the Circle of Willis is thought to be caused by the delay of the blood supply to the cerebral tissues after hyperventilation because of the presence of the abnomal vascular nets. This is also supported by the facts that the appearance of slow wave is markedly suppressed either by the inhalation of 5% carbon-dioxide or by drip fusion of 40% papaverin chloride intravenously. In addition, the absence of the remarkable build up by hyperventilation in adult cases is thought to be due to the less susceptibility to the hyperventilation which causes hypoxia in the juvenile and the faster return to threshold of 20 mg Hg in internal juglar vein which evokes slow wave on EEG (Myer & Goto).

D-14. Cerebrovascular "Moyamoya" Disease among the Japanese, on Dynamic Changes of Vascularization

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A peculiar fine reticular or fibrillar arterial network spreading upward from a point of stenosis or occlusion of the terminal portion of the internal carotid artery in association with absence or poor visualization of the anterior and especially of middle cerebral arteries is a figure complex observed recently in Japan in carotid angiograms.

We reported 6 cases with such abnormal angiographical findings at 22nd Annual Meeting of the Japan Neurosurgical Society held in 1963. We also reported in details 10 cases with this disease in No to Shinkei (Brain and Nerve) No. 8. Vol. 10, 1965, and said on four subjects, as follows:

(1) The disease is not a congenital malformation, but is a acquired disease in which at first stenosis or occlusion occured in the terminal portion of the internal carotid artery due to some causes, and such abnormal vascular network has secondarily developed as collateral path-ways.

(2) The disease is found in only Japanese race now, especially in young female.

(3) Most of them has the inflammations on the upper region from neck in past history, and the abnormal findings in carotid angiograms were seen bilaterally.

(4) The symptoms of the disease is chiefly one of the cerebral ischemia, and the abnormal laboratory findings are not scarcely found.

At this time we are postulating a nomination for the disease as Cerebrovascular "Moyamoya" Disease.

The word "Moyamoya" is a unique expression of our language in that the meaning of the word itself is very obscure, and would be best understood when we depict something like faint cloud or smoke of a cigarette. The shape of the anomalous vascularization seen in cerebral angiograms is not altogether same with that of smoke of a cigarette, but they are resemble in many ways. The latter is distinctly visible at first with eventual fading away, and this is similar with the changes of the vascular network observed and anticipated during the course of the disease.

As mentioned before, the disease is seen almost exclusively among the Japanese, we will be entitled to describe it in Japanese. Besides the word "Moyamoya" is rhythmical and easy to pronounce and is easy to learn for foreigners.

In this report, we said about the entity of the disease. The summary of our statement is as follows:

1) This report is based on 20 patients with anomalous vascularization at the base of the cerebrum experienced clinically until September 1966. In 8 cases including 4 young patients and 4 adult patients the cerebral angiograms are pursued periodically during the course of 8 months to 3 years and a half. Two cases were autopsied.

2) There is little change in the vascular pattern in the adult and this is in contrast with the changes observed in young patients. Whether the "Moyamoya" in the adult is a different entity or remains stationary at a certain stage is not determined.

3) Periodical change on cerebral angiographic findings are markedly noted in young patients. This will deny the hypothesis of congenital origin of the vascular anomaly and supports our original conviction that the disease is an acquired one.

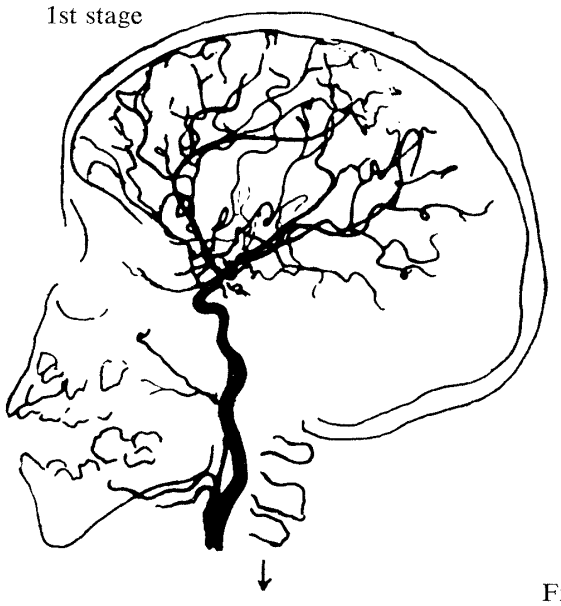
4) The disease begins with a stenosis at the terminal portion of the carotid artery or the original portion of the anterior and middle cerebral arteries. Though the lesion manifests itself unilaterally at time, eventual bilateral involvement is the usual course. According to the changing pattern of the vascularity observed in different periods, the following classification of the "Moyamoya" is made.

First stage of stenosis of carotid fork, 2nd stage of early rise of "Moyamoya" (dilatation of cerebral main arteries), 3rd stage of intensifying "Moyamoya" (fall of middle and anterior cerebral arteries), 4th stage of fibrillation of "Moyamoya" (fall of posterior cerebral artery), 5th stage of fall or reduction of "Moyamoya" (nonfilling of entire internal carotid artery system) and the final stage of 6th stage of vanish of "Moyamoya" (establishment of blood flow from external carotid artery). (Fig. 1, 2 and 3)

5) As the etiology of the stenosis or occlusion of the carotid fork, the vasospasm on this region, being provoked by some inflammation, trauma and soon the internal carotid artery will be presumed.

6) As the treatment for the disease, we tried surgically the perivascular

Fig. 1.



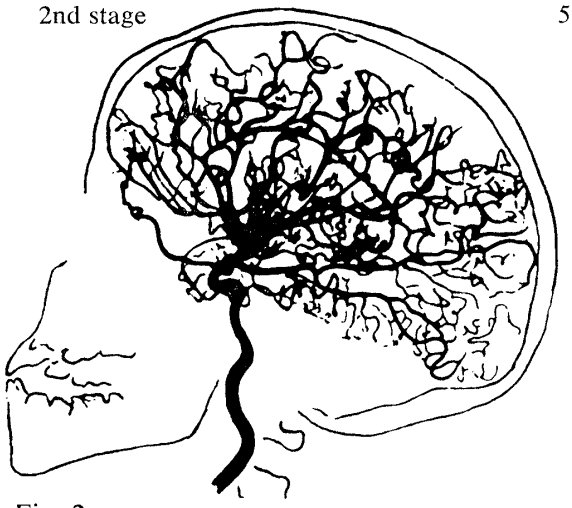
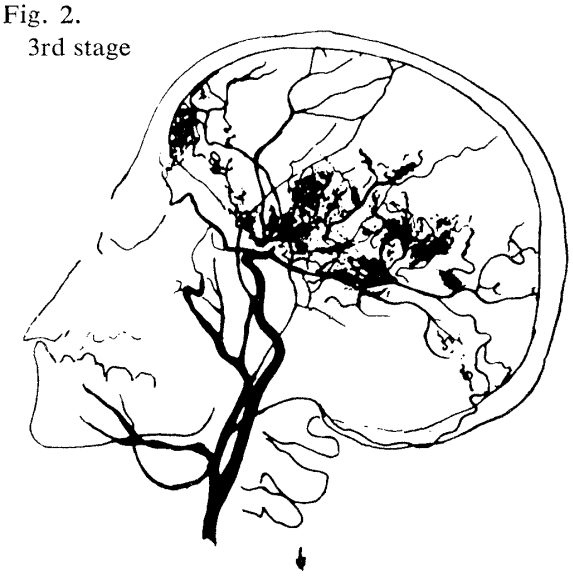


Fig. 2.



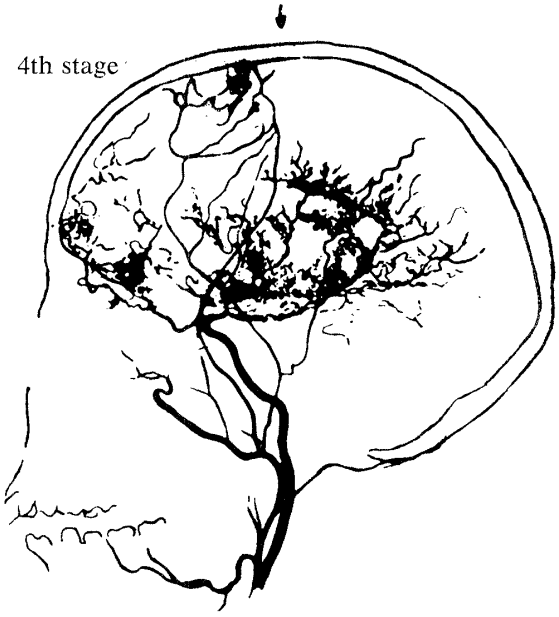
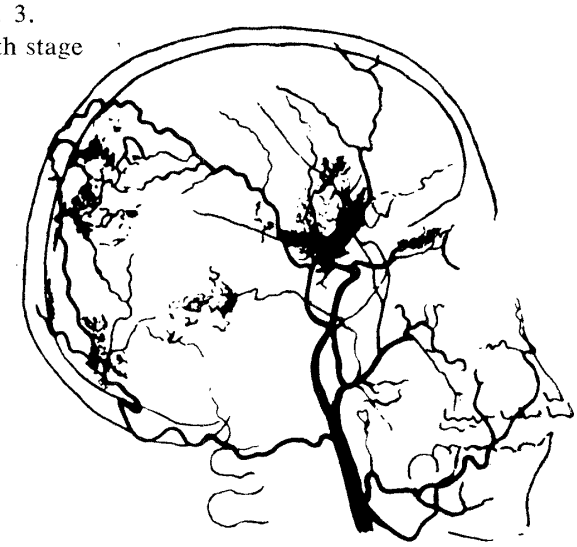
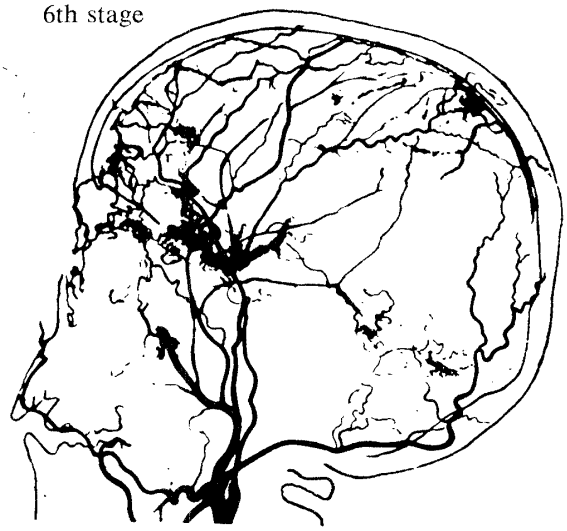


Fig. 3.





sympathectomy of the carotid artery in neck region, and the procedure for the dilatation of the diameter of internal carotid artery, sweeping away of some focuses of inflammation. Thereafter some vasodilators and steroids were prescribed.

Though the result of this treatment is not clear yet, but a case of 4 cases treated by this method showed some improvement on the carotid angiogram after one year and three months.

D-15. On the Net-like Vascular Anomaly of the Cerebral Artery

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Twelve cases of basal net-like vascular anomaly have experienced by us during these eight years. Seven of them are adult cases and another 5 are infants.

In 6 of 7 adult cases, Initial Signs were loss of consciousness of sudden onset due to subarachnoidal hemorrhage. But in a 33-year-old woman, with post-traumatic complaint after traffic accident was demonstrated by angiography the net-like arterial anomaly by chance.

In infants, the chief complaint was almost often transient hemiplegia and the signs often repeatedly observed in various grade.

The Angiographical finding:—The anterior and middle cerebral arteries bilaterally opacified as incomplete shape and distribution with the net-like vascular finding at the base of cerebrum.

The retrograde filling from the posterior cerebral artery via Rr. Splenii was remarkably recognized.

The connection of contrast medium could not visualized between the rostral trunk of the anterior cerebral artery and the peripheral part of the pericallosal artery opacified by retrograde filling.

In two of infant cases, net-like finding also revealed in the orbital region added to the base of cerebrum.

The Postmortem Examination was performed to a 55 year-old woman with net-like vascular finding. Anterior cerebral arteries and middle cerebral arteries