Pulseless and Alive
Absence of All Aortic Arch Branches in a Healthy Toddler

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A cerebral ultrasound study on a neonate revealed abnormal blood flow in all intracranial arteries (Figure 1). The boy was scanned at 5 days of age as part of a Doppler ultrasound research program conducted at our department. Almost all neonates delivered at our hospital undergo imaging of the blood flow in the basilar artery. Perfusion is visualized when the head is turned to the left and right in both the prone and supine position. The aim is to investigate positional hypoperfusion of the brain stem, which is considered a risk factor for sudden infant death.1

The neonate was the second child of a 30-year old white woman. Pregnancy and birth were uneventful. He did not display any signs or symptoms consistent with a congenital abnormality or syndrome. Doppler ultrasound revealed an abnormal cerebral perfusion pattern with low flow velocities and a flattened, veinlike flow profile in all major intracranial arteries (anterior cerebral artery, both internal carotid arteries, basilar and both vertebral arteries) (Figure 1), whereas all abdominal arteries (celiac trunk and renal arteries) showed normal pulsatile flow. Echocardiography demonstrated normal anatomy of the heart. However, the origin of the great neck vessels at the aortic arch could not be identified, ie, absence of the right innominate artery, left common carotid artery, and left subclavian artery. MRI of the head showed normal morphology and vascular anatomy of the brain. The boy showed neither abnormal neurology nor signs suggestive of a circulatory disorder of the upper limb or brain. In the absence of any rational surgical option, it was decided to continue monitoring cerebral blood flow by Doppler sonography. Follow-up studies brought unchanged findings and unremarkable infantile neurological development.

To further assess vascular anatomy and possible therapeutic options, MRI angiography was performed when the boy was at the age of 2 years (Figures 2 through 4 and online-only Data Supplement Movie 1). It confirmed the absence of all branches of the aortic arch. The right innominate artery, right common carotid artery, right subclavian artery, parts of the right axillary artery, parts of the right vertebral artery, and the left subclavian artery were either absent or hypoplastic. The left subclavian artery was not connected to the aortic arch.

Figure 1. Initial findings during cerebral Doppler sonography: pathological Doppler flow in the anterior cerebral artery (ACA) with an almost veinlike flow pattern.
Cerebral perfusion was maintained predominately by the left vertebral artery augmented by collaterals. The collateral circulation supplying the left hemisphere and left upper limb comprised the following vessels: descending aorta, intercostal arteries, left internal mammary artery, left subclavian artery, and left vertebral artery. Both internal mammary arteries were also supplied through anastomoses with the celiac trunk via prominent phrenic arteries. Similarly, the right arm was supplied via connections between intercostal arteries and the right internal mammary and axillary artery. Because the right subclavian and common carotid arteries were absent, the filling of the right vertebral artery and right internal and external carotid artery was presumed to occur via anastomoses to thyroid, spinal, and muscle arteries of the neck.

The venous system, and the intracranial vascular anatomy, as well, appeared normal. Given that the extracranial part of the right internal carotid artery appeared hypoplastic, it was assumed that the intracranial part received retrograde perfusion via the circle of Willis and collaterals to the external carotid artery.

The boy’s anatomy does not match any malformation of the aortic arch and its branches reported to date. Considerations about etiology and prognosis can only be speculative. As with all congenital cardiovascular malformations, abnormal embryological development seems likely, because any alteration at a later stage of prenatal life would not allow the formation of a sufficient collateral circulation. The third, fourth, and sixth primitive aortic arches along with the seventh intersegmental arteries and the left dorsal aorta are the primary contributors to the normal aortic arch and its major branches. Obviously, all these structures must have been altered at some point during embryological development. It seems remarkable however, that the aortic arch, cardiac structures, and the venous system were not at all affected by such profound changes. Ultimately, the underlying etiopathogenesis in this case will remain uncertain.
The anatomy set out in this report is made all the more unique by the fact that the boy remains asymptomatic in the face of such gravely pathological cerebral perfusion. The child is 2 years of age at the time of writing. It remains to be seen whether development of the upper limb muscles throughout adolescence can be sustained despite these variations in vascular anatomy. Cerebral vascular perfusion studies will continue to be conducted at intervals, because this condition may be related to a higher incidence of intracranial aneurysm formation. Any therapeutic approach needs to be discussed when the clinical need arises, eg, in case of circulatory disorder of the right arm.

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Disclosures
None.

References
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