Carotid Arteritis: A Cause of Hemiplegia in Childhood*

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Cerebrovascular accidents are less common in children than in adults but the results may be just as devastating. Until recently, diagnosis of the mechanism of the "stroke" has been made either on the basis of clinical estimation, without arteriographic confirmation, or else by pathological findings.

Specific therapy ordinarily has been directed only to those patients in whom bloody spinal fluid or some other evidence of spontaneous hemorrhage has been present. Children suffering sudden "strokes" with no evidence of intracranial hemorrhage have been less well studied, understood and treated. The etiology of the neurological deficit in many such patients is never determined accurately and a purely clinical diagnosis of arterial thrombosis, embolism or spasm, or of cortical venous thrombosis is made.

Until recently there has been little ante-mortem documentation of cerebral arterial occlusion in childhood, and the number of reported cases is still quite small. A review of the literature made in 1960 by Wisoff and Rothballer yielded 27 cases, to which they added 2 of their own. In 1962 Byers and McLean in their discussion of childhood hemiplegia and aphasia reported cerebral arterial occlusion in 4 children. Three of these are included in this report.

A review of all symptomatic cerebrovascular accidents on file at the Children's Hospital Medical Center has been made for a 14-year period, 1948-1962, in an effort to clarify the frequency of cerebral arterial occlusions in the pediatric age group, and to analyze their clinical course. With the interest and assistance of the Pediatric and Neurology Departments an effort is being made to study by arteriography all cases of "bloodless strokes" as soon as possible after the onset of symptoms (Fig. 1). Neither arteriography itself nor the attendant general anesthesia which is advisable in children has produced any significant neurological or medical complications in these patients during the period covered by this report. The incidence of major cerebral arterial occlusion has been surprisingly high (Fig. 2). A total of 29 arterial occlusions, 4 partial obstructions and 1 recanalization has been documented by arteriography. Unlike bloodless strokes in adult patients, in children spontaneous arterial occlusions have been principally intracranial and less often in the extracranial carotid or vertebral circulation.

In 5 recent cases of cerebral arterial occlusion, surgical attempts were made to relieve the obstruction. With the help of microscopic examination of 2 surgically removed specimens, autopsy findings in a case previously reported by Banker and arteriographic findings in a case of hemiplegia in which only partial arterial obstruction was found, an effort has been made to deduce the pathogenesis of cerebral arterial occlusions in early life.

Historical
A) Embolism. It is well known that arterial embolism can occur in children. In most cases of embolus, however, there has been known congenital heart disease or obvious sepsis of the heart or lungs. 2, 19
B) Thrombosis. There is considerable evidence in the literature that in children thrombi may develop in cerebral arteries secondary to local abnormalities in the wall of the vessel.

1. Ulceration of intima. Ford described
autopsy material from a 3 11/12-year-old boy who died following a sudden hemiplegia in which thrombosis of the middle cerebral artery occurred at a point where there were intimal plaques of connective tissue containing lipoid deposits, with ulceration into the lumen of the vessel. The adventitia was normal and there was nothing to suggest an inflammatory process. Other cerebral arteries showed identical changes without thrombi and the visceral arteries showed similar less advanced lesions.

2. Changes in the media. Calcification in the media of small arteries has been reported in a 4-week-old infant's coronary, meningeal and cerebral vessels. The similarity between such lesions and experimental arteriosclerosis in animals has been suggested. Banker has found deposits of calcium in the media of vessels supplying atrophic areas of brain, such as in hydranencephaly and surgical specimens when hemispherectomy has been performed for intractable seizures. She has found this in the walls of vessels that have recanalized and feels that it is a nonspecific finding signifying only a previous occlusive process.

Several authors indicated that a severe acute generalized illness may somehow produce necrosis in the walls of blood vessels which then become the sites of thrombosis. Several authors have found that cerebral arterial occlusion has occurred at the site of collapse of the lumen by intramural dissection of blood through a defect in the intima. Norman and Urich in 1957 reported finding a recanalized dissecting aneurysm in a 15-year-old boy's middle cerebral artery. He had had a sudden hemiplegia at 6 months of age.

Wolman in 1959 found thrombus in a false passage beside the true lumen at the bifurcation of the internal carotid of a 16-year-old boy. This was associated with a minute aneurysm which arose from a congenital defect of the wall of the vessel, but did not even protrude beyond the adventitia.

Wisoff and Rothballer demonstrated subintimal fibrin thrombus in the internal carotid of an 11-year-old girl. There were multiple areas of focal medial necrosis in the vessels throughout the body suggesting a process such as polyarteritis nodosa.

C) Trauma. Direct trauma to the carotid in the tonsillar area or from contusion by the atlantoid process has been established as a cause of occlusion, presumably produced by periarterial inflammation. The significance of antecedent blows to the head and neck appears real and may be the result of sudden hyperextension of the neck. It has been demonstrated that emboli in the middle cerebral artery can originate from thrombi in the cervical carotid artery.

D) Localized Arteritis. There is evidence already in the literature for another possible mechanism of cerebral arterial occlusion.