Equine Arch Vessel Anomaly
Associated with Coarctation of the Aorta*

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Angiography in a 30-year-old man revealed the unique combination of aortic coarctation and an unusual arch anomaly. Proximal to the coarctation, a single arch vessel trifurcated into the brachiocephalic, left common carotid and left subclavian arteries. This anomalous arch vessel is a normal equine variant. (Chest 1992; 102:634-35)

PCWP = pulmonary capillary wedge pressure; RA = right atrium

Coarctation of the aorta is associated with other congenital anomalies including bicuspid aortic valve, supravalvular and subvalvular aortic stenosis, patent ductus arteriosus, ventricular and atrial septal defects. More unusual associated anomalies include mitral valve atresia, mitral valve prolapse, anomalous origin of the left circumflex artery from the right pulmonary artery, cor triatriatum, sinus of Valsalva aneurysm, complete transposition of the great vessels and double outlet left ventricle.1,2

Anomalies of the arch vessels associated with coarctation are very unusual but include right aortic arch, aberrant right and left subclavian arteries and brachiocephalic arterial stenosis.3-5 In contrast, isolated aortic arch abnormalities are relatively frequent with the left carotid and brachiocephalic arteries arising from a common trunk and the origin of the left vertebral artery directly from the arch being the two most frequent. A rare anomaly in humans, seen frequently in horses,6 is a single arch vessel.

We report a patient with a single arch vessel originating proximal to a true coarctation of the aorta. This vessel trifurcated into the brachiocephalic, left common carotid and left subclavian arteries. Possible embryologic origins are considered.

CASE REPORT

A 30-year-old man was admitted to the hospital with upper extremity hypertension. There was a five-year history of headache with evaluation including a negative head CT and MRI. There was no history of lower extremity claudication or fatigue. Physical examination revealed no clubbing or cyanosis; the blood pressure was 170/110 mm Hg in both arms with diminished and delayed lower extremity pulses. The carotid arteries were 2+, symmetric and without bruits. Cardiac examination disclosed an S4 and a grade 2/6 posterior intrascapular systolic murmur. A chest x-ray film revealed borderline left ventricular enlargement, a low aortic arch at the level of the left pulmonary artery with poststenotic dilatation of the descending aorta. There was no rib notching. The electrocardiogram revealed left ventricular hypertrophy.

Right heart catheterization demonstrated normal hemodynamics (RA, 6; PCWP, 12) and cardiac output (5.1 L/min), without evidence for a left-to-right shunt by oximetry. There was a 45-mm Hg peak-to-peak gradient across the aortic coarctation (ascending aorta, 135/80 mm Hg; distal to the coarctation, 90/70 mm Hg), but no pressure gradient across either the aortic or mitral valves. Coronary angiography demonstrated normal origin of the coronary arteries and a normal right dominant system. An arch aortogram (Fig 1) demonstrated a normal ascending aorta and a tricuspid aortic valve. There was an aortic coarctation distal to the origin of a large single arch.

Figure 1. Left: Lateral arch aortogram shows a normal tricuspid aortic valve and coarctation of the aorta distal to the takeoff of the single arch vessel. Right: LAO arch aortogram shows the takeoff of a single arch vessel which trifurcates into the brachiocephalic, left common carotid and dilated left subclavian arteries. Both right and left internal mammary arteries are enlarged and there is also extensive collateral flow to the descending aorta through a network of small posterior vessels.

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vessel which trifurcated into the brachiocephalic, left common carotid and left subclavian arteries in normal sequence. An extensive network of collateral vessels to the descending aorta was visualized as well as prominent right and left internal mammary arteries. Magnetic resonance imaging confirmed the arch anomaly and demonstrated that the transverse aortic arch was anterior to the esophagus in its usual position. The coarctation arose 3 cm distal to the origin of the single arch vessel at the level of the ligamentum arteriosum.

The patient subsequently underwent successful surgical correction of the coarctation with placement of a 16-mm Goretex graft between the left subclavian artery and the descending aorta. Postoperative physical examination revealed no gradient between upper and lower extremity blood pressure readings (120/80 mm Hg).

**DISCUSSION**

This is the first report, to our knowledge, of a true aortic coarctation associated with an equine-type single arch vessel giving rise to the brachiocephalic, left common carotid and left subclavian arteries. Clarke and Dodrill described brachiocephalic and left common carotid arteries arising from a single orifice with a diminutive left subclavian artery partially fused to the left common carotid artery. This anomaly was associated with indentation of the aorta during systole resembling an eccentric coarctation. In contrast, the current report demonstrates a true coarctation distal to and separate from a single arch vessel. These findings were confirmed at surgery.

The right and left carotid artery systems develop from the third branchial arches and appeared normal in this patient except for their takeoff from the single arch vessel. The normal anterior position of the transverse aortic arch relative to the esophagus argues against a remnant right aortic arch. However, the left subclavian artery, which forms from the cephalad migration of the left seventh intersegmental artery, is abnormally fused with the left common carotid artery. Normally, the aortic arch distal to the left common carotid artery develops from the left fourth branchial arch. Abnormal development and distal hypoplasia of the left fourth branchial arch may have resulted in fusion of the left subclavian and left common carotid artery and distal coarctation.

**REFERENCES**


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**Long-term Prazosin Therapy for COPD Pulmonary Hypertension**

Jerzy Lewczuk, M.D.; Bożena Sobkowicz-Woźniak, M.D.; Piotr Pazko, M.B.; Jerzy Śpikowski, M.B.; and Krzysztof Wrabc, M.D.*

A 51-year-old patient with COPD, obesity, and pulmonary hypertension underwent long-term prazosin therapy after a successful hemodynamic response to 1 mg of oral prazosin. The 18-month administration of prazosin, in a dose of 3 mg a day, resulted in continued hemodynamic and echocardiographic benefits. (Chest 1992; 102:635-36)

\[ PVR = \text{pulmonary vascular resistance; SVR} = \text{systemic vascular resistance} \]

There is a general skepticism about using vasodilators currently available in the treatment of patients with pulmonary hypertension caused by COPD. Some vasodilators do not decrease pulmonary hypertension at all, and in others, an initial vasodilating effect fails to persist during observation. No evidence of prolonged survival of patients with COPD taking vasodilators has been shown.1,2 Among vasodilators alpha-1 postsynaptic nicotinic blockers have some advantages. They inhibit reflex sympathetic activity and do not abolish pulmonary arterial hypoxic pressure response.2,1 In a few clinical acute trials, they produced a potent vasodilating effect.1,2

**CASE REPORT**

A 51-year-old, obese (height, 166 cm; weight, 86.5 kg) iron worker with a several-year history of chronic bronchitis entered the study after having recovered from pulmonary infection. He was in a stable clinical state. An acute trial consisted of Swan-Ganz catheterization before and 1 h after 1 mg of prazosin taken orally. Subsequently, hemodynamic and echocardiographic studies were performed after two weeks, six weeks, and 18 months of prazosin therapy. 1 mg three times a day. Simultaneous respiratory parameters, weight, systemic arterial pressure, and heart rate were measured. Hemodynamic parameters were measured 1 to 3 h after the last dose of prazosin, but a hemodynamic study after 18 months of therapy was performed 12 h after the last dose of prazosin. Pulmonary vascular resistance and systemic vascular resistance were calculated in resistance units according to standard formulae. During echocardiographic examination, we evaluated pulmonary hypertension from the flow velocity pattern in the pulmonary artery using commonly accepted Doppler indices: acceleration time (AT), corrected acceleration time (ATc) = AT/V R – R, right ventricular ejection time (RVET).

Prior to the study, informed consent was obtained, and the protocol was approved by the local Medical Academy committee for clinical investigation. The results of the study are given in Tables 1 and 2.

**DISCUSSION**

In a 51-year-old patient with advanced COPD, obesity, and pulmonary hypertension, 1 mg of prazosin, administered orally, decreased MPAP from 46 to 35 mm Hg (18 percent). *

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