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THE IMPLICATIONS OF EXTENSIVE CEREBRAL VASCULAR DYSPLASIA IN SURGICAL REPAIR OF COARCTATION OF THE AORTA AND VENTRICULAR SEPTAL DEFECT

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Although acute neurologic problems are relatively common postoperatively in patients with arch anomalies,[1] associated anomalies of the head and neck vessels are not routinely excluded preoperatively. In this report we describe the case of a patient with severe dysplasia of the intracerebral vessels and incomplete circle of Willis who underwent successful concomitant repair of a long-segment aortic coarctation and ventricular septal defect (VSD) under hypothermic circulatory arrest. Despite a satisfactory cardiovascular outcome, he had a cerebral infarction during follow-up. Modern magnetic resonance (MR) methods may assist in the diagnostic work-up of patients with arch anomalies, including coarctation. Ischemic

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stroke caused by cerebroarterial disease in children with congenital heart disease will also be discussed.

Clinical summary. An asymptomatic, term, 9-month-old boy weighing 11 kg was referred for evaluation of a grade 3/6 pansystolic heart murmur. Transthoracic echocardiography demonstrated normal situs and intracardiac connections. A perimembranous VSD, partially plugged by accessory tricuspid valvular tissue and with 4 m/s left-to-right flow, was seen. However, the left-sided aortic arch was tortuous, with increased flow velocity up to 4 m/s near the origin of the left common carotid artery, suggesting the presence of a coarctation. Cardiac catheterization confirmed the VSD and demonstrated an aortic coarctation with a prominent ductal dimple at the site of the usual origin of the left common carotid artery, which was absent. Instead, its perfusion territory was supplied by multiple small collaterals. Furthermore, the right subclavian artery had an aberrant origin from an unusually low position on the descending aorta. Because of these irregularities, MR angiography was performed, demonstrating markedly tortuous, dilated, and dysplastic proximal intracranial vessels and lateral carotid arteries, particularly around the right M1, M2, and A1 segments. Extensive collaterals from the posterior circulation were seen on the right side, with a flow gap in the M2 region (Fig 1). An intact circle of Willis was not visualized. With full awareness of the higher associated neurologic risk, elective complete surgical repair of the aortic coarctation and VSD was undertaken.

Intraoperative findings included bilateral superior venae cavae, a perimembranous VSD, slightly dilated ascending aorta, and marked coarctation at the site of ductal insertion.

Only the right common carotid artery and the left subclavian artery were identified to derive from the aorta. Operatively, the ductus arteriosus was ligated, and the VSD was identified through a right atriotomy and closed with a bovine pericardial patch. Under deep hypothermic circulatory arrest (28 minutes), the undersurface of the aortic arch was opened and extended both proximally and distally. The defect was closed with a tailored aortic homograft patch. Measurements showed no gradient across the coarctation repair. The patient's condition remained stable throughout the perioperative period with no neurologic deficits and he was discharged home on postoperative day 5.

The patient remained well until 4½ months after discharge, when he had a sudden onset of focal seizure with twitching of the left side of the face and abnormal tongue movement lasting approximately 5 minutes without loss of consciousness. A diagnosis of febrile convulsion was made, and he was sent home. Twenty-four hours later, he was admitted to our institution with left hemiparesis and left-side facial droop. On examination, the cardiovascular status remained satisfactory; however, he was noted to have a left hemiparesis, affecting the upper limb more than the lower limb, and left VI and VII cranial nerve palsies. During the first hospital day, he had further seizures, which were controlled with diazepam and phenobarbital. Electroencephalography documented focal epileptic activities over the right centroparietal regions. MR imaging of the brain demonstrated extensive acute right temporal and frontal cortical infarction without hemorrhage. MR angiography showed no change from the previous study, but the apparent diffusion coefficient was low on diffusion-weighted imaging, which is compatible with an acute infarct.

After the first day, the patient remained seizure free, and by the end of his 12-day stay he had regained almost all of his left-side strength and was discharged home on a regimen of oral sodium valproic acid. He continues to make uneventful cardiovascular and neurologic recovery 21 months after the initial operation and is developmentally within normal limits. Direct revascularization of his cerebral circulation has been considered but deferred.

Discussion. Certain forms of congenital heart abnormality, particularly those involving the arch and aortic valve, are particularly associated with primary cerebrovascular disease, for example, Moyamoya syndrome,[2] dissection,[3,4] and aneurysm.[3] Abnormalities of vascular embryogenesis at the time of neural crest development and inherited connective tissue disease may be associated with cerebrovascular, as well as cardiovascular, disease. Given that a series from our institution found that acute neurologic problems in the postoperative period were most commonly associated with aortic arch abnormalities, including coarctation,[5] it is possible that the causative role of cerebrovascular disease has been underrecognized. We are aware of only one previously reported case of coarctation in which cerebrovascular disease was specifically looked for as part of the preoperative work-up and none with the extensive malformation, as reported here.[6] Our case was characterized by marked dysplasia of both extracranial and intracranial vasculature with proximal pruning and development of extensive collaterals. Because of the absence of an intact circle of Willis, the bilateral network of collateral vessels essentially becomes the source of cerebral perfusion. Indeed, the constellation of progressive narrowing of the intracranial internal carotid arteries and their main branches and compensatory collaterization is reminiscent of Moyamoya syndrome.[7]

Delineation of the cerebral circulation is not routinely performed in the evaluation of patients with aortic coarctation. Although our patient did not have preoperative neurologic symptoms of cerebral ischemia, abnormal aortic arch branching patterns prompted aggressive and thorough investigations. The combination of MR imaging and MR angiography has been shown in recent years to be a reliable method in the noninvasive evaluation of intracranial diseases. In this case identification of the impaired collateral cerebral circulation was crucial in the planning of the surgical strategy and perioperative management. In more difficult cases invasive 4-vessel cerebral angiogram may be necessary to provide comprehensive assessment of the anatomy.

Delgado and Barturen[6] elected not to offer surgical repair to their patient because of concerns regarding high risks for cerebral ischemia, spinal ischemia, or both. In accord with our policy of single-stage repair for most complex defects, we elected to proceed with complete repair of the VSD and coarctation under deep hypothermic circulatory arrest with both surface and core cooling. Agreeing with Jonas,[8] we believed it was important to reduce left ventricular and ascending aortic pressures and to reduce the deleterious effects of prolonged high pulmonary flow.

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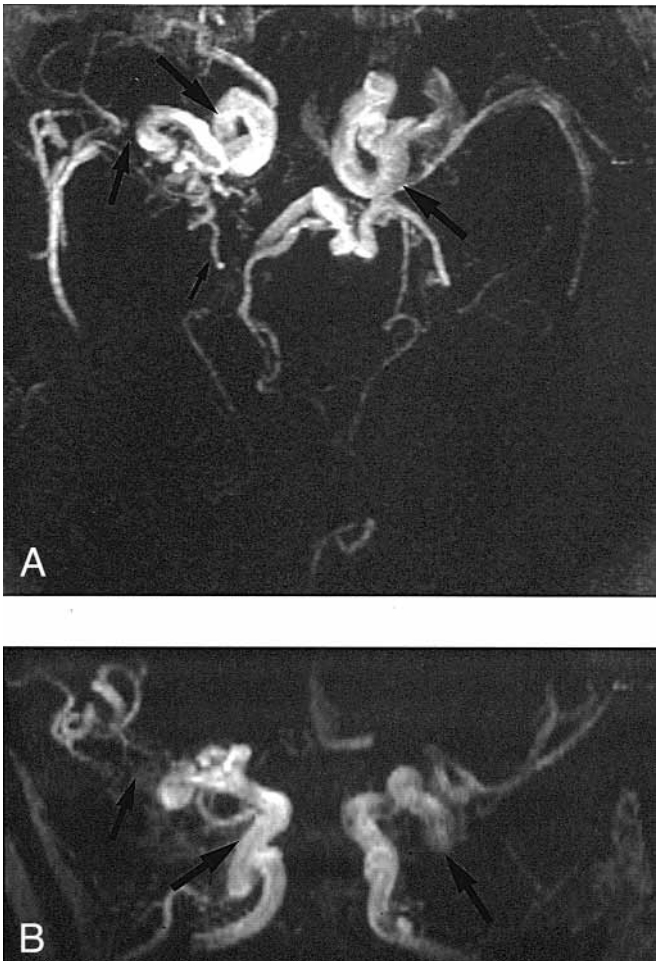

Fig 1. MR angiogram shown in transverse axial **(A)** and frontal **(B)** projections demonstrating dilated, ectatic, terminal internal carotids arteries bilaterally *(large arrows)*. Collateral vessels from posterior circulation are shown on the right *(small arrows)*, as well as a gap in perfusion in the right M2 region *(medium arrow)*.

Later, the patient presented acutely with seizures and a stroke, which could not be causally related to the perioperative period because they were demonstrated to be of very recent origin on diffusion-weighted imaging. There was no evidence for a cardioembolic cause or for venous sinus thrombosis. Moyamoya syndrome commonly manifests as recurrent episodes of transient hemiparesis, seizures, and cerebrovascular accidents as the result of cerebral ischemia. The therapy goal in this disease is to promote the development of collaterization to the brain and surgical pial synangiosis where the superficial temporal artery is anastomosed to the pia matter has shown promise. Our hope is that under close monitoring, natural collaterization would continue in this patient, resulting in freedom from further neurologic insults and a good quality of life.

In summary, we reviewed the implications of extensive cerebral vascular dysplasia and incomplete intracranial collateral circulation in the context of one-stage repair of VSD and aortic coarctation. The finding of abnormal head and neck anatomy in connection with a cardiac lesion should be thoroughly investigated to rule out intracerebral vascular anomalies. In cases such as this, diligent neurologic follow-up is imperative.

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