*British Journal of Psychiatry* (1984), **144**, 538–541

# Ammon's Horn or Hippocampal Sclerosis without Epilepsy in Mental Handicap

P. E. SYLVESTER

**Summary: Neuropathological examination of the brain of eight adult mentally handicapped patients showed mesial temporal sclerosis. Three patients had cerebral palsy, one had Down's syndrome, three were retarded, and one had an unspecific mental handicap. Although there was a suspicion of fits in most patients, temporal lobe epilepsy was not diagnosed in life.**

Sclerosis of Ammon's horn is frequently demonstrated in epileptic patients. Margerison and Corsellis (1966) found damage in 55 per cent of their cases. Falconer (1974) reported an incidence of 80 per cent in the brains of chronic epileptics. Studying schizophrenic-like psychoses of epilepsy, Slater and colleagues (1963) reported that nearly 75 per cent had temporal lobe foci. Inherited conditions (Tay-Sachs disease), congenital abnormalities (micropolygyria), birth injury, anoxia, trauma, infections (herpes simplex and Coxsachie B5 encephalitis), neoplasia (primary and secondary and limbic encephalitis in carcinoma of the lung), degenerative disorders (vascular or senile) are some of the causes of hippocampal damage. Temporal lobe epilepsy is, partly on the evidence of an examination of temporal lobectomy specimens, often associated with changes in Ammon's horn, amygdala, and perhaps also the medial temporal gyri.

Forms of epilepsy other than grand mal and petit mal seizures have for a long time been recognised in the fields of child and general psychiatry. There is a wide range of clinical expression, some features being ictal, others inter-ictal. Epilepsy is also a considerable problem in the field of mental handicap. There are 358 patients (24.4 per cent) with either grand mal or petit mal seizures or combinations of both in a total population of 1,450 in St. Lawrence's Hospital, a hospital for mentally handicapped patients in the South of England.

Difficult behaviour problems—quarrelsomeness, aggressiveness, and destructive behaviour—afflict 101 of these. The problem often arises whether or not a patient with difficult or bizarre behaviour but without overt seizures suffers from occult epilepsy. An opportunity arose to report on eight adult mentally handicapped patients who had not been diagnosed as epileptic during life but who were found to have sclerosis of the Ammon's horn or hippocampus on post-mortem examination.

## Method

Eight mentally handicapped people came to post mortem. Routine neuropathological studies were made on their nervous systems. This consisted of preparation of routine blocks from five territories of the cerebral hemispheres—the temporal lobe with the Ammon's horn, three areas of brain stem, and one of cerebellum. The clinical details of the patients (six male and two female) were taken from the case notes.

## Case Histories

*Case 1:* Male aged 42 suffered from spastic paraplegia. Mental handicap was observed in infancy and attributed to the mother's abnormal health and a fall during pregnancy. As a child he was very baby-like. He failed to develop speech, was never able to stand and remained doubly incontinent. He developed attacks of screaming at six years. Clinically, epilepsy was queried but the diagnosis never confirmed. He was put on diazepam from the age of 39 years. There was no history of any infectious diseases.

The brain was symmetrical and the hemispheres weighed 560 g and 568 g The total weight was 1314 g. The left cerebral peduncle was smaller than the right. There was slight ventricular dilatation. The Ammon's horn on the left side was well formed, but contained a fine gliosis which ran through the pyramidal cell layer, slightly reduced in number: the right Ammon's horn was normal.

*Case 2:* Male aged 62 with right hemiplegia. Deficiency was observed in early infancy and attributed to illness at seven months when he cut his first tooth, had a fit and developed measles. He walked at two years, talked at two years and his habits became clean. He was the third of eight children. Later in childhood he sustained a broken leg and collar bone, and suffered numerous falls. A diagnosis of epilepsy was never made. He was said to be nervous, quiet and not troublesome. He developed 'bad habits' consistent

with adolescence at the age of 18 years and it was felt that he could not be trusted with his young sister. He was impulsive in adult life; otherwise moods and behaviour were normal.

His total brain weight was 1021 g but the left hemisphere weighed only 335 g, contrasting with the right of 530 g. The left peduncle and left pyramid were small. There was atrophy of the left frontal lobe and the left mammillary body. Gliosis was present in the dorsomedial nucleus of the thalamus. There was thinning of the genu of the corpus callosum. Intense gliosis was present throughout the left Ammon's horn, the right being normal. There was no evidence of ageing in the temporal lobe.

*Case 3:* Male aged 62 was mentally handicapped but also suffered from superimposed schizophrenia. He was a breech birth and said to be normal until seven years. There was no history of fits or convulsions. He had the usual childhood illnesses of measles, pertussis, mumps and scarlet fever. Schizophrenia was diagnosed because he kept muttering in a low voice, had numerous mannerisms and appeared hallucinated. He was doubly incontinent. He was illiterate and had no comprehensible speech. There were aggressive outbursts and he was described as solitary. He would bite or hit the nearest person to him for no apparent reason. He used to have outbursts of shouting and muttering to himself for hours on end. At the age of 60 years he had a grand mal seizure and had about seven up to the time of death. An EEG at the age of 60 showed no evidence of epileptic activity. He was treated with phenobarbitone. During the last two years of life he fell out of bed twice and fell on other occasions, bruising and sometimes cutting his forehead.

The brain weighed 1158 g and was symmetrical. There was a marked loss of cells and gliosis in the Sommer sector and end plate of the Ammon's horn on both sides. There was no evidence of senility in the sections examined.

*Case 4:* Male aged 68 suffered from Down's syndrome with typical trisomy. He had measles and pertussis as a child. There was some confusion about a diagnosis of epilepsy in childhood; this was made between the ages of nine and 13 years. Subsequently in life he had no fits. He was friendly and had a helpful disposition.

His brain was symmetrical and weighed 952 g. There were patchy areas of gliosis in the end plates and Sommer sectors on both sides and a mild gliosis in the white matter of the occipital and temporal lobes. Plaques and neurofibrillary tangles were present in the end plate and the pyramidal cell layer on both sides. Around the plaques were substantial rims of gliosis. These patchy gliotic areas were more discrete and therefore different from the more diffuse areas of cell loss and gliosis seen in the Sommer sector in other epileptics. There was a mild gliosis in the white matter of the occipital and temporal lobes.

*Case 5:* Male aged 72 had right hemiplegia and mental handicap, both of which were thought to have followed poliomyelitis in childhood. He had infantile convulsions followed by a period of epileptic fits. However, in later life he had no seizures but was sometimes quarrelsome. During adult life there were several episodes of falling in which he hit the front of his head above the left eyebrow causing lacerations which needed suturing. He did not receive anticonvulsant treatment.

His brain weighed only 856 g but appeared symmetrical. There was internal hydrocephalus and bilateral cortical atrophy in the frontal, parietal, occipital and temporal lobes. Temporal lobe sclerosis affected the end plates on both sides.

*Case 6:* A male aged 75 died from coronary occlusion but also had carcinoma of the prostate. Early history was not available. He was mildly mentally handicapped and able to read and write, but was untidy in dress. He talked to himself. At 28 years he dislocated his elbow in a fall. After reduction he fell again and redislocated the joint within ten minutes. He also received a cut over the right eye at the same age, but there was no record of the cause. On the whole he was a cheerful person and a good worker. At the age of 50 he developed a mannerism in which he did a jerky, shuffling stepdance when he started to walk, as though being hustled from behind. After six strides he resumed his normal gait. These mannerisms persisted throughout the rest of his life. Epilepsy was never diagnosed.

The brain weighed 1029 g and was symmetrical. It was well formed but showed universal marginal perigyral gliosis. There was cell loss in the Sommer sector on both sides. Evidence of ageing was not demonstrated.

*Case 7:* A female aged 55 suffered from mental illness in addition to mental handicap. She walked at the age of two years and talked at three years. She was clean in habits and used to be helpful at home washing up dishes and doing domestic duties, such as errands. In early adult life she deteriorated mentally and became solitary, muttering to herself. She was not subject to epilepsy.

The brain weighed 1297 g and was symmetrical. It was well formed. There was a fine gliosis in the pyramidal cell layer of Ammon's horn on the left side only, the right Ammon's horn being free from changes. Gliosis was increased round both inferior olivary nuclei. Ageing was not demonstrated in the temporal lobe.

*Case 8:* A female aged 65 was mentally handicapped

TABLE

*Summary of clinical and pathological features in the eight subjects*

| Case | Clinical diagnosis | Left | | Right | | Other comments |
|---|---|---|---|---|---|---|
| | | Sommer sector (SS) | End plate (EP) | Sommer sector (SS) | End plate (EP) | |
| 1 | Cerebral palsy | Mild gliosis | Mild gliosis | Normal | Normal | Minimal pyramidal cell loss. Gliosis most marked at outlet of EP |
| 2 | Cerebral palsy | Heavy gliosis | Heavy gliosis | Normal | Normal | Marked cell loss in gliosed areas. Molecular layers damaged |
| 3 | Schizophrenia | Heavy gliosis | Heavy gliosis | Heavy gliosis | Heavy gliosis | Marked cell loss in gliotic areas |
| 4 | Down's syndrome | Patchy areas of gliosis | Patchy areas of gliosis | Patchy areas of gliosis | Patchy areas of gliosis | Senile changes present. Cell loss in patchy gliosed ares |
| 5 | Cerebral palsy | Moderate gliosis | Moderate gliosis | Moderate gliosis | Moderate gliosis | Poor cell complement in both SS and EP |
| 6 | Mental handicap of unspecified cause | Heavy gliosis | Heavy gliosis | Heavy gliosis | Heavy gliosis | Reduced cell numbers in EP. Complete loss of cells in SS on both sides |
| 7 | Mental illness | Normal | Moderate gliosis | Not examined | | Mild degree of cell loss in EP |
| 8 | Mental illness | Normal | Heavy gliosis | Not examined | | Severe neuronal loss in gliotic area |

and mentally ill. She was the sixth of eight children, the others being normal. She had measles, chicken-pox and anaemia in childhood. She learned to read and tell the time but had difficulty in learning to wash and dress. At the age of 26 she was often fidgety. At 31 she grinned and giggled at nothing. By the age of 34 years she was said to be deteriorating and withdrawing into herself and suspected of developing mental illness. At 35 years, she was diagnosed as schizophrenic and catatonic. When 36 she continued to be withdrawn, inaccessible and had catatonic stupor and periods of excitement. Occasionally she was aggressive and noisy. Later, she became hallucinated, and, on one occassion, banged her right eye. She developed diabetes mellitus at 62. Records from the age of 55 years showed that she had received 60 mg. Phenobarbitone daily; the reason for this was not clear. She died of coronary infarction.

The brain, weighing 1196 g, was symmetrical. There was a generalised increase of perigyral gliosis but no evidence of ageing. Fine gliosis in the end plates on both sides was present.

## Results

The clinical diagnosis fell into the following categories: three male patients with cerebral palsy (cases 1, 2 and 5); three (one male and two female) with mental illness (cases 3, 7 and 8); one male patient with Down's syndrome (case 4); and one male patient with mental handicap of unspecified cause (case 6).

The constant pathological features in all eight cases was sclerosis of the hippocampus either in the Sommer sector (H.1) or the end folia (H.3) or both. These are summarised in the Table. Bilateral lesions were present in five patients. The left side only was affected in three.

Senile changes were present in the patient who had had Down's syndrome. They were not demonstrated in five other patients. Two patients, both cerebral-palsied males were not examined for this feature.

## Discussion

The purpose of this paper is to suggest that undiagnosed epilepsy in mental handicap is common.

The dramatic features of grand mal epilepsy and the less startling but nevertheless obtrusive signs of petit mal are the principal forms of epilepsy diagnosed in the mentally handicapped but less severe forms may go unrecognised. Cases 2, 3, 5 and 6 all had numerous falls, sometimes with unpleasant traumatic complications. If such people suffer cerebral palsy (cases 2 and 5) falls may be attributed to physical disability and clumsiness. The presence of Ammon's horn sclerosis in these cases suggests that these patients did have

epilepsy and may have benefitted from suitable anticonvulsant therapy. Cases 3, 7 and 8 had mental illness; it is more difficult to conclude that this should be attributed to epilepsy. It should be stressed that the case histories refer to patients who lived and were under medical surveillance before the introduction of modern drugs (new anticonvulsants, modern tranquillisers and antidepressants). Two of these patients were treated for many years with phenobarbitone without apparently influencing the nature of their illness, which would be expected if the mental illness was an epileptic phenomena due to Ammon's horn sclerosis. However, a case may be argued for trying modern anticonvulsant therapy as a diagnostic test in mentally ill patients, especially in such cases as 3, in whom the normal EEG was at variance with the pathological findings.

Corbett and Pond (1979) are cautious in acknowledging that paroxymal disturbances of behaviour (epileptic equivalents) have an epileptic basis in retarded children. Distinction has to be made between the clinical presentation in children compared with adults. Children have to grow, mature and learn, whereas adults have already done this and achieved experiences. Furthermore, Morel and Wildi (1956) found that Ammon's horn sclerosis was more frequent in older patients. Most accounts of epilepsy dwell on a history of seizures.

The extent of the pathological studies in these patients does not include notes on the amygdala, uncus or medial temporal gyri so that a complete pathological approach to temporal lobe epilepsy cannot be made. Temporal lobe epilepsy sometimes ends in generalised convulsions, but this is the exception rather than the rule. Communication difficulties in the mentally handicapped often make detailed history-taking impossible, so the observer is handicapped when it comes to discovering whether or not a patient experiences sensory hallucinations of smell or taste or delusions of the déjà-vu (dreamy state epilepsy) phenomena. These may be expressed as altered states of consciousness (cases 7 and 8) and disturbed motor activity (case 3), and be mistaken for a schizophrenic form of mental illness.

Ammon's horn sclerosis without epilepsy is rarer in the cerebral palsied than in the mentally ill, presumably because they have other cerebral pathology which can act epileptogenically. An analysis of pathological features amongst 28 cerebral palsied patients who had died over a thirteen-year period (1965–1977) at St Lawrence's Hospital revealed that six (21.4 per cent) did not have epilepsy and in five of them the Ammon's horn was normal. Three (10.7 per cent) did not have epilepsy, but had Ammon's horn sclerosis. There were 15 (53.3 per cent) epileptics with Ammon's horn sclerosis and four epileptics (14.3 per cent) without it. If these figures could be extrapolated to the living, then about 10–11 per cent of cerebral palsied people might be expected to have Ammon's horn sclerosis without overt epilepsy.

It is important to appreciate that while some handicapped adults have serious brain damage, others—approximately one third (Sylvester 1961)—do not and therefore the dogma that as a group their pathology is so complex as to make proper evaluation extremely difficult is not always valid. The fact that there is a possibility for some patients to go through lifetime with undiagnosed epilepsy is stimulation enough to investigate patients further.

**Acknowledgements**

I wish to thank Mr D. F. High and Mrs M. Shaw for technical help.

**References**

CORBETT, J. A. & POND, D. A. (1979) Epilepsy and behaviour disorders in the mentally handicapped. In *Psychiatric Illness and Mental Handicap* (eds F. E. James and R. P. Snaith). Gaskell, The Royal College of Psychiatrists. Ashford, Kent: Headley Brothers.

FALCONER, M. A. (1974) Mesial temporal (Ammon's horn) sclerosis as a common cause of epilepsy. *Lancet*, **ii**, 767–70.

MARGERISON, J. H. & CORSELLIS, J. A. N. (1966) Epilepsy and the temporal lobes. *Brain*, **89**, 499–530.

MOREL, F. & WILDI, E. (1956) Sclerosis ámmonionne et epilepsies. *Acta Neurologica Belgica*, **56**, 61–74.

SLATER, E., BEARD, A. W. & CLITHEROE, E. (1963) Schizophrenia-like psychoses of epilepsy. *British Journal of Psychiatry*, **109**, 95–150.

SYLVESTER, P. E. (1961) Brain weight in mental deficiency *Journal of Mental Deficiency Research*, **5**, 98–107.

P. E. Sylvester, F.R.C.Psych., F.R.C.Path., D.C.H., *Consultant Psychiatrist, St Lawrence Hospital, Caterham, Surrey*

*(Received 8 July; revised 9 September 1983)*