation.^{3 11 13} In this case, however, there was no evidence of pulmonary override, nor was there obstruction of the atrioventricular valves or ventricular outflow tracts. One can only speculate as to what might have been the cause of reduced aortic blood flow. The morphological right ventricle, giving rise to the aorta, is certainly

hypoplastic, and its small capacity may have restricted the normal intrauterine right-to-left flow of blood through the foramen ovale. This would result in a relatively large amount of blood taking the route:

Fig. 3 (a) Real time two-dimensional four-chamber view echocardiogram and (b) corresponding diagram. RA, right atrium; LA, left atrium; PV, pulmonary vein; IVS, interventricular septum; AV valve, atrioventricular valve.

RA-LV-PA-ductus-descending aorta, and relatively little going: RA-LA-RV-ascending aorta. The primary cause of the right ventricular hypoplasia is, however, unidentified.

Despite its complexity, this case was eventually diagnosed as atrioventricular and ventriculoarterial discordance by a combination of M-mode and two-dimensional echocardiographic information with angiography and catheterisation. It therefore appears to be the first reported case of congenitally corrected transposition associated with interrupted aortic arch. It was surgically treated by closure of the persistent ductus arteriosus, restoration of aortic continuity, and later banding of the pulmonary artery. This is only the fifth reported case of successful correction of interrupted aortic arch by direct anastomosis in infancy.

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