

# PATIENTS WITH HYDROCEPHALUS PRESENTING IN THE PSYCHIATRIC CLINIC

REPORT OF TWO CASES

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The purpose of this communication is to call attention to some unusual cases of dementia caused by hydrocephalus and potentially amenable to treatment. Hakim and Adams<sup>6</sup> have described a syndrome of normal pressure hydrocephalus with free communication between the ventricles and basilar cisterns. The two cases presented here had normal pressure of the ventricular fluid but they were of the non-communicating type. They were caused by an obstructive lesion at the level of the aqueduct of Sylvius. Their symptoms differed in several aspects not only from those reported by Hakim and Adams but also from other cases of "occult hydrocephalus" reported in the literature. Some of the clinical aspects shall be discussed.

CASE 1 — A 60-year-old housewife was referred to the Psychiatric Clinic of the University of Michigan Medical Center for evaluation of personality changes and blackout spells. Over the last four years she had developed a feeling of tiredness and lost interest in her household. Lately she had difficulty in getting around. Her memory became impaired especially for recent events. Also, for four years she had occasional generalized convulsions and at times brief episodes during which she would look pale and stare and not understand what was said. Evaluations over the last three years included electroencephalograms which were abnormal and x-ray studies of the skull suggestive of increased intracranial pressure but a brain scan was normal and she was considered to have a depressive or schizophrenic reaction and was treated with electroshocks and medications. Past history revealed that at age 15 she had an acute febrile illness and stayed in bed for two weeks, after which her mind was "not right" for some time and she was kept out of school for nine months. Family history indicated that of her six siblings, three died of carcinoma of the spine or of the throat. Her husband had a long history of drinking and her only son was serving in the armed forces overseas for the last year.

During the interview the patient was mildly depressed. She misinterpreted or misunderstood some questions and tended to get lost in details. She admitted to ruminations and was aware that something was not right with her. An EEG was

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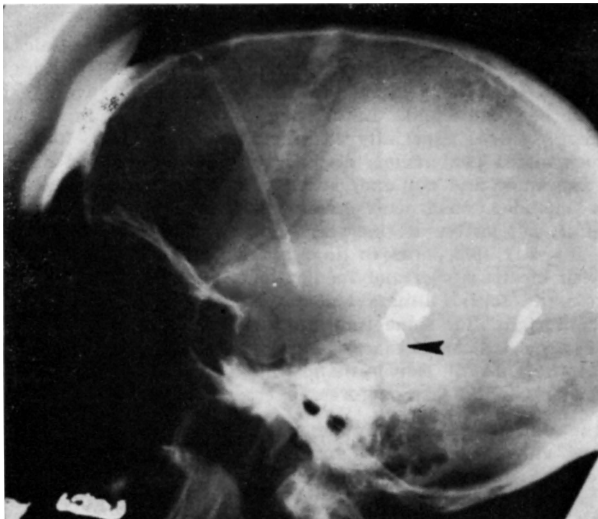
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moderately abnormal, abortively paroxysmal with strong emphasis in the left temporal region. Psychometric tests revealed a full scale I.Q. of 98. There was evidence of visual-motor difficulties and of impaired abstract reasoning. On examination by the neurologist several minor abnormalities were found. Convergence of gaze was poor. Jaw jerk was brisk. Face was asymmetrical with the left corner of the mouth slightly depressed and flattened. Tongue protruded to the left and moved slowly. Rapid alternating movements were performed rather poorly with both hands. With passive movements there was sometimes an irregular resistance in the upper extremities. Deep tendon reflexes were somewhat brisk but symmetrical in the upper extremities. The left prepatellar reflex was elicited more easily than the right. Both ankle jerks were absent. Bilateral grasp reflexes were present.

She was hospitalized for further studies. X-rays of the skull showed demineralization of the dorsum sellae and evidence of enlarged sella turcica. A tomogram confirmed intra and extrasellar type erosion. A left carotid angiogram demonstrated marked ventricular dilatation. A pneumoencephalogram failed to fill the proximal portion of the Sylvian aqueduct and the ventricles above. A Pantopaque ventriculogram showed obstruction at the mid-rostral portion of the Sylvian aqueduct with resultant dilatation of the lateral and third ventricles (figure 1). Opening ventricular pressure was 120 mm of water. A right ventriculo-atrial shunt was performed and the patient had an uneventful post-operative course.

When seen two and a half years after her operation, the patient felt she was back to what she had been before the onset of her illness. Neurological examination still revealed some minor abnormalities, but an EEG was within normal limits.



*Fig. 1 — Case 1. Pantopaque ventriculogram showing obstruction of mid-rostral portion of Sylvian aqueduct.*

*Discussion* — This patient resembles some cases of “occult hydrocephalus” reported by McHugh<sup>8</sup>. Those becoming symptomatic in adult life presented with: (1) a stiff, unsteady gait appearing insidiously in middle life; (2) listlessness, apathy, inattentiveness followed by decline of memory, of abstract thinking, and of non-verbal aptitude; (3) loss of drive and general

slowness; (4) brief attacks of loss of consciousness often preceded or followed by headaches. These were more common than typical epileptic seizures. In none of the cases was there mention of papilledema. The clinical manifestations appeared to depend rather on the character of change in the ventricular pressure than on the location of the obstruction.

The present case differs by the predominance of mental symptoms during a rather lengthy course without a distinct gait disturbance and without the attacks of unconsciousness mentioned by McHugh. The mental picture led to the consideration of a presenile dementing process and the epileptogenic EEG focus to the suspicion of a brain tumor before further studies established the definite diagnosis.

CASE 2 — A sixteen-year-old boy was admitted to Ypsilanti State Hospital with a one-and-a-half-year history of withdrawal, declining school performance, continuous headshaking and, more recently, occasional aggressive behavior, nocturnal enuresis, and inappropriate sexual behavior. Before being transferred he had been hospitalized elsewhere on two occasions. The diagnosis had been Schizophrenic Reaction and during his first admission he seemed to benefit from phenothiazines but, because of some aggressive behavior, he could not be kept in school. He acted hostile and sometimes talked in a strange way at home and when he was re-hospitalized he made occasional sexual advances toward female staff, and demonstrated other sexual disinhibitions. An EEG a few months before admission to Ypsilanti State Hospital was moderately abnormal and paroxysmal and focal to the right temporoparietal region. He was placed on Dilantin and Phenobarbital.

Past medical history was noncontributory. Social history revealed conflicts at home because of his father's and later his step-father's drinking. On examination he was found to be a short, stocky boy, appearing younger than his stated age. The only positive findings on physical examination were a slightly ataxic gait and continuous side-to-side shaking of his head. During psychiatric examination he was cooperative. Speech was spontaneous and coherent and his range of affect fairly broad. He could smile appropriately and make jokes. Thought content was not overtly disturbed. Response to proverbs was concrete. Psychiatric diagnosis was acute schizophrenic episode. Routine laboratory studies were normal except for white blood count of 21,500 cells per cubic millimeter with neutrophilia.

He was referred to the medical clinic where he was found to have bilateral lymphadenopathy. Inguinal node biopsy revealed Hodgkin's disease. He was transferred to the University of Michigan Medical Center for evaluation and treatment. Because of his head shaking he was evaluated by a neurologist. He talked little and reluctantly. He could arrest his head shaking briefly, at request. No new findings were elicited. An EEG was again focally abnormal but there was also slowing of the background. Cerebrospinal fluid examination revealed a protein of 63 mgm.% and a colloidal gold curve of 5553210000; glucose and cell count were normal. Skull films were normal. A right carotid arteriogram showed evidence of dilated lateral ventricles without midline shift. An air ventriculogram showed symmetrically dilated lateral ventricles, third ventricle and proximal portion of the aqueduct which gradually tapered off. There was no air in the fourth ventricle. Pneumoencephalogram showed air in the fourth ventricle without gross deformity or displacement. The ventricular pressure was between 50 and 70 millimeters of water. It was decided not to perform a shunting operation.

After return to Ypsilanti State Hospital he often complained of being hungry and he craved for food and cigarettes. The occupational therapist reported poor coordination, short attention span, and poor work quality though the patient took

pride in his products. A Vineland social maturity test revealed a level of 8 years, 6 months, for an actual age of 16 years and 7 1/2 months. He was released to return to school but he soon again got into fights. His head shaking became worse and he was re-hospitalized. The last nine months of his life he deteriorated mentally and physically. He wandered around and seemed confused. He would say a word or two if he needed something badly enough. Eventually he became lethargic and weak and he died three years after onset of his first symptoms.

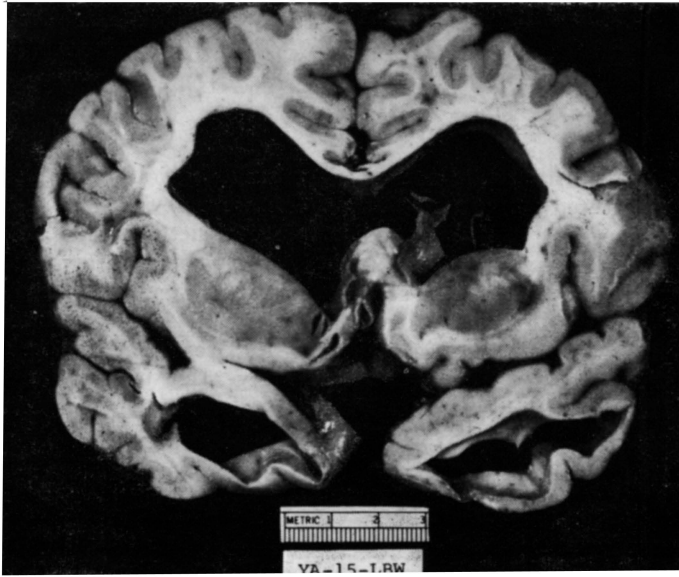
Post-mortem examination revealed massive enlargement of mesenteric, pelvic and retroperitoneal lymph nodes and implants of Hodgkin's sarcoma in various organs including liver, left kidney and left pleura but none in the brain or meninges. The brain weighed 1,350 grams. At the base, both temporal lobes were bulbous and the cortex was markedly thinned, especially over the temporal poles due to anterior extension and dilatation of the temporal horns of the lateral ventricles (figure 2). Both lateral ventricles and third ventricle were greatly dilated with marked thinning of the floor of the third ventricle. A thin membranous septum completely blocked the caudal end of the aqueduct; the aqueduct proximal to the membrane was greatly dilated (figure 3). The septum pellucidum was fenestrated and the fornices thinned and flattened against the thalamus and roof of the third ventricle. The septal area and diagonal band of Broca were partly cystic and gliotic. The choroid plexus was atrophic. Microscopically, the membrane blocking the aqueduct was composed of astrocytes and glial tissue devoid of ependyma on its surfaces. There was bilateral atrophy of the fimbrias of the hippocampi; the fornices were devoid of myelin and showed gliosis.

*Discussion* — This second patient seems to fall between some of the cases reported by McHugh<sup>8</sup> and the juvenile type of hydrocephalus as described by Pennybacker<sup>10</sup> but disturbance of gait, clumsiness or large head were not observed. The dementia was more pronounced than in the cases described by McHugh and it was progressive. In addition there were several striking features not usually observed in cases of occult hydrocephalus and which for their very unusualness led to the diagnosis of a functional psychosis. They were: (1) occasional aggressive and sexually disinhibited behavior; (2) incomplete mutism; (3) to and fro shaking of the head.

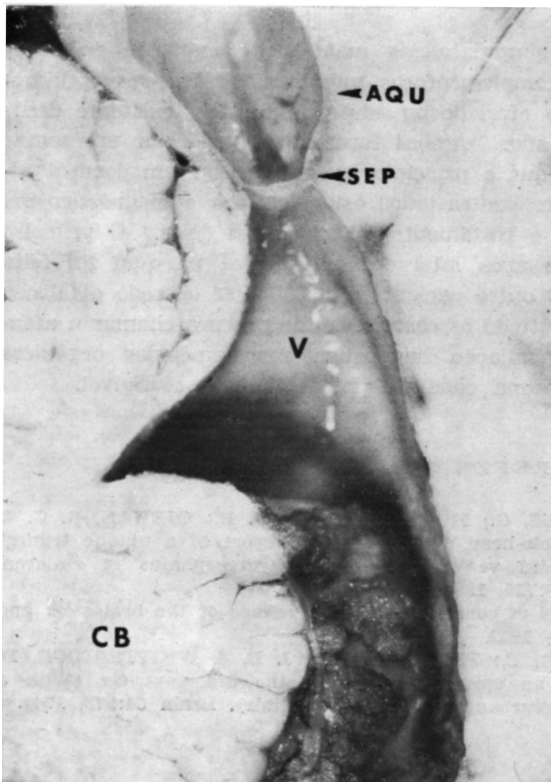
The aggressive and sexually disinhibited behavior might be related to disturbance of the postero-medial nuclei of the hypothalamus or their connections<sup>5</sup>. Such hypothalamic symptoms have been noticed in tumors adjacent to the aqueduct and causing increased cerebrospinal fluid pressure in the third ventricle<sup>7</sup>.

Mutism due to an organic lesion has usually been described in association with akinesia<sup>2,4</sup>. Cairns and associates<sup>3</sup>, however, have observed an incomplete and reversible mutism after ventriculogram in chronic hydrocephalus due to stricture of the aqueduct and other lesions of the midbrain.

The nearly continuous headshaking represented the most intriguing symptom. Recently Benton and associates<sup>1</sup>, and subsequently, Nellhaus<sup>9</sup> reported on a movement disorder associated with third ventricle cyst and hydrocephalus in children and which they named "The Bobble-Head Doll Syndrome". Of the three cases, one of Nellhaus' is closest to ours. The headshaking was abolished after placement of a Holter valve.



*Fig. 2 — Case 2 — Coronal section of brain at level of anterior limb of internal capsule, showing anterior distension and marked dilatation of temporal horns, and dilated superior horns of lateral ventricles. The cerebral mantle at this level is reduced to a thickness of 15 to 25 mm. dorsally and dorso-medially.*



*Fig. 3 — Case 2 — Sagittal section through Sylvian aqueduct (AQU), fourth ventricle (V) and cerebellum (CB). Septum (SEP) blocks caudal end of aqueduct which is dilated above*

## SUMMARY

Two patients with major mental symptomatology and few neurological signs were presented who on further studies were revealed to have unsuspected hydrocephalus. This was due to an obstructive lesion at the level of the aqueduct present since birth or for many years. Both presented with a rather unusual clinical picture so that they were first considered to suffer from a psychiatric illness. Radiographic contrast studies established the diagnosis in both cases and led to successful treatment in one of them. The first patient made an uneventful recovery after a shunting operation. The other had no such operation and died of an unrelated disease some time later.

A constellation of declining mental functions with or without behavioral disturbance and few or minor neurologic signs and an abnormal electroencephalogram should include in the differential diagnosis "occult" hydrocephalus which is a potentially correctable condition. The psychiatrist might be the first one to see this type of patient and to initiate the appropriate evaluation.

## RESUMO

*Pacientes com hidrocéfalo e predominância de distúrbios psíquicos.  
Relato de dois casos.*

Em dois pacientes com sintomatologia mental predominante e poucos sinais neurológicos, exames complementares mostraram tratar-se de hidrocéfalo por lesão obstrutiva ao nível do aqueduto de Sylvius, existente desde o nascimento ou por muitos anos. Ambos foram internados por apresentar um quadro clínico não usual que a princípio levou a pensar em doença psiquiátrica. Exames radiológicos contrastados estabeleceram o diagnóstico em ambos os casos e permitiram o tratamento eficaz de um deles. O primeiro paciente teve grande melhora após intervenção cirúrgica na qual foi feita derivação ventrículo-atrial. O outro paciente não pôde ser operado e faleceu de afecção intercorrente. Discutindo os casos, o autor procura chamar a atenção para a importância da avaliação diagnóstica das demências orgânicas visando ao despistamento de uma condição potencialmente corrigível.

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