

A case of internal carotid artery occlusion successfully treated by encephalo-duro-arterio-synangiosis

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Abstract. In many cases, intracranial occlusive strokes in children are treated symptomatically, with a diagnosis of acute infantile hemiplegia. This is because angiography is more difficult in children and, even if occlusive cerebral arterial disease is diagnosed, there is no definite therapeutic procedure. A case is reported of left internal carotid artery occlusion presenting with acute infantile hemiplegia and followed by repeated transient ischemic attacks. Encephalo-duro-arterio-synangiosis – developed as an operation for moyamoya disease – was performed on this patient, resulting in a marked revascularization of the brain in 8 postoperative months with good improvement in symptoms. This suggests that the operation may be effective in chronic cerebral ischemic diseases other than moyamoya disease.

Key words: Acute infantile hemiplegia – Pediatric stroke – Internal carotid occlusion – Encephalo-duro-arterio-synangiosis.

Intracranial occlusive cerebral arterial disease in children is a common cause of pediatric stroke. In most cases, treatment is palliative with a diagnosis of acute infantile hemiplegia, without the establishment of a definite etiology. This is probably because cerebral angiography is less easily performed in children, and also because there is no commonly accepted therapy for occlusive cerebral arterial disease.

Recently, we treated a child with left internal carotid artery occlusion, who had repeated attacks of reversible ischemic neurologic deficit or transient cerebral ischemia by encephalo-duro-arterio-synangiosis (EDAS) [9], a treatment developed for pediatric moyamoya disease. The result was a remarkable revascularization, confirmed by cerebral angiography performed 8 months after the operation.

Case report

A 3-year-old boy was apathetic for 1 day before 3 April 1981, when he could not stand up in the morning, suffering weakness in the extremities. He had no appetite and could not talk. There were no remarkable findings on the first computed tomography (CT) examination. After the attack, he began to take meals on the 3rd day, to talk on the 5th day, to sit on the floor on the 7th day and to stand up on the 8th day. CT performed on 13 May 1981 revealed a low-density area in the left frontal lobe. On 30 June 1981, it became difficult for him to walk, again for 2 days, but his gait returned to normal within a week. Cerebral angiography performed on 19 August 1981 revealed that the left internal carotid artery was completely occluded just distal to the origin of the ophthalmic artery (Fig. 1). The left cerebral arteries were opacified by the right internal carotid artery via the anterior communicating artery. On 6 May 1982, it again became difficult for him to stand up, but he returned to normal the following afternoon. On 5 October 1982 he had an attack of weakness following blood drawing, but returned to normal within 15 min.

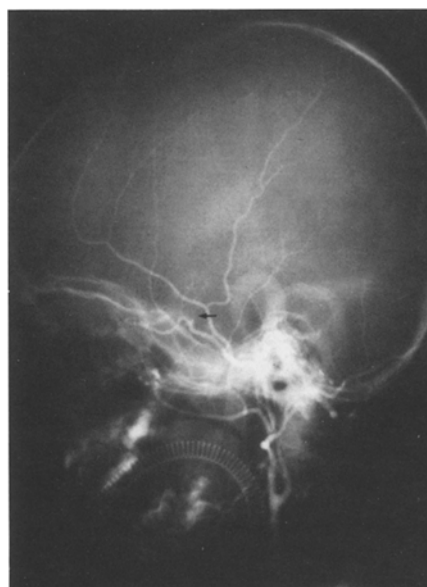


Fig. 1. Preoperative left common carotid angiogram (19 August 1981), 0.5 s after the injection of the contrast medium. Complete obstruction of the internal carotid artery (arrow) visible at the site immediately distal to the origin of the ophthalmic artery

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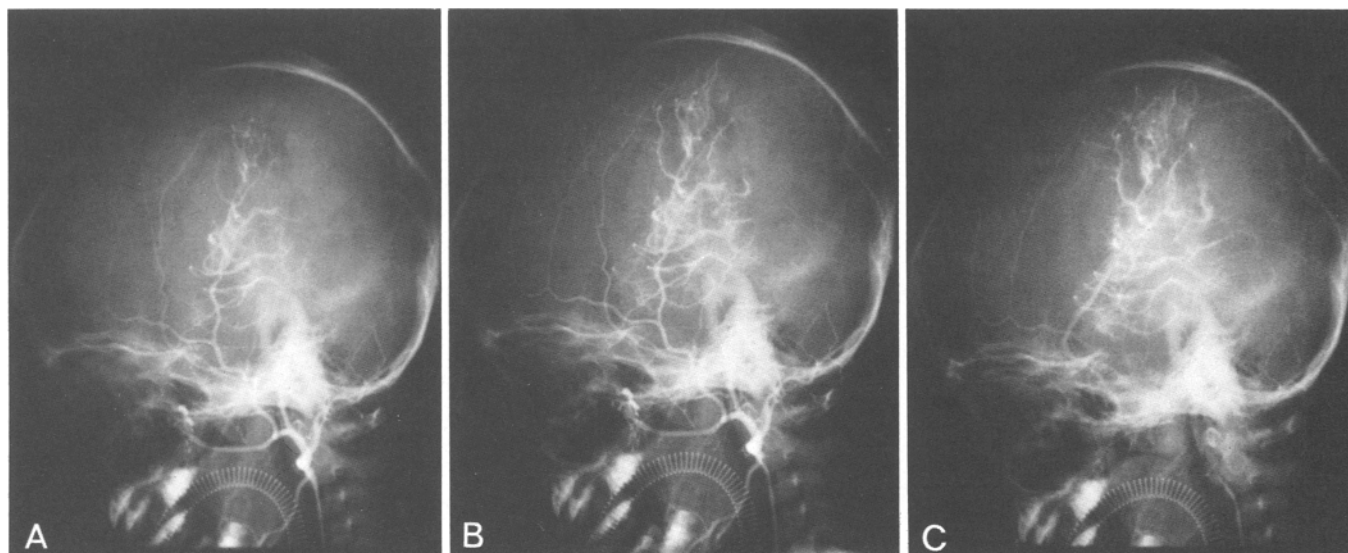


Fig. 2A–C. Postoperative selective left external carotid angiograms (5 September 1983) 10 months after EDAS. Revascularization of left middle cerebral zone is visible, **A**=0.5, **B**=1.5, and **C**=3.5 s after the injection of contrast medium

On 15 October 1982, he was admitted to our hospital. His blood pressure was 129/90 mmHg and his pulse rate was 112/min. Overall general examination was not remarkable, with no cardiomegaly or audible murmurs. The positive neurological signs included slightly hyperactive deep tendon reflexes on the left side, mild right-sided hemiparesis with slightly hemiparetic gait, and awkward usage of the right hand with no grasping power. Skull radiographs revealed a slightly smaller left cranial vault. CT showed slight widening of the left frontal subarachnoid spaces and a larger left lateral ventricle, probably indicating left frontal lobe atrophy.

On 29 October 1982, we performed EDAS [9] using the parietal branch of the left superficial temporal artery. The postoperative course was uneventful, and he was discharged on 13 November 1982. No transient ischemic attacks were observed thereafter, and the right hemiparesis has improved. On 5 September 1983, selective angiography of the left external carotid artery (Fig. 2) was performed: left cerebral revascularization from the donor scalp artery was clearly seen.

Discussion

It has been reported by many investigators that cerebral arterial disease is a common part of the etiology of acute hemiplegia in childhood [1, 2, 3, 11]. Among these reports, the largest series is reported in the textbook by Harwood-Nash [2], who diagnosed 65 cases of cerebral arterial occlusion with idiopathic or acute onset of hemiplegia who had previously been quite normal, with no evidence of intracranial or systemic disease of any kind. They classified them into three types (types I–III) from the viewpoint of cerebral angiography. Our case was confirmed to have unilateral intracranial internal carotid occlusion and belonged to their type IIa. According to Bickerstaff [1] and other investigators [2, 12], cases corresponding to type IIa often recanalize, and the symptoms improve with time. In contrast, our case manifested repeated transient ischemic attacks without recanalization during the 1-year observa-

tion period. There was complete occlusion of the left internal carotid artery (Fig. 1). Since the anterior circulation of the left cerebrum was supplied by the blood flow from the opposite anterior cerebral artery through the anterior communicating artery, sufficient blood flow could not be maintained when the demand for blood in the left cerebrum increased or when PaCO₂ was decreased by hyperventilation, resulting in a decrease in the total cerebral blood flow. Transient ischemic attacks were induced on such occasions. Thus, we thought that EDAS [9] – developed by us for the treatment of moyamoya disease, a typical chronic progressive cerebral ischemia in childhood – might be effective for the chronic ischemia in this case.

In order to explain this, we should describe our concept of collateral circulation in the case of chronic cerebral ischemia. According to our classification [6], the collaterals of the brain are composed of seven systems referred to as A–G surrounding the brain in laminar layers (Fig. 3): the A, B, and C systems are of the internal carotid artery, and the D and E systems are of the external carotid artery. These systems assume different functions during cerebral ischemia. For example, in moyamoya disease, which is a progressive failure of the B system, the collaterals of the A and C systems belonging to the same internal carotid artery system act immediately to compensate for the failure. Although the well-developed picture of the A system is noticed as the moyamoya vessels because of its distinguishing feature, the C system is not so noticeable because it is composed of the cortical arteries seen on a normal cerebral angiogram. When the D and E systems of the external carotid artery serve as collaterals through transdural anastomoses, the moyamoya vessels of the A system become reduced or disappear: spontaneous cure of pediatric moyamoya disease occurs [13]. This process

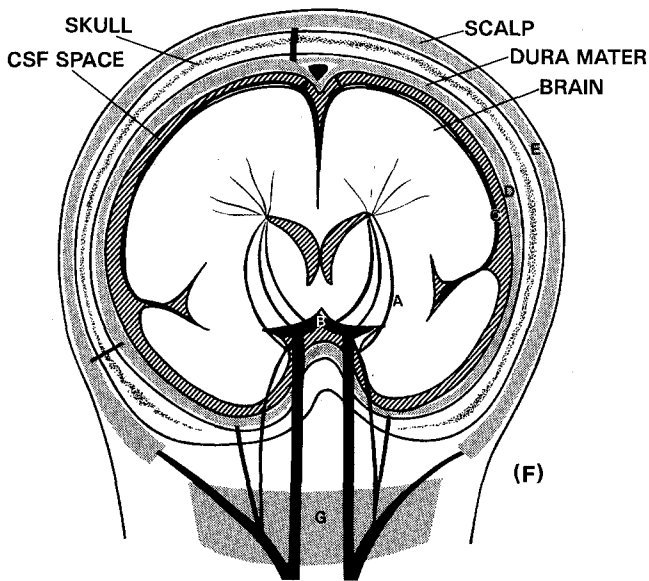


Fig. 3. Schematic illustration of collaterals to the brain. *A* system, Anastomosis intracerebralis; *B* system, basal communications; *C* system, cortical leptomeningeal anastomosis; *D* system, dural networks; *E* system, extracranial networks; (*F* system, functional collaterals); *G* system, ground communications

requires a great deal of time, because the layer of CSF in the subarachnoid spaces between the *C* and *D* systems interrupts their physical continuity. Also, the skull is a barrier between the *D* and *E* systems, inhibiting the formation or growth of collaterals. Therefore, in order to treat moyamoya disease surgically and successfully, one must both minimize the damage to the *D* and *E* systems and eliminate the obstacle, i.e., the subarachnoid fluid space and the skull. Thus, it is possible to produce the spontaneous healing of this disease by establishing the condition in which failure of the *B* system is completely compensated for by the *D* and *E* systems, without waiting for the lapse of time.

Since EDAS is an indirect EC/IC bypass operation based on the above-mentioned concept [6], and is designated to cause minimal damage to the *D* and *E* systems, it was expected that this operation would probably be effective for our case with the already described chronic ischemic condition. EDAS involves making a linear scalp incision along a donor scalp artery and freeing the artery for a certain length with a strip of Galea attached. The artery then is sutured with the strip of Galea onto the linear dural opening through a narrow cranial window made in the skull. The donor scalp artery is not cut, and the blood flow of this artery is not obstructed throughout the procedure. In this operation, incisions made on the *D* and *E* systems are all linear and parallel to the direction of thick arteries, minimizing damage to these systems [7, 9].

Although encephalo-myo-synangiosis (EMS) [4], and superficial temporal-middle cerebral artery (STA-MCA)

anastomosis [5] seem to be similar to EDAS, these methods are quite different in that they cause too much damage to the *E* and *D* systems, sacrificing the naturally and already formed collaterals and reserve collaterals by making large scalp and dural flap and/or cutting the superficial temporal artery, which is the main artery of the *E* system. EMS has further disadvantages in its use of muscle, from muscle contractile action, electrical potential activity, extensive adhesions, compression over the brain as subdural hematoma, wide bone defect to pass the muscle flap, and so on.

STA-MCA anastomosis also has some disadvantages. In children, both the donor scalp artery and the recipient cortical artery are quite narrow.

Skill in the technique is required, and thus the operative results are variable. Since it is necessary to close off the recipient artery for a period of time, focal brain circulation may be impaired. Furthermore, if anastomotic failure occurs, the vessels with abundant blood flow may be lost by thrombosis.

EDAS was developed to eliminate the disadvantages of these two methods and has been proved to work well in the treatment of moyamoya disease in children [7, 8, 10]. It seems that omental transplantation has been widely performed in China [14], but this method has the disadvantages of both EMS and STA-MCA anastomosis from the viewpoint of our concept of collateral formation, and should not be used as a therapeutic method for cerebral ischemia in children.

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Announcement

The 14th Scientific Meeting of the International Society for Pediatric Neurosurgery will take place in Madrid, Spain, September 30–October 3, 1986. *Chairman:* Francisco J. Villarejo, Prado del Rey, 65, Somosaguas, 28023 Madrid, Spain.