

Comparison of memory quotient between non-cerebral hepatolenticular degeneration patients and normal people^{*}

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Abstract

BACKGROUND: Patients with cerebral type hepatolenticular degeneration (HLD) always suffer from decrease of intelligence and memory. If the memory ability of non-cerebral type HLD patients changes is still uncertain.

OBJECTIVE: To make sure the memory ability of non-cerebral type HLD patients.

DESIGN: Case control study on the basis of diagnosis.

SETTING and PARTICIPANTS: Thirty cases of non-cerebral HLD treated in Hospitalization and Outpatient Service in Neurology Research Institute of the Affiliated Hospital of Anhui College of Traditional Chinese Medicine during January 1998 to December 2002 were chosen as patient group with 15 males and 15 females, aged from 16 to 37 years old. Twenty healthy people from this hospital staff or volunteers from community were chosen as control group with 13 males and 7 females, aged from 18 to 43 years old.

INTERVENTIONS: Wechsler memory scale(WMS) was used to assess the memory quotient of the patient group and the control group.

MAIN OUTCOME MEASURES: Score of WMS of two groups and memory quotients.

RESULTS: The memory quotient of the patient group (134.07 ± 12.98) was higher than that of the control group (110.4 ± 13.4) ($P < 0.01$). And 67% of them had excellent memory, 20% had good memory, 13% of them had average memory or above.

CONCLUSION: The memory ability of non-cerebral HLD patient is pretty good. It is supposed that the copper content of them is a bit higher than that of normal people so that the efficiency of signal transmission among brain cells is increased due to the enhancement of activity of copper dependent enzyme.

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INTRODUCTION

Most of the Hepatolenticular degeneration(HLD) patients diagnosed by doctors are in middle or advanced stage of course with serious brain damages. The imaging test shows that there are emoliated focus in basal ganglia area, atrophy in brain stem and cerebral cortex. The learning and memory ability also are impaired except of occurrence of extrapyramidal system and hepatic symptoms, even mental symptoms and intelligence decrease can be found. All the above symptoms are defined cerebral HLD. The author conducted tests of memory quotient for non-cerebral HLD patients to explore the mechanism of intelligence change before and after occurrence of cerebral symptoms in non-cerebral HLD patients.

MATERIALS AND METHODS

Materials

Thirty cases with non-cerebral HLD treated in Neurology Research Institute of the Affiliated Hospital of Anhui College of Traditional Chinese Medicine were chosen as patient group. Among them, 29

cases were hospitalized and one case was outpatient. They all met the HLD diagnosis and typing standards^[1]. There were 15 males and 15 females who aged from 16 to 37 years old with a mean age of (23 ± 6) years old. The disease course ranged from 2 months to 16 years with a mean course of (4.88 ± 4.75) years. There were 4 cases with early stage symptoms, 24 cases of hepatic type, 2 cases with bone muscle type. Among them, 10 cases were good students with high score. The control group was consisted of 20 people came from healthy staff of this hospital and volunteers from community with 13 males and 7 females, aged from 18 to 43 years old with a mean age of (26 ± 6) years old. There was comparability on age, gender, education and living area between the two groups(See Table 1).

Table 1 Comparison on age, education and living area between the two groups (n)

Group	n	Age(year)			
		16-20	21-25	26-35	36-45
Patient	30	14	8	7	1
Control	20	2	7	8	3

Group	n	Education			Area		
		Elementary	Junior high school	Senior high school	University	Urban	Rural
Patient	30	2	11	12	5	25	5
Control	20	3	2	11	4	17	3

Methods

Wechsler memory scale(WMS)^[2,3] edited by Gong Yaoxian was used to assess 30 cases of non-cerebral HLD patients and 20 normal people in quiet room. The original scores of intelligence, figure recall, visual recognition, visual regeneration, associative memory, touch memory, understanding memory, and reciting numbers were got by urban or rural scale. The measurement score could be obtained by table look-up so that the total score could be calculated. Then the memory quotient could be obtained according to age table. The memory quotient was classified by WMS into following grades: excellent ≥ 130 , very good: 120-129; good: 110-119; fair: 90-109; below average: 80-89; critical state: 70-79; mental defect: ≤ 69 ^[4]. Physical examination, relative tests and ultrasound tests of liver and kidney were also provided to non cerebral HLD patients. Some patients took brainstem auditory evoked potential(BAEP), electromyogram(EMG), dynamic electroencephalography(DEEG) and head CT, MRI tests. Statistical analysis: SPSS 10.0 was used to process data and *t* test was used.

RESULTS

Among 30 cases with non-cerebral HLD, there were 20 cases with memory quotient ≥ 130 , 6 cases with 120-129, 4 cases with 110-119. Nerve system examination showed that there were 9 cases with positive pathologic reflex of lower limbs. Ultrasound B test showed that all 30 cases had hepatic disease of HLD, 5 cases had renal disease of HLD. DEEG test was conducted to 17 cases, 1 case showed slightly abnormal EEG with short blast of θ wave but without epileptic attack. Three cases had fast wave EEG, and 13 cases had normal EEG. Nineteen cases took BAEP tests, 7 of them showed normal results and 12 showed abnormal results with 1 case having disappeared single side or bilateral I wave and V wave, 1 case having decreased V wave amplitude, 6 cases having PL prolongation of single side or bilateral V

wave, 3 cases having PL prolongation of single side or bilateral III wave, 2 cases having IPL prolongation of single side or bilateral I-III wave, 5 cases with IPL prolongation of I-V wave, 3 cases of IPL prolongation of III-V wave. Thirteen patients took EMG test which had 7 normal cases and 6 abnormal cases. Among abnormal cases, 4 patients had slight slowered motor nerve conduction velocity of single side or bilateral common peroneal nerve, 1 case appeared prolonged distal latency of bilateral common peroneal nerves, 2 cases suffered from prolonged time limit and decreased amount of motor units of toe short extensor, gastrocnemius, triceps brachii and appeared short spike wave, multiphase potentials. Head CT scan was provided to 8 patients which all had no abnormal findings. Five patients took head MRI test, 4 were normal with only one appearing long T1 and T2 signals of both lenticular nucleus.

It could be seen from Table 2 that the scores of items liked picture recall, visual recognition, associative memory, tactile memory, understanding memory in patients group were higher than those of control group ($P < 0.01$ or 0.05) except for lower score on 1-100 intelligence scale. The memory quotient of patients was about 134.07 ± 12.98 which was much higher than that of control group (110.4 ± 13.4) ($P < 0.01$).

Table 2 Comparison of score of Wechsler memory scale and memory quotient between the two groups ($\bar{x} \pm s$)

Item	Control group (n=20)	Patient group (n=30)	t	P
1-100	12.0 ± 2.4	10.00 ± 2.50	-2.815	< 0.01
100-1	11.4 ± 2.3	12.10 ± 1.85	1.187	> 0.05
Accumulation	12.1 ± 1.7	12.57 ± 5.57	0.365	> 0.05
Figure recall	11.9 ± 2.3	13.23 ± 1.70	2.351	< 0.05
Visual recognition	12.2 ± 1.3	12.83 ± 0.59	2.318	< 0.05
Visual regeneration	11.9 ± 2.1	12.43 ± 1.04	1.130	> 0.05
Associative memory	11.4 ± 2.4	13.77 ± 0.97	4.895	< 0.01
Tactile memory	8.6 ± 2.7	14.43 ± 2.92	7.124	< 0.01
Understanding memory	11.1 ± 2.1	15.37 ± 1.13	9.399	< 0.01
Reciting number	12.7 ± 3.1	13.10 ± 2.72	0.484	> 0.05
Memory quotient	110.4 ± 13.4	134.07 ± 12.98	6.244	< 0.01

DISCUSSION

The long term clinical observation shows that most patients of non cerebral HLD or cerebral HLD before the brain symptoms come into being have normal living ability, working ability, learning and memory ability, some of them study very well. However, the intelligence of them is poorer than normal people with decreased study score after brain symptoms appear. The intelligence quotient and clinical symptoms and signs improve after normal treatment^[5-7]. This study shows that the IQ of non-cerebral HLD patients is much higher than that of normal people ($P < 0.01$) especially the memory ability is greatly increased. And 67% of them have excellent memory quotient, 20% with very good memory quotient, all are higher than that of normal distribution percentage of WMS^[4].

Tian *et al*^[8] used trisect radiate labyrinth box to test the old rats with supplement of CuCl_2 250 $\mu\text{g}/\text{kg}$ per day for 10 days and showed increased learning, memory ability. The copper content in hippocampus and blood serum of old rats was higher than that of control group. It suggested that learning and memory procedure could be improved by altering the brain functions through the protection of nerve cells as well as adjustment of intracerebral neurotransmitter by supplementing copper. Zhang *et al*^[9] has studied the interrelationship between epileptic attack, learning ability of rats, content of copper and zinc, it shows that the copper content in cortex of temporal lobe, hippocampus and hypothalamus decreased in the rats with frequent epileptic attacks which suggests that the effects of learning and memory caused by epilepsy might relate to the change of activity of enzymes dependent on copper and zinc. However, the study of Lin *et al*^[10] shows that long-term oral intake of aluminium will damage the memory of rats. Copper can affect the brain functions by adjusting the metabolism of monamine neurotransmitter through dopamine- β -hydroxylase,

5-hydroxytryptophan hydroxylase and monoamine oxidase. However, if there is too much or too little copper *in vivo*, it will cause cell apoptosis and lead to abalienation and intelligence decrease of patients^[11,8].

The 30 cases of non-cerebral HLD in this study showed no mental symptoms. But 9 cases appeared pathologic reflex of lower limbs. Tests and ultrasound examination showed all the patients suffered from the hepatic disease of HLD caused by copper deposit in liver, 5 cases had renal disease of HLD. 63% of them had abnormal BAEP, 54% with abnormal EMG, it indicated that copper deposit existed in brain tissue and all over the body although head CT and MRI showed no abnormal structural changes. It suggests that the brain tissue is in the stage of slight copper deposit without obvious morphologic changes caused by cell degeneration so that the memory ability may be possibly increased instead of being greatly decreased.

To conclude, in the early stage of HLD, when copper content in brain tissues is slightly increased, the activity of relative metalloenzyme might be enhanced so that the efficiency of signal transmission among brain cells is increased. However, further research need to be conducted for proving.

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非脑型肝豆状核变性患者与正常人记忆商数比较*

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摘要

背景: 脑型肝豆状核变性 (hepatolenticular degeneration, HLD) 患者的智力、记忆能力下降, 但非脑型 HLD 患者的记忆能力是否有改变?

目的: 明确非脑型 HLD 患者的记忆能力。

设计: 以诊断为依据的病例对照研究。

地点和对象: 1998-01/2002-12 到安徽中医学院神经病学研究所附属医院住院及门诊就诊非脑型 HLD 患者 30 例 (患者组), 男 15 例, 女 15 例, 年龄 16-37 岁, 20 例对照组为身体健康的本院工作人员及社会志愿者, 其中男 13 例, 女 7 例, 年龄 18-43 岁。

干预: 使用龚耀先修订的韦氏成人记忆量表 (WMS) 对患者组和对照组的记忆商数分别进行测试。

主要观察指标: 两组患者 WMS 测定量表得分和记忆商数。

结果: 患者组记忆商数 (134.07 ± 12.98) 高于对照组 (110.4 ± 13.4) ($P < 0.01$); 其中非常优秀者占 67%, 优秀者占 20%, 中上者占 13%。

结论: 非脑型 HLD 患者记忆能力较强, 推测其机制可能为脑组织的铜含量略高于正常人, 相关铜依赖酶的活性增强, 脑细胞间信号传递效能增强。

主题词: 肝豆状核变性; 韦氏智力量表; 记忆

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