

## ABSENCE OF PULSE IN BOTH UPPER EXTREMITIES DUE TO AN AORTIC ARCH ANOMALY

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The absence of pulses in both upper extremities, with normal or increased pressure in the lower ones, has been known as the so-called "pulseless disease" or Takayasu disease.

In the reported cases, the cause was an arteritis of the aortic arch and, although mentioned as a possibility, no case due to an aortic arch anomaly has been described.

In the case to be reported, the absence of pulses was due to a rather unusual anomaly of the aortic arch.

### *Case Report*

A 30-year-old man was admitted to the hospital ward with a right-sided hemiplegia, already improving. He was leading a normal life, working as a radio-operator, until 11 days before admission, when he complained of dizziness and headache, and after a few days woke up in the morning unable to walk or talk. Since then, he had improved rapidly. He had been admitted to hospital, when 15 with a diagnosis of heart disease. We were unable to obtain any further data on this previous examination. His father and a brother died of "cerebral apoplexy".

He was well developed, but there was difficulty of movement and speech due to the right hemiparesis. There was an easily visible and palpable collateral circulation in the back, more marked in the right intercostal spaces. Pulses were unobtainable in both upper extremities, while easily and forcibly palpable in both carotid arteries and legs. Blood pressure could not be registered in either arm by the auscultatory method. An external recording of the pulses was made, showing a marked delay and "rounding" of the arm curves (Fig. 1). The intra-arterial pressure (Fig. 2) was recorded in both carotid arteries, both femoral arteries, and the left radial artery, and was high and of normal contour in the legs and in the neck, and greatly diminished in the arm, with an abnormally delayed anacrotic limb. During the puncturing of the radial artery the jet of blood was continuous and very weak.

The apex beat was strong and located at the fifth intercostal space, at the left mid-clavicular line. The aortic second sound was accentuated. No murmurs were heard. Breath sounds were diminished at both lung bases. The abdomen was normal.

The nervous system examination showed a right hemiparesis, already rapidly receding.

The urine was normal. The chest X-ray showed opacification of both costophrenic angles, a heart of normal position and size, and marked erosion of the lower rib margins. The œsophagus was normal. The electrocardiogram showed signs of left ventricular hypertrophy. A heart catheterization was done and the data obtained were compatible with right heart failure, with slight increase in right ventricular, pulmonary artery, and pulmonary "wedge" pressures.

A *selective angiocardigraphy* (Fig. 3), with the catheter placed in the right ventricle, showed during the opacification of the aortic arch a single large vessel coming off from the arch and giving branches only for the neck. In the lateral position the plates showed the same aspect—a single large vessel, giving branches to the neck. Normal pyelograms were obtained at the same time.

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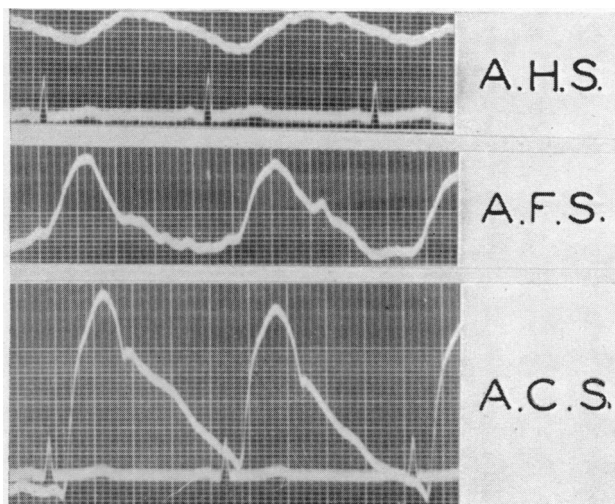


FIG. 1.—External recordings of the arterial pulses. A.H.S.—left humeral artery; A.F.S.—left femoral artery; A.C.S.—left carotid artery.

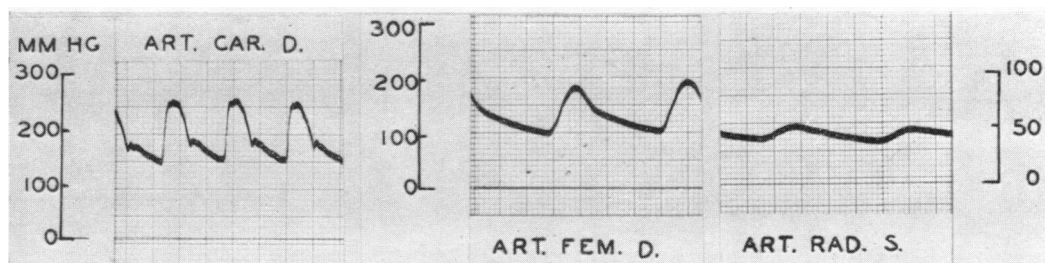


FIG. 2.—Intra-arterial recording of pressure.

*Evolution.* The signs of heart failure cleared up rapidly with digitoxin and the blood pressure came down to normal with reassurance and rauwolfia. Upon discharge, his speech and gait were almost normal.

#### Comments

The decrease or absence of pulses in both upper extremities has been reported in the arteritis of the aortic arch, comprising the so-called “pulseless disease”, the outstanding symptoms of which are visual disturbances and syncopal attacks.

The absence of these last-mentioned symptoms and the normal pulses in both carotid arteries allowed us, on a clinical basis, to discard an arteritis of the aortic arch and to suspect an aortic arch anomaly. Also in favour of a congenital anomaly was the history of a “heart lesion” when 15 years old and the normality of the upper extremities as far as temperature, trophic state, and strength were concerned.

Although mentioned as a possibility (Giffin, 1939), no case of pulseless disease due to an aortic arch anomaly has yet been reported. Another interesting feature is the great increase of the intercostal circulation and the rib notching, listing one more aetiology for this well-known X-ray sign: in this case the circulation probably functions in the reverse direction from aorta to arm.

A case exactly similar to the above reported was seen at the Hospital dos Servidores do Estado, Rio de Janeiro, Brazil. A 46-year-old heavy labourer when examined was found with hypertension in the lower

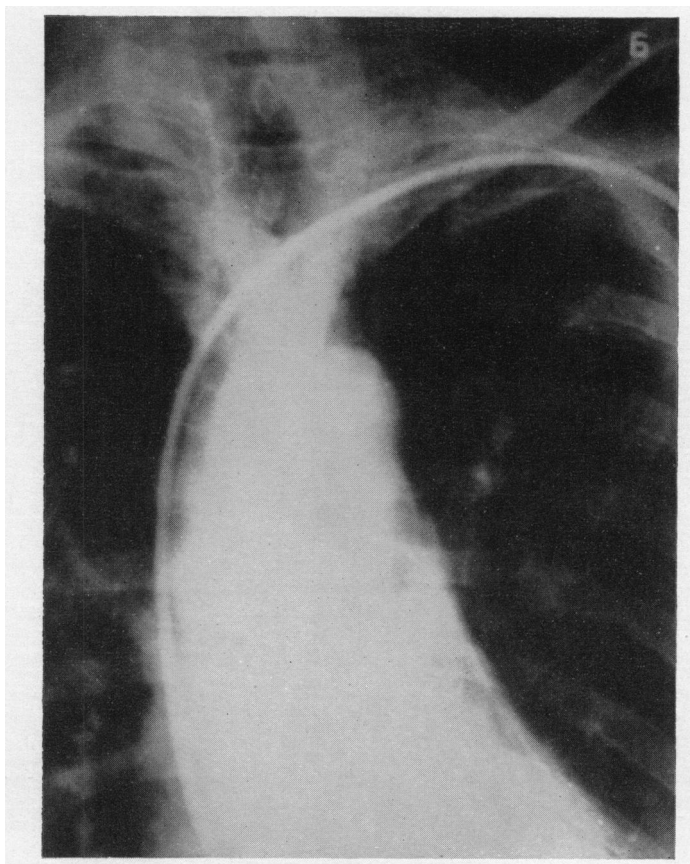


FIG. 3.—Angiocardiographic plate, showing the aortic arch opacified and a single large vessel coming off from it and giving branches only for the neck.

extremities (220/130), normal carotid pulsation, and extremely weak pulses in both arms. An aortography showed a single large vessel coming off from the aortic arch and giving branches only for the neck. A great increase in intercostal circulation was also seen in the aortography plates.

#### Summary

A 30-year-old patient is reported with absence of the pulse in both upper extremities. Pulses were normal in the leg and neck arteries. Intercostal collateral circulation and rib notching were observed. Angiocardiography showed a single large vessel coming off from the aortic arch and giving branches only for the neck.

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