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A Treatable Form of Dementia Due to Normal-Pressure, Communicating Hydrocephalus

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ANALYSIS of cases of apparently hopeless, progressive dementia has very recently revealed a significant, although small number with unsuspected, communicating hydrocephalus and a normal cerebrospinal fluid pressure. It has been found that this condition may be remedied by surgical shunting procedures or even by repeated lumbar punctures. This clinical observation by Foltz and Ward[1] in 1956 and more recently by Hakim and Adams *et al.*[2, 3] is important and significant because it represents a treatable form of progressive dementia. The essential radiological finding (pneumoencephalography) has been ventricular dilatation with air present in the basal cisterns, occasionally over the insular cortex and the midline sulci, but absent over the convexity of the hemispheres.

These cases likely have been undetected until the last few years, because of an erroneous tendency to ascribe most cases of dementia to "hardening of the arteries". Without evidence of repeated small or of massive strokes, or of major arterial obliterative vascular disease or serious prolonged hypertension (with a resultant possibility of marked small vessel disease), it is essential that one regard the demented patient as a problem in *diagnosis*—not a problem in *disposal.*

This paper describes 14 patients manifesting the clinical triad of dementia, gait disturbance and urinary incontinence associated with normal spinal fluid pressures and communicating hydrocephalus. Of the 14, one patient was seen in 1959, another in 1964 and the remainder during 1965-66 inclusive.

Despite the fact that all had "symptomatic normal-pressure, communicating hydrocephalus" and that the symptoms were fairly constant, in some cases there were different clinical signs and different etiological features.

Cases 1 to 5 illustrate progressive dementia in five middle-aged and older persons associated with gait disturbance in four and incontinence in three.

CASE 1.—F.K., a 61-year-old tailor, became aware of a decline in his intellect one year before his neurological evaluation. This progressed, and six months before admission his gait deteriorated; he took small steps and fell frequently. He also developed bladder and later bowel incontinence.

Examination in July 1965 revealed a demented man, disorientated as to time but not as to place. Recent and remote memory were impaired, as was abstract thinking. The only other abnormalities were mild incoordination of heel-shin testing and a severe gait apraxia.

A pneumoencephalogram in July 1965 confirmed a communicating hydrocephalus. Following this, his condition deteriorated for about 48 hours. A lumbo-peritoneal shunt was then performed (July 1965), and after an uneventful convalescence he returned to full work. In February 1966, however, his gait deteriorated rapidly and he began to fall, his memory failed, and he developed intermittent urinary incontinence. It was felt that the shunt was

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blocked. On April 12, a ventriculoatrial shunt was performed. When last seen, he was depressed because of a tense family situation and because his memory remained moderately impaired for both recent and remote events. Although able to walk unaided, he shuffled. He was working as a tailor, but was not as productive as he had previously been.

CASE 2.—A.R., a 68-year-old war amputee, enjoyed good health. About one year before admission he developed memory deterioration; he became forgetful, particularly of recent events, and had difficulty in writing letters and playing cards. One month before admission he developed difficulty in walking and shaving, and unwitting urinary incontinence.

On physical examination he was cheerful but completely disorientated as to time and place. Recent memory was severely impaired, and he was unable to calculate. Upward gaze was limited and he had brisk jaw and snout reflexes. The left plantar response was extensor. (He had had an above-knee amputation on the right.)

A pneumoencephalogram confirmed a communicating hydrocephalus, and following this he became akinetic and mute, with increased urinary incontinence. The deterioration continued until after repeated lumbar punctures, with removal of large quantities of cerebral spinal fluid. He then improved markedly; his incontinence disappeared and he showed improved mentation. On March 20, 1966, a lumboperitoneal shunt was performed and this was followed by continued improvement in his condition. He rapidly became orientated and gradually regained normal walking.

Two months after discharge, he again became demented and incontinent over a period of five days. It was thought that the shunt had occluded; when this was remedied, he again began to show improvement. However, he is neither as bright mentally nor walks as well as in the weeks immediately following his lumboperitoneal shunt.

CASE 3.—E.L., a 46-year-old woman, had a two-year history of apathy and failing memory, a nine-month history of progressive limb tremor and a one-month history of unsteady gait and incontinence.

Physical examination revealed severe dementia, and bilateral upper and lower limb tremor with rigidity and gait impairment. An electroencephalogram was grossly abnormal. An air encephalogram revealed communicating hydrocephalus.

On December 1, 1965, a right ventriculoatrial shunt was performed. Improvement began immediately and continued until mid-January 1966, when deterioration became evident and she became bedridden. Examination revealed severe dementia, lack of spontaneous movement, generalized increase in muscle tone, right-sided hyperreflexia and a right extensor plantar response. She also had brisk jaw, facies and snout reflexes.

Over the next three weeks the shunt was revised on three occasions. On March 12 a large left subdural hematoma was removed. Her postoperative course was stormy and complicated by a septicemia and progressive neurological deterioration. However, with appropriate antibiotics she gradually improved and when last seen she was walking unaided, although cautiously. She had pseudobulbar laughter and was fully continent but continued to have moderate memory impairment, particularly for recent events.

CASE 4.—B.R., a 62-year-old hotel executive, noted memory deterioration six months before admission. His judgment and unpredictable behaviour led his superiors to direct him to seek medical help. One week before admission he noted an unsteadiness of gait and fell once. He also was incontinent on one occasion.

Physical examination revealed only a slightly ataxic gait, impairment of rapid alternating movements bilaterally and mild memory impairment.

Because of the nature of his symptoms, arteriography was performed which tended to exclude disease of large vessels as a basis for his symptoms. A pneumoencephalogram showed air in the insular region. A second air encephalogram was done with the neck fully hyperextended in an attempt to fill the sulci. Despite this posturing no air was seen over the hemispheres.

On November 24 a lumboperitoneal shunt was carried out. His gait impairment cleared. He was able to return to a demanding executive job, but his judgment remained mildly impaired.

CASE 5.—G.H., a 52-year-old man, had an eight-month history of lack of initiative, increasing somnolence and fatigue. Physical examination revealed marked memory impairment with impairment of attention and perception. The only other abnormality was a left extensor response.

A pneumoencephalogram revealed communicating hydrocephalus, but the sulci filled over the medial aspect of the hemispheres and in the insular region. The air encephalogram resulted in marked mental confusion and increased mental impairment. A lumboperitoneal shunt has resulted in no change in his condition.

Of these five, four improved with surgery and one remained unchanged. Of the four who improved, two had setbacks because of occluded shunts, but improved again with revision of the shunts. One of the four had additional complicating features, a subdural hematoma and septicemia but finally emerged from this and was able to return home, although to date she has not been able to resume her household duties. It is of note that the one who failed to improve had air in the sulci over the insula and medial parts of the hemispheres.

An example of symptomatic hydrocephalus with an initial normal cerebrospinal fluid (CSF) pressure that changed to an increased CSF pressure, as a sequel to further leakage of an aneurysm, is seen in another patient (Case 6).

CASE 6.—H.A., a 58-year-old man, suffered a head injury in 1954 necessitating a right frontal craniectomy. Apart from a slight personality change and a tendency to be quarrelsome, he was neurologically normal and able to resume his previous job. In May 1964 he became forgetful and more irritable; within five months he was sent home from work because of forgetfulness. Initial examination revealed only an apathetic but irritable man who was moderately demented. The cerebrospinal fluid pressure was 160 to 180 mm. water and contained 16 to 80 lymphocytes and 134 to 148 red cells with a protein content of 62 to 100 mg. %. A pneumoencephalogram revealed communicating hydrocephalus with insular air. He was discharged unchanged.

He continued to deteriorate over the next three months, with many fluctuations in his behaviour. Again complete examination was unremarkable apart from the dementia. Again the cerebrospinal fluid was under normal pressure (160 mm. water) but contained 177 red cells with a protein of 62 mg. %. He became less responsive and developed a gait ataxia, with minimal right-sided pyramidal signs. The cerebrospinal fluid was now under increased pressure (300 to 350 mm. water) and contained 4000 red cells and 16 white cells. A retrograde brachial arteriogram demonstrated a large aneurysm arising from the basilar artery at the bifurcation of the posterior cerebral artery. He became stuporous and incontinent. A right ventriculoatrial shunt using a Holter valve was performed February 17, 1965.

He showed progressive improvement and by October was able to return to an inside job. His gait was normal, and mentally he had almost returned to his previous state. By December 1965 another subarachnoid hemorrhage occurred and he had again deteriorated. After bed rest, improvement once again began and he has been able to return to work but in a very menial capacity, because of mental impairment.

Four other patients (Cases 7, 8, 9 and 10) exemplify communicating hydrocephalus secondary to subarachnoid hemorrhage. One patient (Case 7) improved with a shunting procedure, while another (Case 8) improved after repeated lumbar punctures; the third (Case 9) showed some improvement in his mental state, but the shunt occluded and although it could not be revised, a lumboperitoneal shunt has been performed in the past month. He remains incontinent, impaired mentally, and is unable to move his lower limbs because of spasticity.

CASE 7.—In November 1963, B.M., a 50-year-old hypertensive woman, had a left middle cerebral aneurysm clipped following a subarachnoid hemorrhage. She recovered uneventfully apart from right-sided seizures which were easily controlled on drugs. When discharged from hospital she had a mild dysphasia, and the family noted subtle mental changes.

Within three months she complained of headaches, increasing fatigability, excessive drowsiness and poor memory. Examination revealed a moderate dementia; her gait was slow and cautious, and she failed to swing her arms when walking. She had residual right-sided hyperreflexia. A lumbar puncture resulted in marked improvement. She remained well for two months and then began to have periods of irrational behaviour. Intellectual deterioration again became evident. A pneumoencephalogram one year after the hemorrhage showed communicating hydrocephalus, and this procedure resulted in marked improvement. Following discharge the symptoms were those of confusion, apathy, headaches, sleep disturbance and poor memory. These improved with lumbar punctures, which were continued until January 1965. Intellectually she has shown marked improvement; her gait is normal, and she has had no further periods of irrational behaviour. She has a chronic mild depression, however, cannot cope with housework, and rarely goes out of doors alone.

CASE 8.—L.W., a 32-year-old known hypertensive man, suffered a subarachnoid hemorrhage on October 12, 1965. He had a stormy hospital course complicated by phlebitis, possible pulmonary embolism and a urinary tract infection. Examination initially had been without lateralizing signs although the fundi showed hypertensive changes without papilledema. The blood pressure was 230/130 mm. Hg. Over five months he showed improvement in his level of consciousness, but he was demented and incontinent, and his limbs were severely spastic. The cerebrospinal fluid, which had been initially under increased pressure, was now under normal pressure and an air encephalograph showed communicating hydrocephalus. On April 6, 1966, a ventriculoatrial shunt using a Pudenz valve was performed.

When reviewed one month later he was markedly improved mentally but could not move his extremely spastic lower limbs. The spinal fluid pressure was 180 mm. water and it was felt that the shunt had blocked. It was not possible to revise this and he was discharged unchanged. In November 1966 a lumboperitoneal shunt was done. No improvement was noted on follow-up one month later.

CASE 9.—H.C., a 69-year-old woman, suffered a subarachnoid hemorrhage in July 1957, and a left internal carotid supraclinoid aneurysm was clipped. She recovered uneventfully, her only residual being a partial third nerve palsy.

In August 1964 she was readmitted because of confusion, disorientation and twitching of the left arm, of sudden onset. Physical examination revealed mild pyramidal signs in the left arm and leg, with sensory inattention. The cerebrospinal fluid was under a pressure of 290 mm. water and was bloody, but the supernatant was clear. This was done 36 hours after her first symptom. She was discharged completely well.

In November 1964 she was readmitted and at this time was felt to be post-ictal. Cerebrospinal fluid pressure was 240 mm. water. She was discharged with a mild hemiparesis and impaired intellect. From November 1964 to May 1965 she showed continuing mental deterioration and presented in May 1965 with a severe dementia and a mild left hemiparesis. A pneumoencephalogram revealed communicating hydrocephalus. A ventriculoatrial shunt was done May 1965 and has resulted in slight improvement. She continues, however, to have impaired recent and remote memory, and functions at home in a limited capacity.

CASE 10.—In June 1958, H.C., a 66-year-old housewife, suffered a subarachnoid hemorrhage. Clinical examination was normal apart from a right extensor plantar reflex. Carotid arteriography failed to demonstrate an aneurysm and she recovered uneventfully. The family subsequently noted mental deterioration, and by November 1958 there was also deterioration of gait.

Examination in January 1959 revealed an obviously demented woman with a mild gait ataxia. An air encephalogram showed gross enlargement of the ventricular system, but no air was seen in the hemispheral sulci. Surgical therapy was considered but was postponed. Readmission one month later was necessitated by a further deterioration intellectually, incontinence, and a complete inability to walk because of a severe gait apraxia. The air encephalogram was repeated but the findings remained unchanged. In February 1959 a ventriculoatrial shunt was performed. She rapidly showed intellectual improvement, her gait became virtually normal and the incontinence cleared.

Over the next five years she resumed her normal domestic duties. In the fall of 1964 her condition deteriorated, with a return of progressive dementia and gait apraxia. Deterioration continued with septicemia from an infected valve, and congestive heart failure eventually led to her death.

Another patient (Case 11) illustrated delayed regression due to a lesion after trauma other than a subdural hematoma. A progressive decline has apparently been arrested by the shunting procedure.

CASE 11.—In 1954, J.A., a 34-year-old man, sustained a compound fracture of the left frontal bone with a post-traumatic right hemiparesis. He did well following surgery and returned to work one year later with minimal neurological deficit. Seizures with a Todd's paralysis of the right upper limb began in May 1955 and continued sporadically.

When admitted to hospital in April 1958 he was thought to be normal mentally. An air encephalogram revealed a large porencephalic cyst in the left frontal lobe, lying beneath the operative bone defect. The remainder of the ventricular system was moderately dilated, as were the sulci. Since 1958 he has been employed only sporadically. In November 1962 he developed dystonia and slight weakness but marked spasticity of the right limbs.

On readmission to hospital in October 1964, he showed mild memory impairment. He was having constant dystonic movements of the right limbs, which were spastic. The right plantar was now extensor. An air encephalogram again showed the porencephalic cyst; the lateral ventricles had enlarged and the previously demonstrated basal cisterns did not show either. No air was present in the subarachnoid sulci. A lumboperitoneal shunt was performed in late 1964. His dystonia and spastic weakness did not progress and his memory remained unchanged. He has not returned to work.

Case 12 illustrates a communicating hydrocephalus secondary to trauma. In this patient improvement was delayed some months after the shunting procedure.

CASE 12.—L.W., a 73-year-old woman, had a one-year history of forgetfulness and declining memory. This became accentuated after surgery for pyloric obstruction; however, she returned to the pre-surgical mental state during her convalescence. During November 1965 she fell down the cellar steps and shortly thereafter she became demented, with incontinence and gait apraxia. Neurological examination was normal apart from the gait disturbance and memory change. A pneumoencephalogram revealed communicating hydrocephalus. The spinal fluid pressure was normal and the protein content 110 mg. %. A lumboperitoneal shunt on April 21, 1966, resulted in a significant improvement in her mental competence and memory. Her incontinence cleared and her gait was improving. However, she fell, probably because of her gait difficulty, and sustained a fractured hip. This resulted in a decline to the pre-shunting state. She was discharged in a state very similar to that noted on admission. However, when seen in October 1966 she was walking with a cane, was quite bright and had the mild memory changes of the aged; she was not incontinent.

An elderly man with chronic meningitis and a communicating hydrocephalus improved somewhat with lumbar punctures, but deteriorated and died postoperatively (Case 13).

CASE 13.—H.L., a 65-year-old labourer, had been in good health apart from an 11-month period beginning in April 1959 when he underwent treatment for pulmonary tuberculosis. In February 1964 he had a febrile illness characterized by headaches, vomiting and nuchal rigidity. Spinal fluid examination revealed 180 white blood cells, mainly lymphocytes, elevated protein and depressed sugar. Cultures were negative and he was subsequently given a three-month course of antituberculous drugs. He returned to work in 1964.

He was hospitalized in March 1965 because of increased somnolence, fatigue, memory impairment and incontinence of fairly abrupt onset. On examination he showed marked apathy and disinterest, and made few spontaneous movements. No spontaneous conversation was heard. He had marked impairment of recent memory although his remote memory was relatively unaffected. His facies were immobile and spastic. Tone in the limbs and neck was increased, and he had right hyperreflexia with a right extensor plantar response. When walking he assumed a flexed attitude and his gait was slow and shuffling. An air encephalogram revealed communicating hydrocephalus. Seven lumbar punctures were done over the next four weeks, and the pressure was never higher than 165 mm. of water. After each lumbar puncture he would be brighter. His cerebrospinal fluid content remained abnormal and included a paretic colloidal gold curve. Cerebrospinal fluid cultures and gastric washes were negative for all organisms and fungi, and antituberculous therapy was reinstituted. It was felt that the infection was tuberculous and might be spread by a shunt; therefore this procedure was postponed.

At home, lumbar punctures were continued and he showed improvement after each one. He was therefore readmitted for surgical therapy in late October 1965. Two lumboperitoneal shunts failed to function and a Pudenz valve was inserted. He aspirated vomitus and died unexpectedly. An autopsy was not performed.

A patient with an encephalopathy of unknown etiology developed a communicating hydrocephalus (Case 14). A lumboperitoneal shunt returned him to his predeterioration state.

CASE 14.—I.P., a 65-year-old man, presented in 1962 with a six-month history of defective memory for recent events, difficulty in concentrating, and indifference. His gait had become shuffling and he had unsteadiness, particularly with the right leg. The right arm was clumsy. Neurological examination revealed mild intellectual impairment. His gait was shuffling and unsteady, but not broad-based. He had generalized hyperreflexia with an equivocal right plantar. There was a coarse tremor of the hands. Detailed general medical investigation was normal. The electroencephalogram showed a mild diffuse dysrhythmia, and an air encephalogram revealed dilated ventricles, but air was present in the basal cisterns and the subarachnoid sulci. The spinal fluid contained 88 mg. % protein, 10 white blood cells and the colloidal gold curve was strongly paretic (5555554210). (Cerebrospinal fluid Wassermann was negative and blood *Treponema pallidum* immobilization and Wassermann tests were negative.) From January 1962 to June 1963 his condition deteriorated slightly. Investigation remained unchanged and he was placed on steroids. The tentative diagnosis was carcinomatous encephalopathy. Within a few months he exhibited mild but definite improvement in gait and memory, and held this modest improvement until May of 1965 when examination showed increased memory impairment, an ataxic broad-based gait and a coarse intention tremor. The right plantar remained equivocal.

In July 1965 he suddenly deteriorated, with increasing drowsiness and clumsiness of the upper limbs. He had more difficulty in walking and required assistance. Physical examination revealed a drowsy but orientated man with only slight mental deterioration. He had sustained coarse nystagmus on horizontal gaze, and mildly increased tone in the lower limbs; he was ataxic on heel-shin testing and had a severely ataxic gait. The plantar responses were equivocal. A pneumoencephalogram revealed communicating hydrocephalus with no air in the hemispheral sulci, but air was present in the insular region. Following this procedure, he deteriorated greatly with increasing confusion and urinary incontinence and became mute and akinetic. A lumboperitoneal shunt was done on September 17, 1965. One month later his condition had not improved; he had psychomotor retardation, and marked disorientation, and was unsteady on his feet. However, six months postoperatively he had improved remarkably, and his gait and memory were similar to what they had been on examination in May. He was able to travel alone by plane to visit friends. The nature of his underlying encephalopathy has not yet been identified.

DISCUSSION

Acute Hydrocephalus

Communicating hydrocephalus has been defined as hydrocephalus in which there is no ventricular obstruction, but rather free communication between the ventricles and the subarachnoid space. However, in hydrocephalus there is always an obstruction of some type. This may be at the tentorial incisura, producing obstruction of anterior communicating pathways or obstruction over the cerebral convexities, or it may occur secondary to leptomeningitis as a result of infection,[4-7] or parasitic infestation[8, 9] or to blood in the subarachnoid space.[1, 2, 10-18] This latter may result from a spontaneous subarachnoid hemorrhage, or be due to trauma, to operative procedures, to rupture of an aneurysm, or to an arteriovenous malformation.

Since the early experimental animal studies of Bagley[19-21] with subarachnoid bleeding, it has been recognized that blood injected into the cisterna magna produced ventricular dilatation in 30% of puppies after a period of time, accompanied by fibrosis of the leptomeninges and infiltration with pigment-containing macrophages. It is the opinion of some[10] that the greater the number of subarachnoid hemorrhages in the same individual, the greater the hydrocephalus. Fibrosis is thought to take 10 to 14 days to develop,[12] but acute hydrocephalus is known to develop before this—within a few days of the bleeding. This early acute hydrocephalus can be the result of a clot obstructing the pathways of cerebrospinal fluid circulation, but on occasion this is not the case and other mechanisms must be sought. One of these mechanisms might be interference with cerebrospinal fluid absorption in the Pacchionian granulations due to an excess of blood in the fluid.

Generally, the symptoms of hydrocephalus which are acute and progressive are those of an increased intracranial pressure.[1, 12] The symptoms respond dramatically to lowering the pressure, either by repeated lumbar punctures or by a shunting procedure. This condition is well known, but in this report all cases with evidence of high CSF pressure have been excluded.

NORMAL-PRESSURE, COMMUNICATING HYDROCEPHALUS

On the other hand, symptomatic normal-pressure, communicating hydrocephalus, idiopathic[2, 3] in etiology or secondary[1, 2, 12] to some other condition, has received little or no attention until recently. At the time of preparing this paper the English literature contained reports of only 11 such cases.[1, 2, 12, 22]

In 1956 Foltz and Ward,[1] in describing 10 cases of communicating hydrocephalus from subarachnoid bleeding, mentioned three patients with normal CSF pressure who had symptoms. One of the patients responded dramatically to ventriculomastoidostomy. A repeat pneumoencephalogram six weeks later revealed the ventricles to have decreased in size. The other two patients whose condition resulted from repeated trauma to the head failed to show symptomatic improvement with repeated lumbar punctures; surgical shunting was not performed.

The authors suggest that the patients with the lower pressure hydrocephalus, while not as critically ill as those with increased intracranial pressure, are more severely affected neurologically. They could not adequately explain the reasons for the neurological picture, but mention that "even with normal intracranial pressure the deficits attributable to the hydrocephalus can still be reversed by a shunt operation".[1] About 10 to 12% of patients bleeding from the midline area of the brain show hydrocephalus and about 5 to 7% have progressive symptoms and require shunting.[14] In some patients the condition is self-limiting.

In 1965 two papers[2, 3] dealt with the clinical features of six cases (three were idiopathic in origin, two followed trauma and one was associated with a tumour). A hypothesis was proposed for the mechanism of symptomatic hydrocephalus with relatively normal or slightly elevated pressure. It was felt that "the expanded state of the ventricles themselves play an important role in determining the effect of the cerebrospinal fluid pressure on the brain".[2] In accordance with Pascal's law (Force = Pressure x Area), a "given pressure of CSF would therefore be expected to have a greater force when applied to the walls of enlarged ventricles than to small ones".[2] Hakim and Adams[2] felt that other factors probably come into play also, but that a distinction between force and pressure must be appreciated. They also felt that a pressure of 180 mm. of water is a pathological increase in their cases with enlarged ventricles and that this may be responsible for the maintenance of hydrocephalic symptoms.

ETIOLOGICAL FACTORS

Some of the etiological features in our cases are summarized in Table I. Several groups were recognized: (a) those with bleeding due to subarachnoid hemorrhage or trauma; (b) one with infection (probably tuberculous); (c) one with an undiagnosed encephalopathy, and (d) the remainder due to unknown causes. Of particular note is the fact that two patients in this series had communicating hydrocephalus secondary to bleeding from middle cerebral aneurysms. Kibler, Couch and Crompton[12] state that communicating hydrocephalus rarely

TABLE I.—ETIOLOGICAL FACTORS

Number of patients—14				
Etiology				
I	Idiopathic			5
II	Subarachnoid bleeding			5
	A *Ruptured aneurysm*		4	
	Middle cerebral	2		
	Internal carotid	1		
	Basilar	1		
	B *Unidentified subarachnoid bleeding*		1	
III	Post-traumatic			2
IV	Chronic meningoencephalitis (probably tuberculous)			1
V	Diffuse encephalopathy —etiology unknown			1

or never occurs with middle cerebral aneurysms, despite the fact that they constitute some 25 to 30% of all aneurysms.[23] Our experience is therefore at variance with this report.

Time of Onset

The onset of symptoms secondary to subarachnoid hemorrhage usually began within two months of the hemorrhage, and although it is difficult to be sure, over 50% appeared to begin almost immediately. In one case, secondary to trauma, the onset of symptoms was delayed eight years.

TABLE II.—SUBARACHNOID BLEEDING

Case No.	6	7	8	9	10
Age at time of hemorrhage (years)	58	50	31	67	66
Sex	Male	Female	Male	Female	Female
Site of aneurysm	Basilar	Middle cerebral	Middle cerebral	Internal carotid	Not identified
Number of hemorrhages	?2	1	?1	2 or 3	1
Length of time after bleeding to onset of symptoms	? At once	2 months	? At once	?	1 month

Tables II, III and IV detail the age, sex and, where applicable, the site of bleeding, the number of hemorrhages and the length of time between bleeding or trauma and the onset of symptoms.

TABLE III.—POST TRAUMATIC

Case No.	11	12
Age at time of injury (years)	22	74
Sex	Male	Female
Time interval after trauma to onset of symptoms	9 years	Immediately

The most constant features were the triad: (1) memory deterioration, (2) gait abnormalities and (3) incontinence. Impaired memory was present in all 14 patients, and confusion and apathy in 11. The changes were severe in 10 and slight in 4. Gait disturbance was present in 12, with six being apractic, three ataxic, one dystonic and spastic, one "cautious" and one unable to walk owing to severe spasticity. Incontinence was a major feature in nine. Pyramidal signs of recent origin were present in six cases.

RADIOLOGICAL INVESTIGATION

Pneumoencephalography revealed communicating hydrocephalus in all 14 patients. The ventricles were dilated but not obstructed. No air was present in the sulci over the cortex except for the fact that air was seen in the insular regions in 33% of the patients. The point of relative obstruction appeared to be most often at the tentorial notch or in the sulci over the hemispheres. These radiological features are shown in Figs. 1, 2 and 3.

Hakim and Adams[2] have emphasized that pneumoencephalography often causes abrupt deterioration in the condition of these patients. Our experience indicates that this is by no means invariable, and indeed in some the reverse phenomenon has been seen. Two patients improved remarkably after a pneumoencephalogram. This improvement, although temporary, appeared more lasting than when a lumbar puncture was done and a large quantity of fluid removed. Four patients deteriorated after the air study; one became akinetic and mute, and remained so until repeated lumbar punctures, with removal of large volumes of cerebrospinal fluid, were carried out.

CEREBROSPINAL FLUID PRESSURE

The cerebrospinal fluid pressure was normal in all patients, as measured in either the left lateral position or in the sitting position at the time of the air study—i.e. the pressure, recumbent, was not higher than 180 mm. water and there was no clinical evidence of increased pressure.

The cerebrospinal fluid content was completely normal in six cases; seven showed pro-

TABLE IV.—CLINICAL FEATURES

Case No.	*1*	*2*	*3*	*4*	*5*	*6*	*7*	*8*	*9*	*10*	*11*	*12*	*13*	*14*
Mental state														
Poor memory	x	x	x	x	x	x	x	x	x	x	slight	x	x	x
Confusion	x	x	x		x	x	x	x		x		x	x	x
Apathy	x	x	x		x	x	x	x		x		x	x	x
Gait disturbance														
Apraxia	x	x				x				x		x	x	
Ataxia			x	x										x
Other							x	x			x			
Incontinence	x	x	x			x		x		x		x	x	x
Miscellaneous														
Pyramidal signs		x	x		x				x		x		x	
Extrapyramidal signs			x								x		x	
Pseudobulbar signs		x	x											
Tremor			x											x

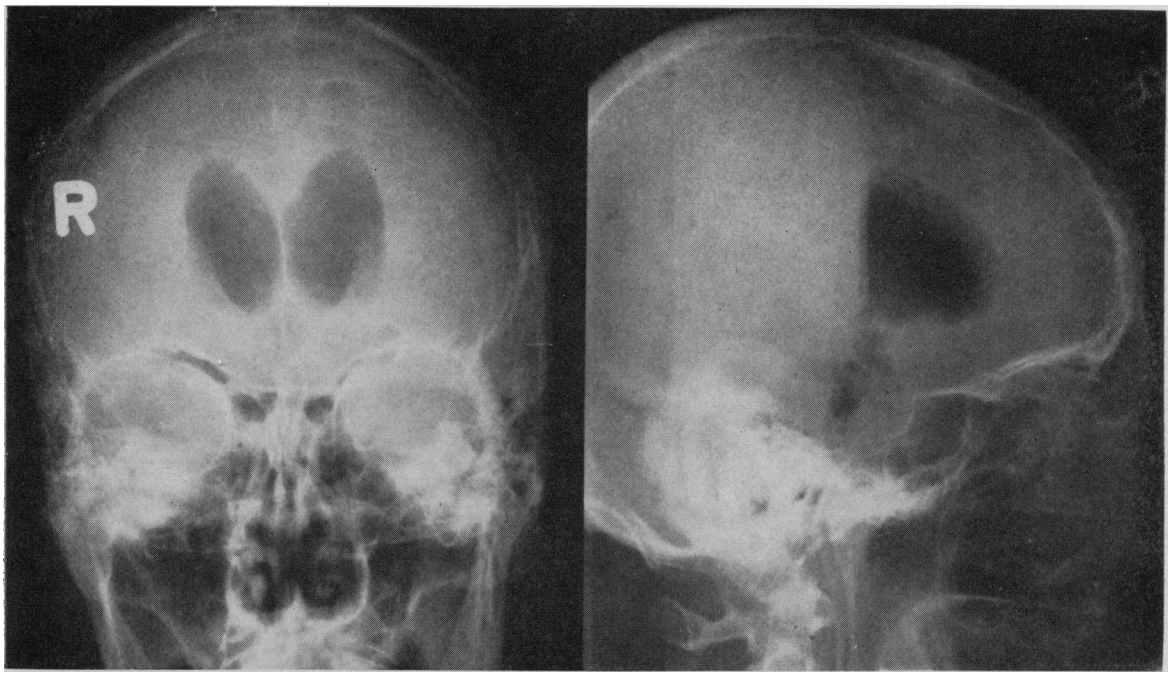

Fig. 1.—A.P. and lateral brow up films showing marked ventricular dilatation and air in the prepontine and interpeduncular cistern. The ventricular span is 57 mm. (normal: 40 mm.[27]).

tein elevation as high as 110; four showed increased cells and two had paretic colloidal gold curves. These abnormalities are likely meaningless in terms of communicating hydrocephalus, as they appear to reflect the associated intracranial lesion.

TREATMENT

In twelve patients shunts were performed: seven had lumboperitoneal shunts and five ventriculoatrial shunts (one lumboperitoneal shunt was revised to a ventriculoatrial shunt, and one ventriculoatrial shunt was later revised

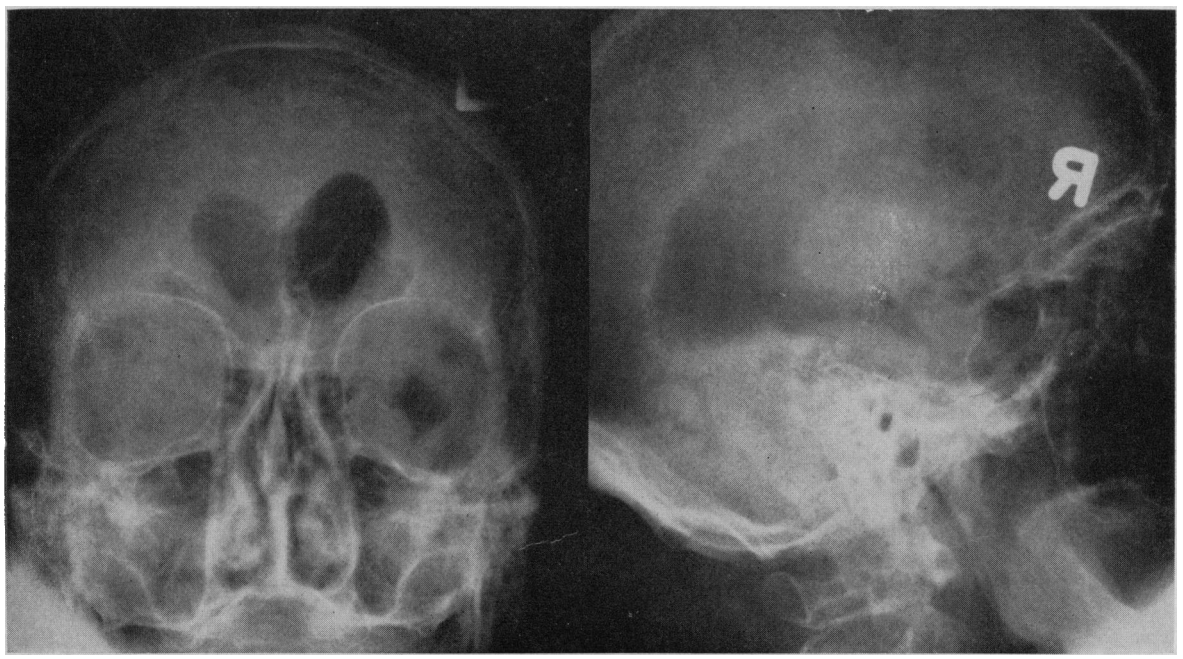

Fig. 2.—Temporal horn films done at the termination of the pneumoencephalogram procedure. These show left temporal horn dilatation with air in the sulci of the left insular area but failure of other sulci to fill.

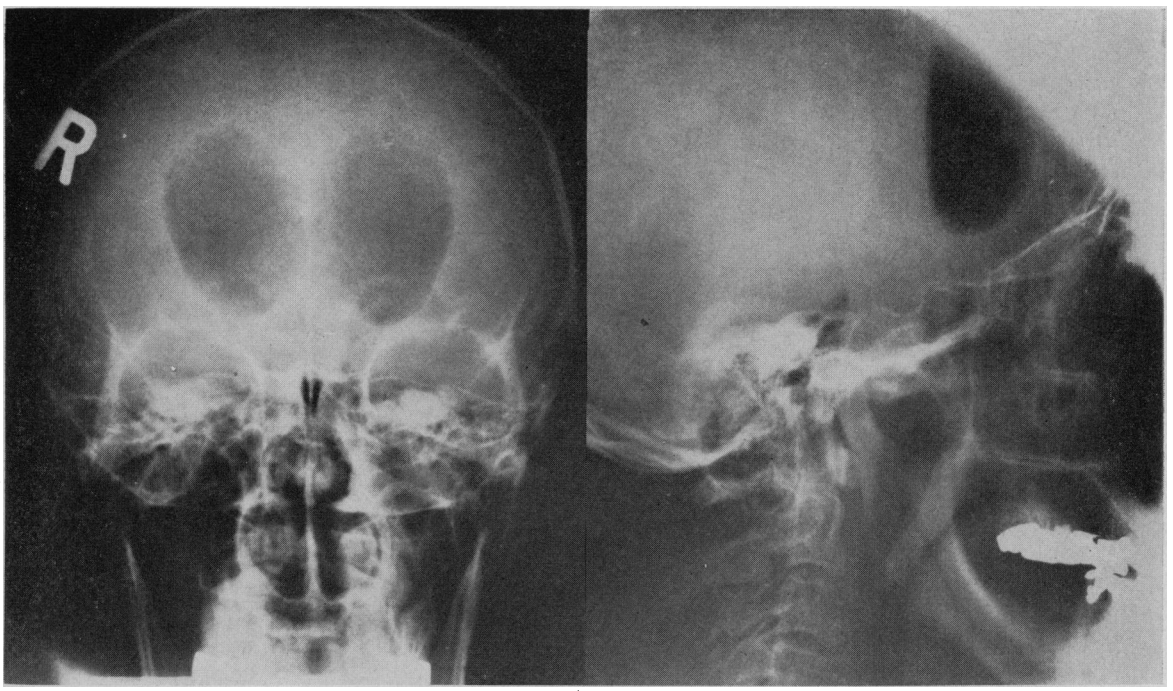

Fig. 3.—A.P. and brow up films showing marked ventricular dilatation with a ventricular span of 88 mm. Air is present in the prepontine cistern, but none is present in the other basal cisterns or in the insular sulci.

to a lumboperitoneal shunt). Two patients were treated with repeated lumbar punctures (one subsequently had a shunt performed but died in the postoperative period). It was our preliminary policy to try the effect of several lumbar punctures as initial therapy. We felt that this might prove to be a useful guide to the patients who might benefit from a shunting procedure. Our subsequent experience has led us to doubt the universal validity of this guide. Significant early improvement was lacking in one patient (Case 12), but she went on to gradual improvement after the shunting had been effected. Conversely, one of the patients (Case 7) improved enough that a shunting procedure did not seem justifiable. Perhaps in this and other cases, particularly after bleeding, spontaneous re-establishment of a balance between cerebrospinal fluid formation and resorption can be attained. To wait for this in all cases, though, might lead to irreversible damage that might be avoided by earlier shunting.

Follow-up of these patients ranged from 3 to 24 months, with an average of 12¾ months (excluding one patient followed up for 6¾ years). Results are shown in Table V and summarized in Table VI.

As can be seen from these tables, the greatest number of patients were functionally improved in regard both to mentation and to gait. Of 14 with mental impairment two remained unchanged. Gait abnormalities were improved in 10, remained unchanged in one, and became worse in one. This latter patient somewhat improved mentally but his limb power got worse. He was found to have a blocked shunt and this could not be revised. A lumboperitoneal shunt was done some months later, but examination one month after the procedure revealed no improvement. Incontinence cleared in eight patients in whom it was present and was unchanged in one. While they are encouraging, these results are not meant to imply that the patients became completely normal. Some could, however, carry on household duties or return to jobs in a lesser capacity.

While repeated lumbar punctures may not produce improvement, this should not be taken as evidence that a shunting procedure will not be of value. In fact, repeated lumbar punctures in one patient (Case 12), failed to bring about improvement, but the patient eventually responded well to a shunting procedure. Our observations suggest that the improvement will continue for a number of weeks or months in some, and may therefore not be evident after only a few lumbar punctures.

Complications from any shunting procedure are quite common and include obstruction of the ventricular catheter,[13] thrombosis of the jugular vein (when the cardiac end is too high),[13] septicemia,[13, 24] pulmonary emboli, men-

TABLE V.—RESULTS

Case No.	Duration of symptoms in months before therapy	Procedure	Mentation	Gait	Incontinence	Follow-up in months	Results
1	12	July 1965—L.P. shunt which occluded in April 1966 and a V.A. Pudenz valve was inserted.	Moderately improved	Moderately improved	Cleared	18	Returned to work but is not as productive as previously. Recently memory again failing although the shunt is functioning.
2	12	March 1966—L.P. shunt which occluded in April but became functional spontaneously.	Moderately improved	Improved after the L.P. shunt but declined when it was blocked. Has not returned to his best state after the shunt.	Cleared	12	Retired but probably couldn't work. Extensor plantar remained.
*3	24	December 1965—V.A. shunt, Pudenz valve. Revised three times from Feb. 1 to 23, 1966.	Slightly improved	Slightly improved	Cleared	12	Subdural removal February 1966, and septicemia treated. Has returned home but can't cope with household duties.
*4	6	November 1965—L.P. shunt.	Slightly improved	Normal	—	6	Returned to work for one year but efficiency diminished.
*5	8½	December 1965—L.P. shunt.	No change	—	—	6	Not able to return to work.
*6	8½	February 1965—V.A. shunt, Pudenz valve	Moderately improved	Became normal	Cleared	21	Returned to work in a lesser capacity. Bled again but again able to return in a menial capacity. Further hemorrhage December 1966 and died.
7	4	Repeated lumbar punctures over three months.	Slightly improved	Became normal	Cleared	15	Function at home is impaired by an emotional reaction. Continues to have severe headaches.
8	6	April 1966—V.A. shunt, Pudenz valve. This occluded in May and could not be revised. November 1966 L.P. shunt.	Slightly improved	Became worse	Unchanged	8	Bed-ridden.
9	6	May 1965—V.A. shunt, Pudenz valve	Slightly improved	—	—	12	Able to return home and to cope with household duties under supervision.
10	7	February 1959—V.A. shunt, Holter valve.	Moderately improved	Became normal	Cleared	6¾ years	Died 6¾ years after Holter valve was inserted. Had carried on adequately at home for 5½ years.
11	18 (nine years after the head injury)	November 1964—L.P. shunt.	No change (symptoms did not progress)	No change	—	24	Dystonia did not progress. Has not worked since 1958.
*12	4	April 1966—L.P. shunt	Moderately improved	Moderately improved	Cleared	7	Deteriorated when hip broken but gradually improved and returned home to do part of the housework.
13	14	Repeated lumbar punctures.	Slightly improved	Slightly improved	Cleared	9	Died after V.A. (Pudenz) shunt.

TABLE V.—RESULTS (continued)

Case No.	Duration of symptoms in months before therapy	Procedure	Mentation	Gait	Incontinence	Follow-up in months	Results
*14	? 2 - 3	September 1965—L.P. shunt.	Moderately improved	Moderately improved	Cleared	15	Has resumed his usual activity of retirement. In recent weeks, mental and gait deterioration. Shunt functioning.

Key: *Insular air. —Symptoms never present.

ingitis[13] and subdural hematoma.[25] With lumboperitoneal shunts there is a high incidence of permanent failure of the shunt.[13] Recently another complication has been reported in which a communicating hydrocephalus has been turned into a stenosis or occlusion of the aqueduct, presumably owing to a continuation of the basic process producing the hydrocephalus.[26] The incidence of complications in this series included five revisions of ventriculoatrial shunts—three in one patient and one each in two others—and one instance each of subdural hematoma, septicemia and probable emboli.

Failure of a shunting procedure (lumboperitoneal shunt) to cause reversal of symptoms or to lead to early improvement should raise doubt as to the patency of the shunting catheter. In recent cases, in an endeavour to settle this question, we have been assessing the value of an intrathecal injection of radioiodinated serum albumin (RISA) with subsequent counts over the abdomen. Initial results suggest that this may be a valuable adjunct in the follow-up of these patients.

The lumboperitoneal shunt is a simple procedure and we prefer to start with it even though revision or change to a ventriculoatrial shunt will be required in a high percentage of the cases which have had an early favourable response.

Patients showing the least improvement were those: (1) whose main symptoms were other than impaired intellectual capacity, gait deterioration or incontinence, as for example those with dystonia; or (2) in whom there is reason to suspect that the symptoms may be related to an additional process as well as to communicating, normal pressure hydrocephalus. In this group of patients the duration of symptoms alone has failed to affect the eventual prospects of improvement.

TABLE VI.—SUMMARY OF RESULTS

Intellectual decline		(14 cases)
Improved	10	
No change	4	
Gait disturbance		(12 cases)
Improved	10	
No change	1	
Worse	1	
Incontinence		(9 cases)
Cleared	8	
No change	1	

The pathophysiology of the condition of "symptomatic normal-pressure, communicating hydrocephalus" remains unexplained. What is needed is an experimental model of the brain, its membranes, its pulsatile vasculature and its cerebrospinal fluid medium. The production of this model will be an elaborate feat. In the meantime, we may learn more about this condition by repeated intraventricular pressure readings combined with microcirculation studies.

Still unexplained is the reason for improvement when a "normal" cerebrospinal fluid pressure is reduced even further by a shunting procedure or by repeated lumbar punctures. It may be, as Hakim and Adams[2] suggest, that a pressure of 180 mm. of water in enlarged ventricles is pathological. Repeated post-treatment pneumoencephalograms, not yet available, may be helpful to study in conjunction with the results of treatment. It will be interesting to know if detectable decline in ventricular size is present with those who improve the most. The nature of the histopathology in the idiopathic types is obscure but may represent a circulatory and absorptive failure secondary to arachnoiditis.

Asymptomatic Cases

Since we have become interested in this condition, several patients have been seen with the radiological findings of communicating hydrocephalus, in whom it was difficult to know whether their symptoms could in any way be related to this condition. The question at issue is whether or not there is a symptomatic, mild or reversible form of normal-pressure, communicating hydrocephalus. The question also arises whether or not this radiological finding may be seen and be of no significance. Since the study was begun, several cases have been seen without any of the classical features of this clinical condition, without dementia, and yet air was

not present over the convexity of the brain in a pneumoencephalogram. We are following up these cases to see what ultimately develops.

Summary Fourteen patients with symptomatic communicating hydrocephalus and normal cerebrospinal fluid pressure are described. The main features were: (1) progressive dementia, which is often considered to be hopeless; (2) gait disturbance and incontinence; and (3) communicating hydrocephalus, which is often unsuspected. The pathophysiology of the condition is obscure, but shunting procedures or even repeated lumbar punctures have produced symptomatic improvement in some cases, most markedly in those with the triad of symptoms. The complications associated with shunting procedures are described. One patient had a 5½-year interlude of reasonable normality after this treatment was carried out.

Résumé Les auteurs présentent 14 malades atteints d'hydrocéphalie communicante symptomatique à pression du LCR normale. Les principales caractéristiques de ces sujets consistaient en: (1) une démence évolutive qui est généralement considérée comme étant désespérante; (2) des troubles de la démarche et de l'incontinence et (3) une hydrocéphalie communicante qui reste souvent ignorée. La pathophysiologie de la maladie demeure inconnue mais on a noté, en quelques cas, une amélioration symptomatique sous l'influence d'un shunt chirurgical ou même par des ponctions lombaires répétées. Cette amélioration est surtout frappante chez les malades qui présentent la trilogie de symptômes. On expose les complications qu'entraînent les shunts opératoires. Un des malades étudiés a bénéficié d'une période de 5½ ans de normalité raisonnable après cette opération.

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REFERENCES

1. FOLTZ, E. L. AND WARD, A. A., JR.: *J. Neurosurg.*, **13**: 546, 1956.
2. HAKIM, S. AND ADAMS, R. D.: *J. Neurol. Sci.*, **2**: 307, 1965.
3. ADAMS, R. D. *et al.*: *New Eng. J. Med.*, **273**: 117, 1965.
4. FOLTZ, E. L. AND SHEEHY, T. F., JR.: *Amer. Rev. Tuberc.*, **74**: 835, 1956.
5. JACKSON, I. J. AND THOMPSON, R. K., editors: Pediatric neurosurgery, Charles C Thomas, Publisher, Springfield, Ill., 1959, p. 176.
6. LORBER, J.: *Arch. Dis. Child.*, **26**: 28, 1951.
7. RUSSELL, D. S.: *Med. Res. Counc. Spec. Rep. (London)*, No. 265: 1, 1949.
8. CÁRDENAS, J. C.: *J. Neurosurg.*, **19**: 635, 1962.
9. DIXON, H. B. F. AND LIPSCOMB, F. M.: *Med. Res. Counc. Spec. Rep. (London)*, No. 299: 1, 1961.
10. ASKENASY, H. M., HERZBERGER, E. E. AND WIJSENBEEK, H. S.: *Neurology (Minneap.)*, **3**: 213, 1953.
11. MERWARTH, H. R. AND FREIMAN, I. S.: *Brooklyn Hospital Journal*, **1**: 149, 1939.
12. KIBLER, R. F., COUCH, R. S. C. AND CROMPTON, M. R.: *Brain*, **84**: 45, 1961.
13. SCARFF, J. E.: *J. Neurol. Neurosurg. Psychiat.*, **26**: 1, 1963.
14. SHULMAN, K. *et al.*: *J. Neurosurg.*, **20**: 1040, 1963.
15. STRAIN, R. E. AND PERLMUTTER, F.: *Bull. Univ. Miami Sch. Med.*, **8**: (No. 3): 23, 1954.
16. STRAUSS, I., G., GLOBUS, J. H. AND GINSBURG, S. W.: *Arch. Neurol. Psychiat. (Chic.)*, **27**: 1080, 1932.
17. SWEET, W. H.: *Ibid.*, **45**: 86, 1941.
18. PENFIELD, W. AND CONE, W.: *Canad. Med. Ass. J.*, **48**: 99, 1943.
19. BAGLEY, C., JR.: *Arch. Surg. (Chicago)*, **17**: 18, 1928.
20. *Idem*: *Ibid.*, **17**: 39, 1928.
21. *Idem*: *Ass. Res. Nerv. Dis. Proc.*, **8**: 217, 1929.
22. MESSERT, B. AND BAKER, N. H.: *Neurology (Minneap.)*, **16**: 440, 1966.
23. WALTON, J. N.: Subarachnoid haemorrhage, E. & S. Livingstone Ltd., Edinburgh, 1956.
24. COHEN, S. J.: *Develop. Med. Child Neurol.*, **4**: 298, 1962.
25. DAVIDOFF, L. M. AND FEIRING, E. H.: *J. Neurosurg.*, **10**: 557, 1953.
26. FOLTZ, E. L. AND SHURTLEFF, D. B.: *Ibid.*, **24**: 520, 1966.
27. DAVIDOFF, L. M. AND DYKE, C. G.: The normal encephalogram, 3rd ed., Lea & Febiger, Philadelphia, 1951, p. 104.