Figure-Eight Kinking of the Aorta (Pseudocoarctation) Coexistent with Coarctation*

Liang-Miin Tsai, M.D.; Morgan Fu, M.D.; Chau-Hsiung Chang, M.D.; and Jui-Sung Hung, M.D.

An asymptomatic young man presented with a cardiac murmur and hypertension only in the right arm. Angiograms showed kinking of the aortic arch with a figure-eight appearance consistent with pseudocoarctation. Before the kinked segment, also demonstrated were a stenotic lesion in the aortic arch proximal to the left carotid artery and profound collateral circulations in the right thorax. These findings strongly suggested a rare combination of pseudocoarctation and true coarctation. His hypertension was corrected following surgery with a bypass graft between the ascending aorta and the abdominal aorta.

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Congenital kinking or buckling of the aorta is an abnormal elongation or redundancy of the aortic arch with a sharp downward angulation of the aorta at the fixed level of the ligamentum arteriosum.1,2 It is also termed pseudocoarctation because it yields similar chest x-ray film findings to those of classic coarctation, but it is different from coarctation by the absence of the following: (1) stenosis of the aortic lumen; (2) pressure gradient across the lesion, (hence, clinical hypertension); and (3) collateral circulation manifested by rib notching on the chest x-ray film or palpable collateral pulses in the thorax. Since Dotter and Steinberg3 introduced this term in 1951, there have been more than 150 reported cases of pseudocoarctation. Of these, only two cases had significant pressure gradients across the lesions, suggesting coexistence of coarctation;4,5 however, both cases did not have hypertension or evidence of collateral circulations. Herein, we report a case of kinking of the aorta with a figure-eight appearance not previously described. In addition, the patient had marked hypertension of the right upper limb and profound collateral circulations.

CASE REPORT

A 24-year-old asymptomatic man was referred for evaluation of hypertension and a cardiac murmur. His right arm was better developed than the left arm and both legs. The pulse was regular, with a rate of 88 beats per minute. The right carotid and right radial pulses were bounding, while the left carotid, left radial, and both femoral pulses were weaker and delayed. Blood pressures were 190/100 mm Hg in the right arm, 120/60 mm Hg in the left arm, 100/60 mm Hg in the right leg, and 100/60 mm Hg in the left leg. The left ventricular impulse was sustained with a palpable S4. There was a grade 4/6 systolic murmur accompanied by a thrill in the aortic area, with transmission to the right supraclavicular fossa and the interscapular area. A grade 3/6 continuous murmur was audible in the left supraclavicular fossa, with transmission to the posterior chest. The chest x-ray film showed a space-occupying lesion in the left upper mediastinum. Rib notchings were noted only on the right side. There was no typical sign of so-called figure "3" of the descending aorta. The ECG revealed left ventricular hypertrophy.

![Aorta Image](attachment:image.jpg)

**Figure 1.** A (left), Aortogram showing normal-sized ascending aorta (As Ao), markedly dilated brachiocephalic artery (BCA), slender aortic arch (Ao Ar) with figure-eight kinking, and dilated and tortuous right internal mammary artery (IMA). Ds Ao, Descending aorta. B (right), Delayed visualization of left common carotid artery (CCA) and left subclavian artery (LSC).
Two dimensional echocardiography showed discontinuity of the aortic arch and a markedly dilated brachiocephalic artery.

Cardiac catheterization was performed via the right brachial approach. The ascending aortic pressure was 190/108 mm Hg, and there was no systolic pressure gradient across the aortic valve. The pressure gradient between the ascending and the descending aorta was not measured due to an inability to pass the catheter from aortic arch to descending aorta, and due to the difficulty of puncturing the feeble femoral arteries. Aortograms (Fig 1A) showed a normal-sized ascending aorta and a huge brachiocephalic artery giving rise to a dilated right subclavian artery and right common carotid artery. The aortic arch tapered to a stenotic lesion, followed by an elongated and markedly kinked vessel with a figure-eight or "dumbbell" appearance. The right internal mammary artery was enlarged and tortuous. The left carotid artery and the left subclavian artery were not immediately visualized but were faintly opacified in the delayed phase after visualization of the collateral circulations, suggesting an obstruction proximal to the origin of the left carotid artery (Fig 1B). Selective angiography clearly showed the course of the figure-eight kinking of the aortic arch (Fig 2). There was no vessel or collateral circulation arising from this kinked aortic arch. No patent ductus arteriosus was demonstrated.

Surgery was performed using a woven Dacron bypass graft between the ascending aorta and the abdominal aorta. After surgery, blood pressures were 130/70 mm Hg in the right arm, 120/70 mm Hg in the left arm, 110/70 mm Hg in the right leg, and 102/68 mm Hg in the left leg. The patient was well without need of antihypertensive therapy at the latest visit three years after the surgery.

FIGURE 2. Aortogram by selective contrast injection into proximal aortic arch, showing arch tapering to constricted segment before assuming figure-eight course. BCA, brachiocephalic artery; As Ao, ascending aorta; and Ds Ao, descending aorta.


discussion

Our case was unique on several grounds. First, the patient had kinking or buckling of the aortic arch; however, in contrast to the usual "S"-shaped or corkscrew-like deformity, the kinking had a figure-eight appearance which has not been previously described. Secondly, a stenotic lesion in the aortic arch between the brachiocephalic artery and the left carotid artery was demonstrated by angiograms. Thirdly, the stenosis was functionally significant, as evidenced by hypertension in the right arm and right-sided unilateral collateral circulations manifested by rib notchings on the chest x-ray film and an enlarged right internal mammary artery on angiography. The previous findings strongly suggested that the patient had both pseudocoarctation and true coarctation; however, the coarctation was not histologically proven.

Pseudocoarctation of the aorta usually presents as a mediastinal density in the left upper mediastinum. Unlike coarctation, in which the aortic arch does not reach the clavicle, its aortic arch is elongated and may arise higher than the clavicle.1 It is occasionally mistaken for a mediastinal neoplasm or an aortic aneurysm. In such an instance, cardiac catheterization is usually indicated to confirm the diagnosis. Surgery is usually not indicated for this malformation. Since kinking of the aorta may rarely be accompanied by coarctation, careful clinical evaluation should be done, with particular attention to the peripheral pulses and blood pressures in all limbs, and to the presence of rib notchings on the chest x-ray film. On the other hand, in a patient with proximal hypertension and clinical results suggestive of coarctation, a relatively high aortic arch on the chest x-ray film, as seen in our case, should alert one to consider associated kinking or buckling of the aorta.

REFERENCES