IV. 2 Measurement of CBF and CMRO$_2$ in Childhood Moyamoya Disease
-An Investigation into "Re-build up Phenomenon" by Positron CT-

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The appearance of abnormal EEG induced by hyperventilation is one of the
most specific finding in childhood Moyamoya disease. We have previously noted
that in childhood Moyamoya disease the EEG changes seen after hyperventilation
are not merely build-up which appeared during hyperventilation, or its
prolongation, but that subsequent to the disappearance or attenuation of build
up, which we have labelled "re-build up". And we have emphasized the fact that
EEG examination is an effective means for screening childhood Moyamoya
disease. ²) However, the nature of this re-build up phenomenon has been poorly
understood. In this paper, our preliminary experiences of positron emission
computed tomography in two cases of Moyamoya disease will be presented,
especially for the purpose of claryfying the relationship between the mechanism
of re-build up and brain functions.

Cases

Case 1 was a 13 year old male who suffered from paraventricular hemorrhage
on February, 1983. The angiographical stage ⁴) was V in the right side and III
in the left. Positron emission computed tomography was performed 7 months
later, at this point he had no neurological deficits.

Case 2 was a 17 year old male who had a frequent TIA attacks since he was 4
years old. Perivascular sympathetectomy and superior cervical ganglionectomy ⁵)
was performed on both side 7 years ago. Angiographical stage was VI on both
side.

Positron Emission Computed Tomography

Regional cerebral blood flow (CBF) and cerebral metabolic rate of oxygen
(CMRO$_2$) was measured by ECAT II ⁶) using cyclotron produced C$^{15}$O$_2$ and O$_2$
continuous inhalation technique.¹) The patients lay quietly on the ECAT bed
with eyes and ears open, essentially seeing and hearing the dark ambient
condition in the laboratory. Patients were asked to hyperventilate for 3
minutes, and 4 minutes scannings were performed before and after
hyperventilation. Since ECAT II is a single slice scanner, scanning was
performed at the same single plane parallel to orbito-meatal line which includes
basal ganglia. During the study, serial arterial blood samples were taken for
analyzing $^{15}$O radioactivity, PaO$_2$ and PaCO$_2$. Continuous EEG recording was also performed throughout the study.

Results

Pre-hyperventilation value revealed no differences between the two hemispheres in both cases, even though case 1 had different angiographical stages.

Post-hyperventilation 5 minutes scanning showed remarkable change in both CBF and CMRO$_2$. In case 1, decrease in CBF was observed mainly in the right hemisphere where the angiographical stage was more advanced, however CMRO$_2$ was decreased much more than CBF in the whole brain regions (Fig. 1). In case 2, where there was no difference in the angiographical stage between the hemispheres, marked decrease in both CBF and CMRO$_2$ was observed, and the decrease in CMRO$_2$ was also more than that of CBF (Fig. 2).

Simultaneous EEG recording revealed typical re-build up in both cases during the post-hyperventilation 5 minutes scanning. Also the sequential changes of PaO$_2$ and PaCO$_2$ showed characteristic findings in both cases. A gradual decrease in PaCO$_2$ due to hyperventilation was observed in the first place, however, which gradually recovered following the cessation of hyperventilation and returned to pre-hyperventilation level at the time of post-hyperventilation 5 minutes scanning. In contrast, high level of PaO$_2$ due to hyperventilation fell sharply following the termination of hyperventilation probably because of the decrease in the rate of respiration. And even though it gradually recovered, PaO$_2$ was still remained low level at the time of post-hyperventilation 5 minutes scanning (Fig. 3).

Discussion

Moyamoya, a Japanese word meaning 'something hazy like a puff of cigarette smoke drifting in the air' is the descriptive term we apply to a peculiar angiographic picture consisting of abnormal net-like vessels at the base of the brain, together with bilateral occlusion or stenosis of the internal carotid artery at the level of its terminal bifurcation, anterior and middle cerebral arteries. In 1963, we first suggest that this type of vascular abnormality consisted a new disease entity, since then, many cases have been reported and many studies have been performed from various aspects. One of the interesting feature of Moyamoya disease is the striking difference in the clinical presentation between children and adults. Children typically presents with recurrent episodes of sudden motor disturbances which are often induced by hyperventilation like crying or playing harmonica, while adults presents with evidence of intracranial hemorrhage. It is of great interest that polymorphous high voltage slow waves on EEG seen following the termination of hyperventilation, which we have labelled re-build up, deeply correlates with these kinds of attacks characteristically in children. However, the mechanism of re-build up has been poorly understood.
From the above findings, it is not likely that only the hemodynamic change following the hyperventilation is responsible for the emergence of re-build up on EEG, because there was a dissociation between the decrease in CBF and CMRO$_2$ in both cases.

Instead, on the bases of our results the mechanism of re-build up might be explained as follows; cerebral vessel constriction is produced due to the decrease in PaCO$_2$ following hyperventilation and it appeared as CBF decrease on positron emission computed tomography in the first place. And in addition to this decreased CBF, delayed decrease in PaO$_2$ following hyperventilation results in the decrease in CMRO$_2$ which causes cerebral dysfunction, and thus it appears as re-build up phenomenon on EEG. In other words re-build up is the manifestation of not only hypoxic but also ischemic state seen characteristically in Moyamoya disease.

Although further studies have to be performed before the definite conclusion is reached, the positron emission computed tomographic study seems to give us a important clue to elucidate this disease.

References

Acknowledgement

Collaborations of all the members of Cyclotron Radioisotope Center, Tohoku University were greatly appreciated.
Fig. 1. Case 1: 13 year old boy. Decreased CBF was observed mainly in the right hemisphere, where the angiographical stage was more advanced, at post-hyperventilation 5 minutes scanning. In contrast, decrease in CMRO₂ was much more than CBF in the whole brain regions.

Fig. 2. Case 2: 17 year old male. Marked and symmetrical decrease in both CBF and CMRO₂ was observed at post-hyperventilation 5 minutes, and they almost returned to pre-hyperventilation level at post-hyperventilation 15 minutes. Careful comparision between the pre- and post-hyperventilation 5 minutes scanning revealed that the decrease in CMRO₂ was more than that of CBF.
Fig. 3. Sequential changes of PaO$_2$ and PaCO$_2$ in Case 1. At the time of post-hyperventilation 5 minutes scanning PaCO$_2$ returned to normal range, however, PaO$_2$ still remains low level due to the decrease in the rate of respiration following hyperventilation.