COARCTATION OF THE AORTA AT OR ABOVE THE ORIGIN OF THE LEFT SUBCLAVIAN ARTERY

BY

R. H. BAYLEY AND J. E. HOLOUBEK

From the Charity Hospital of Louisiana and the School of Medicine of the Louisiana State University, New Orleans, Louisiana, U.S.A.

Received February 14, 1940

Coarctation of the aorta, according to Evans (1933), was first described by Morgagni in 1760. Blackford (1928) cites Barié to the effect that the first case was recorded by Paris in 1791. Abbott (1928) wrote a comprehensive discussion of 200 reported cases with autopsies, and estimated the statistical incidence of the various anatomical abnormalities. Blackford (1928) estimated the incidence of coarctation of the aorta as 1 in every 1550 autopsies, with the frequency in males twice as great as in females.

The so-called adult and infantile types of coarctation were discussed by Parker and Dry (1938). In the former the constriction is localized abruptly in the region of the aortic isthmus ; the lumen may be completely closed, the collateral circulation is extensive, and a patent ductus is rare. The latter, on the other hand, is characterized by a diffuse constriction between the origin of the left subclavian artery and the attachment of a patent ductus arteriosus.

Of the several theories advanced to account for the malformation, Blackford's (1928) seems the most reasonable. He explains it as due to the absence, atrophy, or imperfect development of the fourth left branchial arch. The various diagnostic features (Eisenberg, 1938 ; Dock, 1932 ; Ernstene and Robins, 1931 ; Ferris, 1935 ; East, 1932 ; Amberg, 1932 ; Woltman and Shelden, 1927 ; Deneke, 1925 ; and Hamilton and Abbott, 1928) are all illustrated in our case, except that murmurs were absent over the intercostal vessels.

It is apparently a rare form of the so-called adult type in which the localized constriction of the aorta lies at or just proximal to the origin of the left subclavian artery. Only 15 instances of this type seem to have been reported. Abbott (1928) discussed the post-mortem findings in 7 such cases and Parker and Dry (1938) reported another with a necropsy. King (1937) collected 10 additional clinical examples ; the reports of only 6 of these were available to us and only 3, in our opinion, fall into this classification.

These cases differ from the usual type in that the left shoulder and arm are for the most part excluded from the distribution of high-pressure blood flow. Erosion of the ribs by enlarged intercostal arteries, which is often absent in the rare variety of coarctation, is confined to the right hemithorax.

DESCRIPTION OF THE CASE

A Puerto Rican labourer, 32 years of age, was referred to the Charity Hospital of Louisiana at New Orleans for a determination of employability. When first seen (November 2, 1939) he stated that four years before, when he had a broken wrist, he had been told that he had high blood pressure and had been advised to do light work only. For the past 18 months palpitation and dizziness had become progressively worse, and finally permitted only the easiest kind of work without discomfort. Dyspnoea appeared only on marked exertion. Precordial pain had never been felt. There was no history of rheumatic or syphilitic infection.

Examination showed a well-developed man of sthenic habitus, 157 cm. tall, and weighing 62.2 kg. The retinal arteries were tortuous and slightly compressed the veins at the arteriovenous crossings. The eye grounds as well as the lips and mucous membrane of the mouth showed a purplish cyanosis. Pronounced pulsation of both carotid arteries was present and also a synchronous bobbing of the head; the tension was apparently equal and was increased above normal. The thyroid showed a moderate general enlargement without any nodules or bruit. Pulsation of the subclavian artery was palpable in the right supraclavicular fossa and was both visible and palpable below the clavicle. No corresponding pulsation was present in the left supraclavicular and infraclavicular fossæ. There was no visible or palpable pulsation in the supra-sternal notch and no venous distention was present.

The apex beat was in the fifth space, 10 cm. from the midsternal line, and was well localized and forceful. The heart sounds at the apex were loud. The first sound was split, and a soft, blowing systolic murmur was present. The intensity of the second sound was moderately and equally increased in the aortic and pulmonary areas. A moderately loud, harsh systolic murmur in the aortic area was transmitted upward to the sternoclavicular joint and was not accompanied by a thrill. The rhythm was regular and the rate 88 per minute. The blood pressure was 154/78 in the right arm, 98/78 in the left arm, 84/78 in the right leg, and 90/80 in the left leg. A distant soft systolic murmur in the right scapular region was thought to be transmitted from the aortic area. No pulsations, murmurs, or thrills were detected over the intercostal vessels. The volume of the right radial pulse was considerably greater than that of the left. Compression of the left carotid artery failed to alter the character of the left radial pulse. There were no visible or palpable pulsations of the abdominal aorta. The pulsations of the femoral arteries were soft, equal, and synchronous. There was a distinct lag of these pulsations behind those of the right radial artery. They were synchronous, however, with those of the left radial artery. The pulsations of the posterior tibial and dorsalis pedis arteries were easily felt.

The muscular development of the right shoulder and arm was abnormally heavy. The circumference of the right upper arm was 3 cm. larger than that of the left, and the impression of asymmetrical muscular development was thus confirmed. The other physical findings were normal.

The red blood cell count was 5,500,000 per c.mm. and the hæmoglobin was 98 per cent. (Sahli). The urine and the renal output of phenolsulphonophta-

lein were normal. The blood reactions for syphilis were negative. Radioscopy of the chest in the postero-anterior, left and right oblique, and lateral diameters showed marked cardiac enlargement, chiefly the left ventricle and the left auricle. There was also moderate enlargement of the pulmonary conus. A small aortic knob could be seen. The inferior borders of the dorsal segments of the ribs of the right hemithorax were eroded (Fig. 1). Barium in the

FIG. 1.—Radiogram, showing erosion of the ribs of the right hemithorax.

œsophagus confirmed these findings. Several attempts to visualize the aortic arch by taking radiographs of the chest following the injection of a 70 per cent. solution of diodrast into a large vein of the arm (Robb and Steinberg, 1939) were not very successful because of the abnormally small calibre of the deformed aortic segment (Fig. 2). Fig. 3, taken with a double string galvanometer of the Einthoven type, which recorded any two leads simultaneously, showed that the P-R interval was 0.16 sec. and the QRS duration 0.12 sec. The form of the curve indicated a complete left bundle branch block. From curves taken by pairing lead I, first with semidirect leads first over the right and then over the left ventricle, it was possible to determine (after the method of Wilson *et al.*,

FIG. 2.—Second anterior oblique view showing the descending aorta beyond the stenotic region. Diodrast was used.

FIG. 3.—(A) Leads I and II recorded simultaneously. (B) Leads I and III recorded simultaneously. The standardizations, reading from above downwards and from right to left, are such that 1 mV. is equivalent to 1.0, 1.05, 1.1, and 1.25 cm., respectively. The small time division is 0.02 sec.

1932) that the accession wave arrived at the epicardial surface of the left ventricle 0.04 sec. later than at the epicardial surface of the right ventricle.

DISCUSSION

The clinical picture in this case is considered typical enough to establish the diagnosis of coarctation of the aorta. The difficulty found in attempts at radiographic visualization of the aortic arch was taken as evidence that this region of the vessel was hypoplastic. The basal systolic murmur can be explained by a trigonoidation (Chisholm, 1937) of the aortic semilunar valves or, in the same dynamic sense, on the basis of a bicuspid aortic valve. The presence of a left bundle branch block is not surprising, for this tends to appear in the ventricle subjected to the greatest strain (Bayley, 1934). This case is apparently the first of the kind to be reported in which erosion of the ribs was present. As might be expected, the erosion was confined to the right hemithorax.

Because of the striking difference in the volume of the radial pulse in the right as compared with that in the left arm, the rare variety should prove easier to differentiate from essential hypertension than the common variety of the adult type of coarctation. In both, the differentiation is important for therapeutics and prognosis. Simultaneous palpation of the radial and femoral pulsations is probably the most convenient method of routinely excluding aortic coarctation; it would, however, seem unwise to rely on this procedure alone.

REFERENCES

- Abbott, M. (1928). *Amer. Heart J.*, **3**, 574.
 Amberg, S. (1932). *Libman Anniversary Volume*, **1**, 55.
 Bayley, R. H. (1934). *Amer. J. med. Sc.*, **188**, 236.
 Blackford, L. M. (1928). *Arch. intern. Med.*, **41**, 702.
 Chisholm, D. R. (1937). *Amer. Heart J.*, **13**, 362.
 Deneke, T. (1925). *Virchows Arch.*, **245**, 336.
 Dock, W. (1932). *J. Amer. med. Ass.*, **99**, 2024.
 East, T. (1932). *Proc. Roy. Soc. Med.*, **25**, 796.
 Eisenberg, G. (1938). *J. Pediat.*, **13**, 303.
 Ernstene, A. C., and Robins, S. A. (1931). *Amer. J. Roentgen.*, **25**, 243.
 Evans, W. (1933). *Quart. J. Med.*, **2**, 1.
 Ferris, H. A. (1935). *Canad. med. Ass. J.*, **32**, 276.
 Hamilton, W. F., and Abbott, M. E. (1928). *Amer. Heart J.*, **3**, 381.
 King, J. T. (1937). *Ann. intern. Med.*, **10**, 1802.
 Parker, R. L., and Dry, T. J. (1938). *Amer. Heart J.*, **15**, 739.
 Robb, G. P., and Steinberg, I. (1939). *Ann. intern. Med.*, **13**, 12.
 Wilson, F. N., Macleod, A. G., and Barker, P. S. (1932). *Amer. Heart J.*, **7**, 305.
 Woltman, H. W., and Shelden, W. D. (1927). *Arch. Neurol. and Psychiat.*, *Chicago*, **17**, 303.