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Satoshi Sanada* Nagako Murakami† Isaac Horiuchi‡
Eiji Oka** Shunsuke Ohtahara††

*Okayama University,

†Okayama University,

‡Okayama University,

**Okayama University,

††Okayama University,

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Satoshi Sanada, Nagako Murakami, Isaac Horiuchi, Eiji Oka, and Shunsuke Ohtahara

Abstract

Using a transcranial Doppler blood flowmeter, the blood flow velocity (BFV) ratio of the middle cerebral artery (MCA) to the basilar artery (BA) was investigated in 12 patients with severe motor and intellectual disability syndrome. The BFV of the MCA was also investigated in 58 handicapped children, classified according to the severity of their motor and intellectual disability. The ratio of the MCA to the BA was lower by 2 SD from the mean of our previously reported standard value in 8 out of the 12 cases with severe motor and intellectual disability syndrome, suggesting a more profound decrease in the level of brain activity in the MCA area than that of the BA area. The BFV of the MCA mainly decreased in cases belonging to the category of the most severe motor disability (bed-ridden). Hence, it is suggested that motor disability is the main factor related to the decrease in the BFV of the MCA.

KEYWORDS: transcranial blood flowmetry, blood flow velocity, handicapped children, severe motor and intellectual disability syndrome

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Brief Note

Cerebral Blood Flow Velocity in Handicapped Children

Satoshi SANADA*, Nagako MURAKAMI, Isaac HORIUCHI^a, Eiji OKA and Shunsuke OHTAHARA^b

Department of Child Neurology, Okayama University Medical School, Okayama 700, Japan

Using a transcranial Doppler blood flowmeter, the blood flow velocity (BFV) ratio of the middle cerebral artery (MCA) to the basilar artery (BA) was investigated in 12 patients with severe motor and intellectual disability syndrome. The BFV of the MCA was also investigated in 58 handicapped children, classified according to the severity of their motor and intellectual disability. The ratio of the MCA to the BA was lower by 2 SD from the mean of our previously reported standard value in 8 out of the 12 cases with severe motor and intellectual disability syndrome, suggesting a more profound decrease in the level of brain activity in the MCA area than that of the BA area. The BFV of the MCA mainly decreased in cases belonging to the category of the most severe motor disability (bed-ridden). Hence, it is suggested that motor disability is the main factor related to the decrease in the BFV of the MCA.

Key words: transcranial blood flowmetry, blood flow velocity, handicapped children, severe motor and intellectual disability syndrome

Transcranial Doppler is a noninvasive method that can provide information regarding blood flow velocity (BFV) in major cerebral arteries. Using this method, we have reported (1, 2) developmental changes in the BFVs of the middle cerebral artery (MCA) and the basilar artery (BA), which showed a maximum velocity at age 4 in the MCA, and at age 5 in the BA. It was also reported (2) that the BFV ratio of MCA to BA (MCA/BA ratio) was stable through infancy to adulthood. Therefore, this ratio is considered useful for estimating the distribution of intracranial hemodynamics, irrespective

of age.

Assuming that the MCA/BA ratio is useful as an indicator for evaluating brain dysfunction, this ratio was investigated in cases with severe motor and intellectual disability syndrome, and the results were compared with our previously reported standard reference data (2). This data was obtained from 72 healthy subjects, ranging in age from 0 to 30 years and divided into 9 age groups.

The factors which might affect the BFV of the MCA were also examined in relation to the manifestation and the severity of the handicap in these children.

Subjects and Methods

Patients

Severe motor and intellectual disability syndrome.

The MCA/BA ratio was investigated in 12 patients with severe motor and intellectual disability syndrome who visited our clinic from 1986 to 1996. Age and etiology of these patients are shown in Table 1.

Handicapped children. The BFV of the MCA was investigated in 58 handicapped children, who visited our clinic and the Asahigawa Jidoin Children's Hospital from 1986 to 1996. According to the level of motor and intellectual disability, these patients were subdivided into bed-ridden and intermediate categories for motor disability. Subdivisions of intellectual disability include IQ/DQ below 25, IQ/DQ between 25 and 50, IQ/DQ between 50 to 75, IQ/DQ between 75 to 85. The BFV of the MCA data was then compared with our previously reported standard reference data (1). Informed consent was obtained from all subjects and/or families.

* To whom correspondence should be addressed.
Present address: ^aAsahigawa Jidoin Children's Hospital,
^bDepartment of Health Science, Kibi International University.

Table 1 Age and etiology in 12 patients with severe motor and intellectual disability syndrome

Case	Age	Etiology
1	2 mos	Unknown
2	4 mos	Birth asphyxia
3	11 mos	Birth asphyxia
4	1 yrs 2 mos	Birth asphyxia
5	1 yrs 6 mos	Cerebral dysgenesis
6	1 yrs 6 mos	Mitochondrial disease
7	1 yrs 8 mos	Tay-Sachs disease
8	1 yrs 8 mos	Cerebral dysgenesis
9	2 yrs 2 mos	Anoxic encephalopathy
10	3 yrs 11 mos	Acute encephalopathy
11	8 yrs 0 mos	Canavan disease
12	11 yrs 11 mos	Cerebral dysgenesis

yrs: years; mos: months

was obtained from all subjects and/or families.

Methods

A transcranial Doppler flowmeter (EME TC 2-64, Eden Medizinische Elektronik GmbH, Germany) was used for the measurement of BFVs of the MCA and the BA. For the measurement of the BFV of the MCA, a 2-MHz probe was placed on the temporal area anterior to the auricle. Then, ultrasound was applied vertically to the skull, and the flow was recorded. For the detection of the BFV of the BA, a 2-MHz probe was placed between the second cervical vertebra and the occipital node. Then, ultrasound was applied to the BA, and the reverse flow was recorded.

The obtained data of the BFVs of the MCA and the BA, and the MCA/BA ratio below 2 SD from the mean of our previously reported standard value (1, 2) was assessed as abnormal.

Results

The MCA/BA ratio in severe motor and intellectual disability syndrome. The BFVs of the MCA and the BA and the MCA/BA ratio in the 12 patients with severe motor and intellectual disability syndrome are shown in Table 2. Asterisks in this table indicate abnormal data of below 2 SD from the mean of our previously reported standard value for the same age group. The BFV of the MCA was low in all cases, and that of the BA was low in 7 of the 12 cases. The MCA/BA ratio was low in 8 of the 12 cases, suggesting a more

Table 2 The BFV of MCA, the BFV of BA and the MCA/BA ratio in 12 patients with severe motor and intellectual disability syndrome

Case	BFV of MCA	BFV of BA	MCA/BA ratio
1	44* (65.1 ± 7.3)	36 (35.5 ± 2.9)	1.22* (1.60 ± 0.08)
2	42* (66.0 ± 6.3)	30* (41.9 ± 3.0)	1.40 (1.62 ± 0.11)
3	48* (79.2 ± 10.2)	30* (43.6 ± 3.0)	1.60 (1.64 ± 0.13)
4	46* (92.2 ± 18.0)	40 (48.8 ± 7.8)	1.15* (1.63 ± 0.11)
5	48* (92.2 ± 18.0)	42 (48.8 ± 7.8)	1.14* (1.63 ± 0.11)
6	50* (92.2 ± 18.0)	36 (48.8 ± 7.8)	1.39* (1.63 ± 0.11)
7	56* (92.2 ± 18.0)	32* (48.8 ± 7.8)	1.75 (1.63 ± 0.11)
8	42* (92.2 ± 18.0)	52 (48.8 ± 7.8)	0.81* (1.63 ± 0.11)
9	32* (92.2 ± 18.0)	28* (48.8 ± 7.8)	1.14* (1.63 ± 0.11)
10	32* (92.2 ± 18.0)	32* (48.8 ± 7.8)	1.00* (1.63 ± 0.11)
11	32* (84.1 ± 15.8)	40* (49.2 ± 3.1)	0.80* (1.57 ± 0.14)
12	46* (86.6 ± 18.0)	33* (44.0 ± 2.1)	1.39 (1.62 ± 0.17)

(mean ± SD)

*: Date with a decreased BFV or MCA/BA ratio below 2 SD from the mean of our previously reported standard value.

Abbreviations: BFV, blood flow velocity; MCA, the middle cerebral artery; BA, the basilar artery.

Table 3 The BFV of MCA in handicapped children

Intellectual disability IQ/DQ	Motor disability					
	Bed-ridden		Intermediate		Not disturbed	
	A	B	A	B	A	B
0-25	11	1	0	2	1	8
25-50	2	0	0	3	0	13
50-75			0	3	0	8
75-85			0	2	0	4

A: Patients with decreased BFV of the MCA below 2 SD from the mean of our previously reported standard value.

B: Patients with a normal BFV of the MCA.

Abbreviations: See Table 2.

profound decrease in the BFV of the MCA in the majority of cases with severe motor and intellectual disability syndrome.

The BFV of the MCA in handicapped children. In order to investigate the factors which may affect the BFV of the MCA, 58 handicapped children were subdivided into 12 categories and analyzed. The results are shown in Table 3.

In the category without motor disability, out of 34 cases, only one case, belonging to the IQ/DQ category below 25, was abnormal.

In the category indicating intermediate motor disability, all 10 cases showed no abnormalities in the BFV of

the MCA: two cases belonged to the IQ/DQ category below 25, 3 belonged to the IQ/DQ category between 25-50, 3 belonged to the IQ/DQ category between 50-75 and 2 belonged to the IQ/DQ category between 75-85.

In the category indicating the most severe motor disability (bed-ridden), 13 out of 14 patients showed abnormally low BFV value.

Discussion

Although the transcranial Doppler technique can not directly measure cerebral blood flow (CBF) volume, the BFV measured by this technique shows a close correlation (3) with the CBF volume measured by the ¹³³Xenon clearance method. Accordingly, the BFV measured by the transcranial Doppler technique can be considered to reflect the cerebral metabolism, which is proportional to the CBF. Moreover, the BFV measured by this technique can be considered to reflect the state of brain activity. There have already been a considerable number of studies (4-6) showing the increase in the BFV in response to various brain activations.

The MCA irrigates a wide cerebral area which includes the frontal, temporal and parietal lobes, and the BA irrigates the brain stem, cerebellum and occipital lobe. Therefore, the BFVs of the MCA and the BA can be considered to reflect the level of brain activity of the above mentioned areas, respectively. In the present study, both the BFVs of the MCA and the BA, and the MCA/BA ratio were low in the majority of cases with severe motor and intellectual disability syndrome. These results indicate an imbalance in intracranial hemodynamics, due to a more severe reduction in the level of brain activity in the MCA

area than that of the BA area, which is essential for life maintenance.

The present study on handicapped children revealed that the BFV of the MCA was reduced primarily in the category of the most severe motor disability (bed-ridden). Hence, it is suggested that motor disability is the main factor related to the decrease in the BFV of the MCA.

The MCA/BA ratio can be considered useful for estimating the distribution of intracranial hemodynamics, and a wide range of clinical applications can be expected in the field of child neurology.

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