Cerebrovascular occlusion in childhood

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Among 2493 patients undergoing cerebral angiography 43 children were found to have cerebrovascular occlusion. The diagnosis, aetiology, management and care of the condition is outlined. It is stressed that angiography is indispensable in diagnosis.

Since the advent of cerebral angiography, occlusion of the blood vessels of the brain has been recognized with increasing frequency as a cause of acute hemiplegia in children. In several reviews special attention has been paid to the aetiology of hemiplegia in addition to the importance of clinical symptoms, localisation of the obstruction and its outcome [2, 5, 6, 11]. Solomon et al. [14] found 16 patients affected by cerebrovascular obstructive disorders among 86 children with acute hemiplegia. Chiofalo et al. [3] described detailed clinical and electroencephalographic findings of 26 children and young adults afflicted by occlusion of a cerebral artery. Livet et al. [8] reported 16 patients with cerebrovascular occlusion and reviewed the clinical and computer-tomographic diagnosis of the disorder in childhood. Hungarian authors [11] reported on 1200 angiographies carried out in children; among these 16 patients had cerebrovascular occlusion. Single cases with rare aetiology have also been described [4, 12, 13].

In the period 1955 to 1981, 2493 angiographies were performed in chil-

dren admitted to our institute. In 43 cases occlusion and in 14 stenosis was encountered in the supply area of the internal carotid arteries.

The localization of the occlusion was as follows. In 24 patients the main trunk of the carotid artery was occluded, in 9 the cervical and in 15 the intracranial section. In 15 patients the occlusion was on the right and in 8 on the left side and in one case on both sides. Complete occlusion of the cerebri media artery occurred in 10 patients, of the distal branches in 9 cases; of these, 11 were on the right and 4 on the left side.

Of the patients 20 were boys and 23 were girls. The mean age at onset is not known since several years may have elapsed between the appearance of the first, mild symptoms and the time of angiography.

The aetiological factors were as follows. In 11 cases the occlusion could be ascribed to infection, in 5 to congenital heart defect leading to embolization, in 7 to progressive proliferation of the intima, in 13 to cervical or skull injury and in 7 children no aetiological factor could be identified.

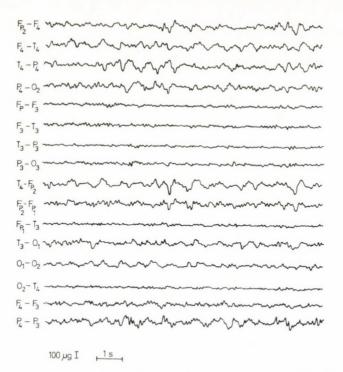


Fig. 1. H.B. 12 years old girl. EEG. The first four channels show the right side activity damaged by vascular occlusion and consisting mostly of theta and delta waves. On the left side slow alpha and fast theta activity indicating an affected circulation

Headaches and convulsions were the most frequent presenting symptoms, not infrequently acute hemiparesis or hemiplegia were the reason for admission. Hemiparesis or hemiplegia was often complicated by cerebral nerve palsy, in some instances by aphasia or partial reduction of the visual field. Sometimes symptoms of meningeal irritation or loss of consciousness were present. The eyeground findings were usually negative except in cases with pronounced cerebral oedema. Electroencephalography frequently revealed diffuse or focal deceleration accompanied in some instances by paroxysmal activity. The diagnosis was based on the angiographic finding (Figs 1, 2, 4).

The diagnosis can only be verified by angiography. Computer tomography may be helpful in determining the extension of the damaged area or in judging the subsequent evolution of collateral circulation within the supply area of the affected blood vessel (Fig. 3).

Earlier, anticoagulant (heparin) treatment was the rule. At present, improvement of the cerebral microcirculation and the prevention and relief of vasospasms by plasma expanders and papaverine or vinpocetin

over several days are the main aims of therapy. Adequate fluid intake, control of systolic blood pressure, treatment of eventual cerebral oedema, adequate oxygenation of the brain eventually by intubation, tracheostomy and mechanical ventila-

tion, treatment or prevention of eventual convulsions are of importance. After the acute phase of the disease, oral vinpocetin therapy is indicated for months or years. In one case a bypass was performed after the acute phase in order to improve



Fig. 2. Same patient as in Fig. 1. Anteroposterior and lateral carotid angiogram. Marked obstruction of the intracranial portion of the common carotid. The cerebri anterior artery does not fill from the right side, incomplete filling of the cerebri media artery

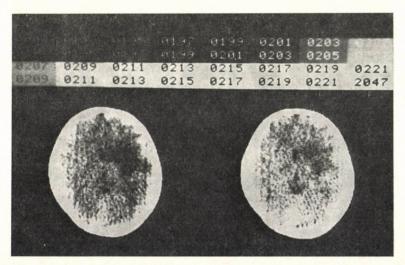


Fig. 3. Same patient as in Fig. 1. Computer tomogram of brain. Hypodensity of irregular shape in right precentral region corresponding to the vascular occlusion



Fig. 4. A. P. 11 years old boy. Anteroposterior and lateral cerebral angiograms show complete occlusion of cerebri media artery

collateral circulation, without any convincing effect.

In the symptoms are severe during the acute phase, lethal outcome may occur. No special measures are indicated in cases with mild symptoms. In the majority, motor handicap due to hemiparesis, aphasia, mental damage or convulsive disorders affect the quality of the patients' life.

Discussion

Occlusion of the trunk or some branch of the internal carotid artery is a comparatively frequent cause of hemiplegia in infants or children. The sites of predilection are the section adjacent to the branching of the common carotid, the intracranial section of the trunk, the initial part of the cerebri media artery and its distal branches. Occlusion of the anterior cerebral artery is an infrequent finding.

The onset may be acute or the symptoms may develop in several steps. The occlusion may be due to vascular malformation, toxic damage (drugs, narcotics, X-rays), inflammatory changes due to infection, tumours, injury, intimal proliferation of unknown aetiology or embolisation, especially in children affected by congenital heart malformations. Thrombosis of the internal carotid artery is a well-known complication of cervical injury or surgery. In blunt skull injury, damage to the temporal lobe may deteriorate microcirculation and provoke focal cerebral oedema leading to vasospasms or occlusion of the cerebri media artery [15].

In cases due to infection or toxic damage, symptoms pointing to cerebral vasospasm, such as headache, numbness in the extremities, eventually convulsions usually precede the full-blown clinical picture of complete occlusion. Fibromuscular dysplasia is a condition characterized by generalised malformations of the connective tissue of the blood vessels; the appearance of the carotid artery resembling a string of pearls is only one of the manifestations. The condition sooner or later leads to complete occlusion and is frequently complicated by arterial hypertension due to similar renovascular involvement.

The severity of clinical symptoms may vary from mild transitory hemisymptoms to severe hemiplegia or coma.

The patient's fate is largely determined by the nature of the underlying disease and the development of collateral circulation. In our experience, the occlusion is definite and leads to permanent neurological sequelae in most cases caused by a tumour or progressive intimal proliferation or when it occurs postoperatively.

Traumatic occlusion may cause severe neurological symptoms but often there is hope for remission. Occlusion due to inflammatory changes is frequently transitory and complete recovery may ensue. Collateral circulation usually develops in children with an occlusion of the trunk of the internal carotid provided that no other lesions are present. The prognosis is worse if the occlusion develops in the cerebri media artery near to its origin.

In therapy, acute thrombectomy is a possibility, but the time elapsing until angiography is usually too long, and after some hours no recirculation can be expected from surgery. After the acute phase it may be attempted to create a bypass. In patients needing non-surgical intensive care the main aim is to improve cerebral microcirculation and oxygenation. Surgical treatment for space reducing conditions like tumours or haematomas and medical treatment of diffuse or local cerebral oedema accompanying the occlusion may be indicated. Later on, these children need regular care and follow-up. Drugs such as vinpocetin preventing subsequent vasospasms caused by toxic agents or hypertension must be administered for months or even years.

The majority of survivors are affected by motor disorders, mental damage, and aphasia.

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