Isolation of the left common carotid or left innominate artery

LANCE V FONG, ALEXANDER W VENABLES

From the Department of Cardiology, Royal Children's Hospital, Victoria, Australia

SUMMARY Isolation of the left common carotid or left innominate artery from the aortic arch is rare. A six week malformed infant with a right aortic arch had isolation of a left innominate artery and connection to the pulmonary artery by a left ductus arteriosus. A right ductus arteriosus had been ligated. Another infant with a right aortic arch and ostium primum atrial septal defect was shown to have an aberrant left subclavian artery arising from the lower descending aorta. The left common carotid artery filled retrogradely and drained into the pulmonary artery by a left ductus arteriosus. This abnormality has not been reported before.

Many forms of aortic arch abnormalities have been described.^{1,2} Isolation of a left subclavian artery with connection to the pulmonary artery by a ductus arteriosus has been reported,³ but isolation of the left innominate artery from a right aortic arch and connection to the pulmonary artery by a ductus arteriosus is rare. Isolation of the left common carotid artery from a right aortic arch with an aberrant left subclavian artery has not been described before. We report one case of a right aortic arch with isolation of the left innominate artery and bilateral ductus and another of isolation of the left common carotid artery with an aberrant left subclavian artery.

Case reports

PATIENT 1

The first case was a six week old female infant with the CHARGE association (colobomata, choanal atresia, retarded growth and development, hearing deficit, and micrognathia). She was tachypnoeic with full peripheral pulses and a continuous murmur at the upper sternal edge consistent with a ductus arteriosus. Cross sectional echocardiography demonstrated a right aortic arch with normal intracardiac anatomy and a large ductus. After ligation of a right ductus a continuous murmur persisted at the upper sternal edge. Doppler echocardiography revealed persisting turbulence in the main pul-

monary artery. Aortography revealed early filling of three major vessels arising from the right aortic arch: the right common carotid, the right vertebral, and right subclavian arteries. These vessels joined plexuses of vessels in the neck, mediastinum, and right and left scapular areas. The left common carotid artery was noted filling late from above. Dye passed

Fig 1 Necropsy specimen of patient 1 with left common carotid artery (LCCA) and left subclavian artery (LSA) entering the left pulmonary artery (LPA) via a ductus arteriosus. The right common carotid artery (RCCA) arises from the right aortic arch (RAA). MPA, main pulmonary artery.

Requests for reprints to Dr Alexander W Venables, Department of Cardiology, Royal Children's Hospital, Parkville, Victoria 3052, Australia.

Accepted for publication 8 January 1987

Fig 2 (a) Aortogram (right anterior oblique view) of patient 2 shows the left subclavian artery (LSA) arising from the lower descending aorta. AA, ascending aorta. (b) Selective angiogram in the left vertebral artery via the aberrant left subclavian artery demonstrates late retrograde filling of the left common carotid artery (LCCA). Dye passes to the main pulmonary artery (MPA) via a ductus arteriosus (PDA).

from the plexuses into the left subclavian and left vertebral artery, which then joined the left common carotid artery. The left innominate artery then emptied into the left pulmonary artery, presumably via a left ductus. Dye could be seen spilling into both pulmonary arteries. This created a left to right shunt at the pulmonary artery level. There was no vascular ring.

Elective implantation of the left innominate artery to the ascending aorta was planned to remove the left to right shunt and prevent a steal syndrome. The infant died suddenly at home at six months of age. Necropsy confirmed the described abnormality of aortic arch vessels (fig 1) and the normal intracardiac structure. The vessels of the right neck and upper limb were connected to the left innominate artery by the circle of Willis and by collaterals in the neck and both scapular areas. The cause of death remained uncertain. The heart, brain, and other organs were normal.

PATIENT 2

A 10 month old female infant with primordial dwarfism presented with signs suggestive of a ductus arteriosus. Cardiac catheterisation showed an ostium primum atrial septal defect with a right aortic arch and a late filling and a left sided vessel entering the left pulmonary artery from the head and neck vessels. At five years of age follow up angiography demonstrated the right aortic arch from which arose the right common carotid, right subclavian, and vertebral arteries. An aberrant left subclavian artery arose from the right lower descending aorta (fig 2a). Selective injection of the right common carotid, right vertebral, and left vertebral artery via the aberrant left subclavian artery (fig 2b) showed late retrograde flow into the left common carotid artery, which received blood from the circle of Willis, vertebro-basilar system, and costocervical collaterals. There were no arteriovenous malformations in the cranium. Dye passed from the left common

Fig 3 (a) The hypothetical double arch of Edwards.⁷ (b) Interruption of the left arch in zones A and D with persistence of the left and right ductus allows development of the aortic arch of patient 1. There is a right aortic arch with isolation of the left innominate artery and bilateral patent ductus. (c) Distal migration of the left subclavian artery beyond the left ductus and concurrent interruption of the left arch in zones B and D would explain development of the aortic arch of patient 2. There is a right aortic arch with isolation of the left common carotid and aberrant left subclavian artery. AA, ascending aorta; DA, descending aorta; LCCA, left common carotid artery; RCCA, right common carotid artery; LSA, left subclavian artery; RSA, right subclavian artery; LPDA, left ductus arteriosus; RPDA, right ductus arteriosus.

carotid into the left pulmonary artery by a ductus arteriosus, creating a left to right shunt. This vessel was selectively outlined by passing a catheter from the pulmonary artery through the left ductus. There was no right patent ductus and no vascular ring.

The isolated left common carotid artery was ligated. The ostium primum atrial septal defect was repaired and a large left superior vena cava draining into the upper pole of the left atrium was channelled to the right atrium. Postoperative progress was uneventful.

Discussion

The Edwards' hypothetical double aortic arch explains aortic arch abnormalities by selective regression of various parts of either arch¹ (fig 3a). Right aortic arch with isolation of a left innominate artery connecting to the left pulmonary artery by a left ductus has been described rarely.^{4,5} Complete separation with regression of the left ductus arteriosus and the late development of a subclavian steal syndrome has also been reported.⁶ These abnormalities can be explained by interruption at two sites in

the double aortic arch—in the left posterior arch distal to the ductus and in the left anterior arch proximal to the left common carotid artery. The first infant had this abnormality with bilateral ductus arteriosus (fig 3b).

The second abnormality of isolation of the left common carotid artery with an aberrant left subclavian artery has not been described before. An explanation based on the Edwards' model is that distal migration of the left subclavian artery leading to the left ductus that enters the left dorsal aortic root just proximal to the origin of the left subclavian artery produces concurrent isolation of the left common carotid and an aberrant left subclavian artery. Interruption could occur in the left anterior arch proximal to the left common carotid and distal to the left patent ductus but proximal to the left subclavian artery and left dorsal aortic root (fig 3c). The variable proximity of origin of the left subclavian artery and ductal area has been noted in other conditions such as coarctation of the aorta.²

Late filling of left neck vessels providing a left to right shunt into the pulmonary artery was recognised fortuitously in both children, who had associated non-cardiac abnormalities. In both patients elective surgery was planned to prevent a later steal syndrome.^{3,6}

We thank Dr D W Boldt, Director of Radiology, Royal Children's Hospital, Victoria, and Dr C W Chow, anatomical pathologist, Royal Children's Hospital, Victoria, Australia, for their help.

References

- 1 Stewart JR, Kincaid OW, Edwards JE. *An atlas of vascular rings and related malformations of the aortic arch system*. Springfield, Illinois: Charles C Thomas, 1964:3–13, 80–129.
- 2 Freedom RM, Culham JAG, Moes CAF. Vascular rings and associated anomalies. In: *Angiocardiology of congenital heart disease*. New York: Macmillan, 1984:487–513.
- 3 Nath PH, Castaneda-Zuniga W, Zollikofer C, et al. Isolation of a subclavian artery. *Am J Roentgenol* 1981;137:683–8.
- 4 Karczenski K, Wozniwicz B. Abnormal site of origin of the common carotid and left subclavian arteries. *Pol Tyg Lek* 1978;33:1041–2.
- 5 Martin EC, Mesko ZG, Griep RB, Haller JO, Gordon DH. Isolation of the left innominate artery, a right arch and a left patent ductus arteriosus. *Am J Roentgenol* 1979;132:833–5.
- 6 Garti IJ, Aygen MM. Right aortic arch and isolation of the left innominate artery from the aorta. *Cardiovasc Intervent Radiol* 1982;5:235–7.
- 7 Edwards JE. Anomalies of derivatives of aortic arch system. *Med Clin North Am* 1948;32:925–49.