Persistence of third aortic arch with fourth aortic arch agenesis

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SUMMARY This report describes a case in which a persistent third aortic arch connection partly palliated interruption of the aortic arch. The malformation occurred without other cardiovascular abnormality in a newborn infant in whom the principal diagnostic clinical finding was the absence of all peripheral pulses except for those of the right brachial and carotid arteries. Echocardiographic and angiocardiographic examination showed a continuation of the left internal carotid artery to the descending aorta by way of an arching communication.

Interruption of the aortic arch is an unusual anomaly which most frequently occurs in association with other, often complex, cardiac malformations.¹⁻⁴ In such cases the condition is invariably ductus dependent and is associated with a high mortality rate in infancy.^{2,3}

Only rarely does the abnormality occur as a solitary malformation that is not associated with other cardiac defects and is not ductus dependent.⁵⁻⁷ In such instances the circulation is adequately maintained, as in simple coarctation of the aorta, by collateral vessels.⁸

We describe an infant with aortic arch interruption in whom the descending aorta was supplied only by a small persistent third aortic arch.

Case report

When we first saw this three day old female infant (4300 g) she was in a state of shock. She was pale and mottled, tachypnoeic, and pulseless—although when she had been discharged from the nursery 36 hours before she had been thought to be healthy. After admission and stabilisation procedures, which included peritoneal dialysis for anuria and azotaemia, we found that only the right carotid and axillary pulses were palpable. Blood pressure in the right arm was 130/70 mm Hg. A systolic ejection murmur radiating only to the right carotid artery was heard.

The electrocardiogram showed biventricular hy-

pertrophy and the chest radiogram showed cardiomegaly and prominent pulmonary vasculature. There was a prominent supracardiac shadow on the left of the cardiac contour.

ECHOCARDIOGRAPHY

Cross sectional echocardiography (Fig. 1) with multiple viewing projections did not show any intracardiac malformation. Left ventricular wall thickness, however, was increased and the internal diastolic dimension was reduced. The valves and the septa appeared normal. The suprasternal notch view showed an interruption of the aorta distal to the left carotid artery. This vessel followed an abnormal course, turning inferiorly to communicate directly with the descending aorta. The communicating vessel was smaller than the descending aorta which also appeared to be hypoplastic.

CARDIAC CATHETERISATION

Right heart pressures were slightly raised especially right atrial and right ventricular end diastolic pressures (Table). An atrial left to right shunt was present and was considered to be secondary to an incompetent foramen ovale. Left atrial pressure was higher than right atrial pressure. Left ventricular pressure was considerably increased.

Left ventriculography (Fig. 2) showed reduced cavity volume (31 ml/m²) and mitral regurgitation. The aorta appeared to be normal up to the level of the left carotid artery where it terminated. The left carotid artery continued normally to the level of the left clavicle where it arched abruptly to communicate

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Fig. 1 Suprasternal long axis echocardiogram. Ascending aorta (aao) terminates at level of origin of left carotid artery (lca). Area where normal aortic arch is expected is identified by open arrow. Note continuation of left carotid artery to descending aorta (dao) by way of an arching communication (encircling arrows). Arrows (aov) identify level of aortic valve.

Table Haemodynamic data

Site	Pressure (mm Hg)	Oxygen saturation (%)
SVC		62
IVC		61
RA	A = 13, V = 12, m = 10	79
RV	60/2/13*	79
PA	59/23 m = 37	78
LA	A = 20, V = 18, m = 13	94
LV	149/4/18*	93

IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava; A, atrial "a" wave; V, atrial "v" wave; m, mean. *Systolic pressure/absolute diastolic pressure/end diastolic pressure.

with the tortuous hypoplastic descending aorta below. A left subclavian artery of normal appearance arose from the arch formed by the left carotid artery, and gave rise to an enlarged left vertebral artery in the usual fashion.

No cause for the circulatory collapse, other than the cardiovascular malformation, could be identified. The infant is now 13 months old and remains in satisfactory condition, although her blood pressure is considerably raised (average 180/100 mm Hg). She currently requires no anti-congestive treatment. Her electrocardiogram shows

only mild left ventricular hypertrophy; her heart size is within normal radiographic limits but the cardiac contour remains abnormal.

Discussion

Aortic interruption occurring without other cardiac abnormalities is a rare malformation that is apparently better tolerated than its ductus dependent counterpart, which is found in association with other cardiac malformations.⁵⁻⁷ Indeed, the eight currently recorded cases of isolated interruption have all occurred in older individuals, whereas nearly 300 infants with combined additional cardiovascular abnormalities have been reported.¹⁻⁴

Our patient is the youngest case that we know of with isolated aortic interruption. The shock-like condition which caused her to be evaluated suggests acute ductal closure. If this did occur, however, the hypoplastic appearance of the descending aorta would be an unexpected finding, because in these circumstances prenatal ductal flow would be assumed to be adequate.

Our patient has aortic arch interruption. Unlike the usual interrupted aorta, however, there is direct communication of the left carotid artery with the descending aorta. There thus appears to be fourth

Fig. 2 Cineangiogram in the frontal projection after left ventricular injection. Note continuity of left common carotid artery (lca) with the hypoplastic descending aorta by way of a structure resembling the embryonic third aortic arch (aa3, small arrows). Aortic segment between black arrows appears to represent the left dorsal aorta (ldao). Aortic segment between third aortic arch and origin of left subclavian artery (lsa) represents persistence of dorsal aorta occurring between embryonic arches three and four. Faint pulmonary arterial opacification can be seen; this occurred because contrast material was regurgitated to left atrium and the atrial left-right shunt. The origin of the right subclavian artery (rsa), which is not well seen in this illustration, is from the innominate artery (inna). lva, left vertebral artery; rca, right common carotid artery.

aortic arch agenesis with the interruption partly corrected by persistence of a patent third aortic arch connection. This anomaly therefore resembles the malformation of cervical aortic arch⁹ but with hypoplasia of a large segment of the aorta. To our knowledge such anomaly has not been recognised before.

EMBRYOLOGICAL CONSIDERATIONS

The third aortic arch can be identified in human fetuses at between 27 and 42 days' gestation.¹⁰ Although it maintains truncal and dorsal aortic continuity throughout this period it only briefly forms the principal connecting pathway. At maturity each third aortic arch forms the proximal portion of the internal carotid arteries.¹⁰

As the portion of the dorsal aorta between the third and fourth arches (the carotid duct) undergoes degeneration, the fourth and to some extent the sixth aortic arches become the principal routes of truncal-aortic continuity.¹⁰ The left fourth aortic arch continues to undergo maturation to become that aspect of the aorta between the origin of the left common carotid artery and the entrance to the aorta of the ductus arteriosus, which is itself derived from the sixth aortic arch.

The malformation we describe therefore seems to be a primitive left carotid continuation with the aorta, which is analogous to a persistent carotid duct. No fourth aortic arch is represented. We found no evidence of the persistence of a sixth (ductal) arch. On the contrary, the hypoplastic appearance of the proximal descending aorta at the level at which ductal entrance usually occurs suggests a failure of development of the left dorsal aorta. This, in turn, indicates that the ductus failed to contribute its expected volume of blood from the fetal pulmonary circulation. Nowhere was there angiographic evidence of the usual outpouching from the aorta (ductus ampulla) that represents the point of previous ductal entry into the aorta.¹¹

This malformation is clearly not amenable to the usual direct approach to surgical repair of aortic interruption.¹² The calibre of the hypoplastic segment of the descending aorta seems to preclude its use in a primary repair necessitating an ascending aortic to abdominal descending aortic graft.

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