Some Observations on C.S.F. Pressure. Hydrocephalic Syndrome in Adults with "Normal" C.S.F. Pressure. (Recognition of a new syndrome.)

Salomon Hakim, M.D.
Bogotá, Colombia, S.A., March 10, 1964

Translated from Spanish:

**Algunas Observaciones Sobre la Presion del L.C. R. Sindrome
Hidrocefalico en al Adulto con "Presion Normal" del L.C.R.**
(Presentacion de un Nuevo Sindrome)

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Foreword to English Translation

This monograph, published originally in Spanish in 1964, was written after it became evident that certain peculiar, not well identified, and poorly understood cases of hydrocephalus ought to be classified as a special group. These patients, who exhibited progressive hydrocephalus and other neurological and mental symptoms in the presence of normal cerebrospinal fluid pressure, improved unexpectedly when the C.S.F. pressure was permanently lowered below normal.

While searching for a rationale to explain this paradox, a case with hydrocephalus secondary to subarachnoid bleeding was reviewed and speculated upon. Research was conducted into the possible physical principles involved. Then the importance of the area of the ventricles was recognized, and the mechanism of symptomatic hydrocephalus with normal C.S.F. pressure was explained on the basis of increased force due to the pressure-area relationship of the ventricles $(F = P \times A)$

After a patient was seen with this combination of symptoms and findings but without any history that would account for the hydrocephalus (intracranial hemorrhage, trauma, meningitis, surgery, etc.), a search was begun for the so-called idiopathic cases. These patients are most important and surprising because of the danger that they will be misdiagnosed and placed in a hopeless category (organic brain diseases). It is in the large group of patients with late-life dementia that further cases must be sought. Finally, these observations were communicated by lectures, motion pictures, and the 1964 monograph.

Dr. Raymond D. Adams encouraged the author to write a summary of the monograph in English, and this paper appeared under the title "The Special Clinical Problem of Symptomatic Hydrocephalus with Normal Cerebrospinal Fluid Pressure" in the *Journal of Neurological Sciences*, volume 2, pages 307-327, 1965.

Since the appearance of the 1965 summary and a second paper* written in English with the Massachusetts General Hospital group, interest in the syndrome of symptomatic hydrocephalus with normal C.S.F. pressure has intensified and become widespread. To fill the many requests for the full text of the 1964 monograph in English, the accompanying translation has been prepared. It is hoped that the reader will find the many details described here to be useful, especially the explanations of the experiments conducted into the physical principles related to the causation of the syndrome. All of these hypotheses show that the symptoms perhaps are related not to the C.S.F. pressure alone but to the total force, which equals pressure times the area of the enlarged ventricles. The hydraulic press mechanism might explain why these patients can show a dramatic improvement in a short time when the C.S.F. pressure is lowered below what had been considered normal.

The author wishes to express special gratitude to Doctors Raymond D. Adams, C. Miller Fisher, William H. Sweet, and Robert G. Ojemann for their encouragement and cooperation in helping to inform the medical profession about the recognition of this syndrome and the ideas developed in this work.

Bogotá, Colombia   Salomon Hakim, M.D.

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*Adams RD, Fisher CM, Hakim S, Ojemann RG, Sweet WH: Symptomatic occult hydrocephalus with "normal" cerebrospinal fluid pressure: a treatable syndrome. *New Eng J Med* 273: 117-126, 1965.

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I Introduction

We have observed in a number of patients the paradoxical combination of symptomatic hydrocephalus and normal cerebrospinal fluid (C.S.F.) pressure. In each of these cases, an unexpected improvement was noted in the progressive neurologic and mental symptoms when this "normal" pressure was reduced by repeated lumbar punctures or by a ventriculo-atrial shunt.

We are submitting this report because our search of the literature did not reveal any clear and well documented cases of progressive hydrocephalic symptomatology in the presence of normal C.S.F. pressure. Also, we want to describe the benefits of the shunt procedure, which in most cases produces what amounts to a cure. We wish to call attention to the importance of recognizing hydrocephalus with normal pressure and the opportunity for rescuing from oblivion at least some of those patients who, inasmuch as this syndrome has not been recognized previously, have been diagnosed as hopeless cases of post-traumatic encephalopathy, arteriosclerotic and degenerative brain disease, and, particularly, Alzheimer's dementia. We have made this mistake ourselves several times, and other neurologists and neurosurgeons probably have done the same.

Another important point disclosed by our study of symptomatic hydrocephalus with normal C.S.F. pressure is that until now the area over which this pressure is applied has not been taken into consideration, even when the ventricles might possibly be enlarged. Therefore, the surface of the ventricular system may be an important factor in determining the effect of C.S.F. pressure on the brain. The expansive force within the ventricles is not equal to the intraventricular pressure alone; rather, it is the product of the ventricular pressure and the ventricular area. In other words, the larger the area of the ventricular system, the lower the pressure that can produce symptoms of cerebral disturbance. This aspect of C.S.F. hydrodynamics has thus far been overlooked, and it constitutes a new concept in C.S.F. physiopathology. The possibility of applying this important physical principle in our cases will be discussed, as will its possible role in other abnormalities of C.S.F. circulation, such as subarachnoidal cysts, porencephalia, infantile hydrocephalus, and syringomyelia.

In this report we will describe only three of our patients. The inclusion of additional cases would only result in a duplication of the principles presented in the three cases to be described.

The Bases for these Observations Originated in the Following Case:

Case 1. In December of 1957, a 16-year-old boy was hit by a car. He immediately lost consciousness and was brought to the hospital. Neurological examination was not remarkable except for his confused and agitated state and a 3-cm scalp wound in the right parietal region. X-rays of the skull were normal. Three hours later the patient was comatose, and his right pupil was dilated. A carotid arteriogram showed a right subdural hematoma, which was drained.

He then improved to the point that he seemed to be conscious but without having contact with his surroundings. The patient opened his eyes, looked around, did not speak, did not follow commands, lacked control of his sphincters, but moved all four of his extremities and assumed catatonic-like postures. He remained in this state for about one month, interrupted by long periods of sleep. A bilateral carotid arteriogram, which was performed to rule out a chronic subdural effusion, demonstrated hydrocephalus. A spinal tap showed an initial pressure of 150 mm, and 15 cc of clear, slightly yellow fluid was removed. Final pressure was 135 mm. The day after this procedure the boy said a few words and followed simple commands. An encephalogram showed dilatation of the entire ventricular system. After the air studies he deteriorated. A new lumbar tap showed initial pressure of 180 mm. A ventriculo-atrial shunt was then performed to lower the C.S.F. pressure, and soon thereafter the patient improved.

History and Examination

This youth was admitted to the hospital after he was hit by a car while playing in the street and was rendered unconscious.

On admission to the hospital the patient was confused, agitated, pale, and breathing stertorously. Both pupils were pinpoints and the eyes were moving conjugately from side to side. Pulse was regular at 115 per minute; blood pressure was 80 systolic, 60 diastolic.

The heart, lungs and abdomen were normal. In the right frontal-parietal region there was a 3-cm wound of the scalp with bleeding from macerated edges. There were edema and bruising of the right cheek. He was alternately agitated and drowsy, and he vomited once. He had a short seizure consisting of shaking of the left arm and simultaneous dilatation of the pupils. Tendon reflexes were sluggish. The plantars were flexor on the right, uncertain extensor on the left. He reacted to painful stimuli in a symmetrical way, perhaps better on the right side. X-rays of the skull were normal. The wound was sutured. Antibiotics, oxygen, and intravenous fluids were given along with 0.10 grams of phenobarbital intramuscularly to prevent further convulsions.

The patient was kept under observation. Three hours later his right pupil dilated. At that time he responded only to strong, painful stimuli. There was a left hemiparesis with very clear Babinski's signs on that side. An arteriogram showed an avascular area over the convexity of the right hemisphere.

Under local anesthesia a burr hole was made above and behind the right frontal bone. The dura appeared bluish and under tension. When an incision was made in the dura, a small amount of partially clotted blood escaped. After the subdural space was washed with warm saline solution, it was observed that the brain expanded. The cerebral cortex in the parietal region was bruised and had subarachnoid hemorrhages.

The next day the pupils were equal and reacted to light. Both eyes were conjugately deviated to the right. Left plantar response was extensor, right was flexor. The patient reacted more to painful stimuli than on the previous day. His breathing was regular. Blood pressure 100/65. Pulse 100. Temperature 37.5°C. There was neck stiffness. He was maintained on 100 mg of

oxytetracycline (Terramycin) I.M. every 6 hours and 1500 cc of dextrose I.V. every 24 hours. After the second day he was fed through a tube. He also was given 50 mg of phenobarbital three times a day through the gastric tube. A catheter was placed in the urethra.

After three days the temperature was normal, blood pressure 120/70, pulse rate 70. Neck stiffness was still present. Respiration normal. Spontaneous eye movements were now normal. Pupils reacted normally to light. The eyegrounds showed engorgement of the veins, but the discs were normal. Tube feeding was continued for 15 more days, and he was kept on the same medication with the addition of 500 mg of sulfadimethoxine (Madribon) daily. During this time he remained unchanged. A week before discharge he started to open his eyes, look around, moved all his limbs, but did not communicate, nor did he make any sounds. The boy was sent home because it was felt that no further treatment would be of help for his post-traumatic encephalopathy.

A month later on his father's insistence he was readmitted to the neurological service. At home there was no improvement in his condition except that he could be fed orally. To awaken him to eat, it was sometimes necessary to slap his face.

When examined on his return to the hospital he was pale and weak. He opened his eyes but did not follow or attempt to look at any of the persons around him. He withdrew from painful stimuli but did not make any sounds.

There were some nystagmoid jerks when he looked to either side. The eye fundus discs were sharply outlined, and the vessels were normal. The neck was supple. When loud noises were made near him, he immediately closed his eyes. He did not understand or follow simple commands. There was some resistance to passive movements in all four limbs. Tendon reflexes were active, and he displayed bilateral extensor plantar responses. Bilateral carotid arteriograms were done again to rule out a chronic subdural effusion. These arteriograms were normal except for a widened sweep of the anterior cerebral artery consistent with hydrocephalus, which was not present in the first arteriogram (Figure 1, A and B). A spinal tap showed an initial pressure of 150 mm of water and a final pressure of 135 after 15 cc of cerebrospinal fluid were removed. Fluid was slightly xanthochromic and contained one lymphocyte; protein was 60 mg%.

The next day the patient was better. He looked at his hands and followed the examiner with his eyes. He mumbled a few words but he remained incontinent. A lumbar air encephalogram was done. This showed symmetrical dilatation of the ventricular system (Figure 1, C and D). This procedure seemed to aggravate the patient's condition. He again became stuporous, and the next day he was in the same condition. The C.S.F. pressure was measured and was found to be 180 mm. Since it was thought that the hydrocephalus could contribute to the symptomatology, a catheter was introduced in the right lateral ventricle to drain the C.S.F. Once again improvement was noticed.

In view of the patient's improvement after the first lumbar puncture when 15 cc of C.S.F. were removed and his worsening with the air study, it was decided to perform a ventriculo-atrial shunt to permanently lower the C.S.F. pressure.
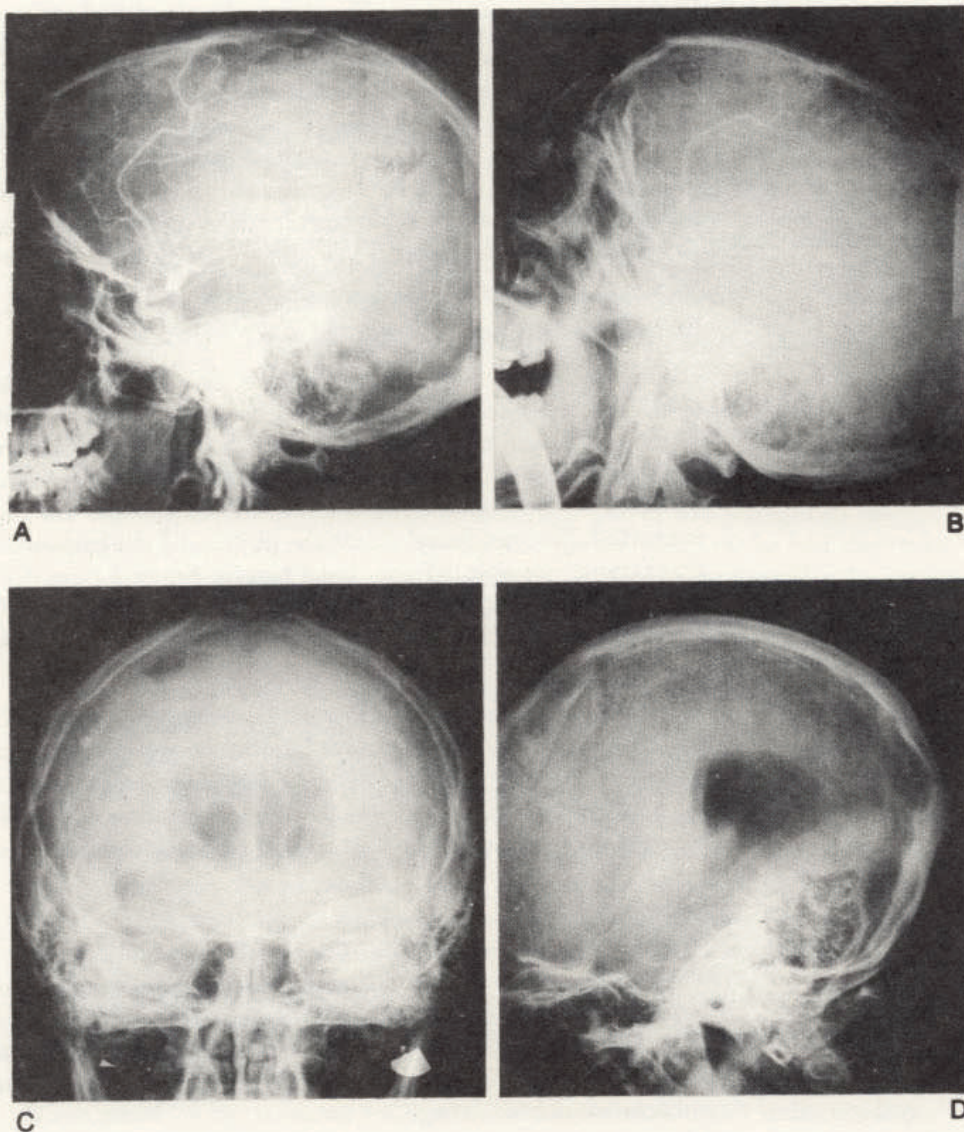
Figure 1. Case 1. A — Lateral view of the carotid arteriography done soon after the accident. B — Similar view taken one month later, showing stretching of the anterior cerebral artery due to ventricular dilatation. C and D — Air studies done at the time of the second arteriogram (B), confirming the hydrocephalus.

Course of His Illness

A ventriculo-atrial shunt was performed under local anesthesia, utilizing a Hakim valve. Shortly before this procedure, C.S.F. pressure was 170 mm. On the day after surgery, the patient showed a dramatic improvement, and he began to talk in phrases, although slowly and in a monotone. He obeyed simple commands. Two days later he was more alert although still incontinent. On the fifth postoperative day he ate by himself and was continent of urine and feces. He was able to carry on a simple conversation with a little dysarthria. He was able to get out of bed and walk a few steps with help, but he was very unstable.

Ten days after the operation a lumbar puncture showed a C.S.F. pressure of 80 mm, and the patient appeared much better.

During one of the neurosurgical rounds, one of the residents stated that he could not see the reason for the improvement in this patient after the removal of the C.S.F. and the lowering of the C.S.F. pressure since his pressure had been normal. This was the moment when we thought that a possible explanation could be based on the relation between the C.S.F. pressure and the area on which it is exerted. We speculated about the physics of the hydraulic press principle. As an example, we pointed out that a bicycle tire and a car tire may have approximately the same pressure, although there is a great difference in the weight of the two vehicles. This is possible because of the different areas of the two tires.

The patient left the hospital fifteen days after the ventriculo-atrial shunt was performed. He was seen a month later in the outpatient department. He had gained weight and was alert. His gait and neurologic examination were normal; his conversation, however, was still slow and monotonous. Three months later his speech was completely normal and he was attending school.

Comment

The obvious cause for the initial unconsciousness in this patient was the cerebral concussion aggravated by the acute subdural hematoma. At the beginning of this case, the hydrocephalus was produced by an augmentation in C.S.F. pressure as a result of cerebral edema and the blockage of the C.S.F. pathways by the subarachnoid hemorrhage.

Lumbar puncture was not performed during the initial days of his illness but ophthalmoscopic examination showed engorgement of the vessels, which could indicate a beginning of intracranial hypertension. Without gaining consciousness, the patient passed from a state of coma to a state of akinetic mutism without sphincteric control and indifferent to his surroundings, although he appeared to be conscious. His condition did not improve, and this was interpreted as being due to irreversible cerebral damage. A month later a second arteriogram showed his ventricles were enlarged, which was not shown in the first arteriogram (Figure 1, A and B). C.S.F. pressure was normal. It was surprising that when 15 cc of fluid were removed there was an improvement, probably as the result of lowering the C.S.F. pressure below normal levels.

The pneumoencephalogram probably had worsened the patient's condition by increasing the C.S.F. pressure (180 mm) due to the dilatation of the air volume (Cronqvist, Lundberg and Pontén, 1963) and by the consequent blockage of the subarachnoid space (Cushing, 1926; Foley, 1957; Johnson, 1958), thus suggesting an important hydrocephalic factor. The favorable response to the shunt supported this suggestion.

Case 2. Male, age 52, musician. This patient was admitted to the hospital because of progressive loss of memory of one year's duration, lack of interest in living, carelessness in dressing, difficulty with his gait of 6 months' duration, occasional vertigo and diplopia, and incontinence of urine for the previous 4 months.

The patient was studied in the hospital for 2 months, and after a thorough workup the following differential diagnoses were considered: Wernicke's disease, demyelinating disease, and progressive cerebro-cerebellar degeneration. While he was in the hospital it was observed that he improved after a spinal tap and had alternate periods of improvement and worsening. Then because of a rapid worsening of his condition, bilateral carotid arteriograms were done to rule out intracranial lesions. The arteriograms showed anterior bowing and elevation of the anterior cerebral artery, suggestive of hydrocephalus. A pneumoencephalogram demonstrated a clear picture of dilatation of the entire ventricular system. A ventriculo-atrial shunt was then performed and the patient recovered to almost his normal state.

Clinical History

On February 12, 1958, a 52-year-old trombone player was brought to the hospital by his wife with the complaint that for the past year he had shown an impairment of memory and inability to play his instrument as well as he used to; he was less steady in his balance, dull and apathetic, careless in his appearance, stiff in the legs, easily fatigued, unsteady on turns, and unable to climb stairs.

With the onset of this gait difficulty, there was a considerable change in his mental state. He became slow in thought and conversation and at times was confused and exceedingly forgetful. He was untidy in his dress, and his clothes were often soiled because of dribbling of urine. About a month before admission he began to have a tremor of his trunk when walking and needed the help of two canes. He was stiff and remarkably slow in his movements, had a wide-based gait, and at times fell to the floor when he closed his eyes. His wife reported that he had gained weight because he ate well and did nothing but sit around most of the time. He wasn't interested in or worried about anything.

Physical examination showed blood pressure of 140/90, pulse of 108, and respirations of 25. The patient was obese, appeared forgetful and perplexed, and was poorly oriented.

When asked "What is wrong with you?" he answered, "I can't walk, my legs don't obey me." He was incorrect in naming the day and date. He did not remember the names of the doctors who had examined him earlier. He did not remember the name of his musical instrument. Nevertheless, he knew the clef in which the music for his instrument was written. He performed arithmetic calculations slowly and reached the correct answer only after many attempts. It seemed that he had no aphasia; he just seemed to be slow in finding his words. His gait was unstable, slow, broad based with stiffness and weakness of the limbs, and he could walk only with the help of two canes. His speech was slurred. There was definite lateral nystagmus, mainly on right lateral gaze. Both optic discs seemed slightly pale, but the eyegrounds were otherwise normal. The pupils reacted to light, the left normally, the right slightly sluggishly. Visual acuity was 20/25 in the right eye and 20/20 in the left. Color vision was normal in both eyes. Ocular motility was normal. There was no apparent strabismus or diplopia. His face moved symmetrically. Hearing was normal. The rest of the cranial nerves were intact.

There was a terminal tremor on the finger-to-nose test, more pronounced on the left side. Sensation to touch, pin prick, vibration, and position sense seemed to

be intact in all four limbs. There was weakness with spasticity in both lower limbs; they were stiff to passive manipulation. There was a definite ataxia that increased when the patient was ordered to perform repetitive movements. The tendon reflexes were much more active in the lower limbs than in the upper ones. There were bilateral sustained ankle clonus and extensor plantar responses. Chest X-rays revealed pulmonary emphysema.

On March 14, 1958, skull X-rays showed the pineal body to be calcified and in midline. There was slight thinning of the posterior clinoid processes, but the examination was otherwise normal.

An EEG showed a generalized nonspecific slowing of the electrocencephalogram with some alpha waves.

A lumbar tap was done with the patient in the lateral recumbent position. Initial pressure was 180. The fluid contained 6 lymphocytes, 15 mg% total protein, 77 mg% glucose, 407 mg chlorides. Cultures were negative. Hinton reaction, Wassermann reaction, and cardiolipins were negative. The blood sugar was 94.5 mg%, hemoglobin 15 gm, sedimentation rate 3 mm in one hour, hematocrit 50% and WBC 8,000 with 60% polymorphonuclear neutrophils, 2% eosinophils, 0 basophils, 35% lymphocytes, and 3% monocytes. Cholesterol was 188 mg, and non-protein nitrogen was 23 mg%.

After the spinal tap the patient seemed to improve. His wife noticed that his answers were quicker, he was less forgetful, and his gait was faster and somewhat better coordinated. He remained in the hospital for further study with the tentative diagnoses of Wernicke's desease, demyelinating process, or a progressive degenerative disease.

He remained in the same condition for about a month and had two more spinal taps with slight but definite improvement after each one.

On March 18, 1958, lumbar puncture showed an initial pressure of 160, and the fluid contained 10 lymphocytes and 40 mg% total protein. There was good movement of the manometer column with the patient's pulse and breathing.

Two days later, March 20, another lumbar tap was done; initial pressure was 170, and the protein was 25 mg%. There were 18 lymphocytes, but the culture was negative.

On March 24, 1958, the patient was definitely worse. His breathing was stertorous and apneic. He was cyanotic and stuporous. Both pupils reacted sluggishly to light; right pupil was 1.5 mm and the left 2.5 mm in diameter. Temperature was 37.8. There were some generalized rales over both lungs. Blood pressure rose to 170/110 in the morning. There was need to suction some saliva and mucus from his trachea on several occasions; pulse rate was 60. There was retention of urine (14 hours without voiding). On palpation, the urinary bladder was found to be near the umbilicus. A Foley catheter was inserted, and 850 cc of urine were obtained.

Because of the patient's history of mental and neurological deterioration, with alternating periods of improvement and worsening and a respiratory infection, it was decided to do a carotid arteriogram to rule out the possibility of a chronic subdural hematoma. A bilateral percutaneous carotid arteriogram was done under local anesthesia. The A-P films showed that the anterior cerebral artery was in the midline but there were no avascular zones indicative of a subdural collection of fluid. The lateral view showed considerable bowing and upward displacement of the anterior cerebral artery (Figure 2, A).
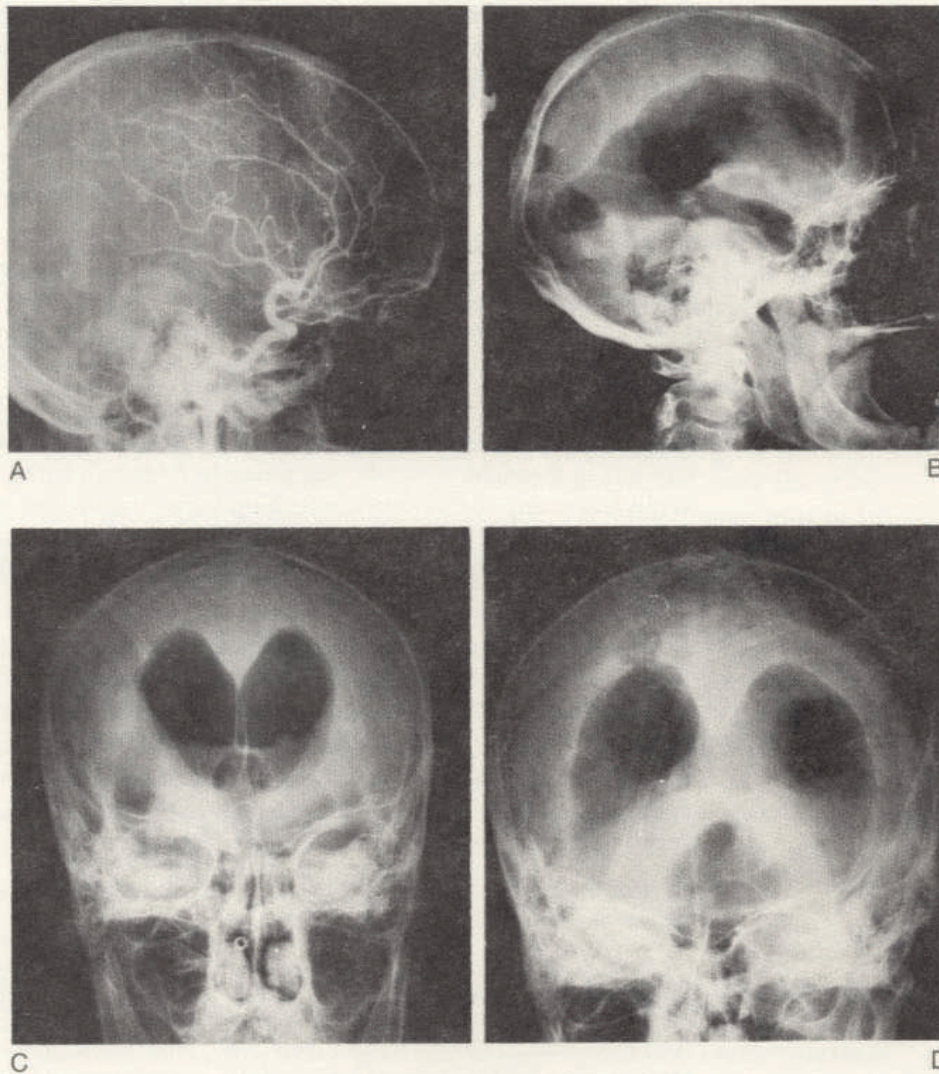
Figure 2. Case 2. A — Carotid arteriogram showing stretching of the anterior cerebral artery, consistent with hydrocephalus. B, C, and D — Air studies confirming hydrocephalus. The dilatation of the fourth ventricle can be seen in B and D.

On March 26, 1958, the patient became duller. There was a proptosis on the left side, and this eye deviated outwardly. There were bilateral Babinskis. Blood pressure was 160/100, pulse 58.

Laboratory findings: Creatinine 1.8, NPN 44.3 mg%, hemoglobin 16.5 gm, hematocrit 55, WBC 9,250, polymorphonuclears 75%, eosinophils 0, basophils 0, and lymphocytes 24%, urine pH 4.5, specific gravity 1.015, albumin 1+. Prothrombin time was 14 seconds.

At 8 a.m., on March 28, 1958, a right occipital ventriculogram and a lumbar pneumoencephalogram were done under local anesthesia. A total of 215 cc of C.S.F. were withdrawn and replaced by air. Films were taken in A-P, P-A, lateral, and facedown positions. The X-rays showed generalized enlargement of the ventricular system (Figure 2, B, C, and D).

There was no air over the convexity of the brain. A number 10 French catheter was left in the right ventricle, according to Poppen's technique, in order (a) to avoid worsening the patient's condition by increasing the intraventricular pressure as a result of air dilatation and (b) to let the air escape quickly. Thus it would be possible to perform the shunt as soon as X-rays showed an absence of air.

At 3 p.m. on the same day, the patient was much better. His temperature was 37°C, and he understood and followed simple commands. The left plantar was extensor, the right indifferent. The left eye moved conjugately. The left palpebral fissure was smaller than the right, and the left pupil was larger than the right. Since 8 a.m., 25 cc of C.S.F. had drained into the ventricular bag connected to the catheter. At 8 p.m., 72 cc of C.S.F. were removed from the ventricular bag. Blood pressure 110/60, pulse 80.

At 4 a.m. on March 29, 1958, 66 cc of C.S.F. mixed with air were removed. Blood pressure 110/70, pulse 86, temperature 37°C.

At 3:20 p.m. on the same day, 27 cc of C.S.F. and a smaller quantity of air were removed. Blood pressure 120/70, pulse 120, temperature 37.5°C.

At 11:30 p.m., 11 cc of C.S.F. were removed. Blood pressure 130/60, pulse 120, temperature 38°C. The patient was fully conscious and lying quietly in bed.

At 4 a.m. on March 30, 1958, 8 cc of C.S.F. with foam and a fair amount of air were removed. Then at 8 a.m., X-rays of the skull were done, and these revealed a bit of air in the frontal horns of the ventricles. It was then decided to do a ventriculo-atrial shunt.

At 10 a.m. a ventriculo-atrial shunt was performed under local anesthesia, utilizing a Hakim valve and the same occipital burr hole used in the ventriculography. The catheter was replaced by a new one.

The next day, there was a remarkable change in the patient. The anisocoria had cleared up. Eye movements were completely conjugated. The left plantar was still extensor, the right flexor. Ankle and knee jerks were 3 plus. Blood pressure was 130/70, pulse 90, temperature 37°C. When we entered the room, the patient said: "Doctor, do you think I shall walk again?" On examination there were some nystagmoid jerks on lateral gaze on both sides. He correctly named five objects which were shown him. He said his name clearly and knew the name of his musical instrument.

On April 2, 1958, the patient was more alert and responsive than at any time since his entrance to the hospital. He was less confused and better oriented in

time and place. He moved his legs more easily. Ankle and knee jerks were 2 plus. Left plantar reflex O, right flexor. Blood pressure 120/70, pulse 95, temperature 37°C. The patient still had an indwelling urethral catheter.

April 3, 1958. The patient asked for permission to stand up and did so without help. He walked with a broad base still using a cane, but his gait was fairly satisfactory. The urinary catheter was removed at the patient's request.

On April 4, 1958, the patient's mental state was almost normal. He had improved so much in such a short time that he was feeding himself and walking with a cane without help. He still had some dribbling of urine. The sutures had not yet been removed.

On April 6, 1958, another spinal tap was done. Initial pressure was 70. No fluid was removed. The patient continued his improvement both neurologically and mentally. He was coherent and well oriented. He seemed brighter and followed the examiner's questions more attentively. He remembered more about his illness than he had before. He now dressed more carefully, in contrast to his previous carelessness in clothes and appearance. He was worried about his economic and family situation. He spoke well and articulated his words clearly. His sutures were removed. Examination did not reveal any diplopia or nystagmus; there was still some stiffness of his lower limbs on passive motion; reflexes were normal and both plantars were flexor. There was some retraction of both Achilles tendons, which did not allow full dorsiflexion of the feet. He was no longer incontinent. He was transferred to the Physiotherapy Department.

On July 13, 1958, his mental status was entirely satisfactory, but he still walked with the help of a cane.

Comment

This case has no apparent history of trauma and the nature of the hydrocephalus is not clear. In a slow and progressive manner the patient began to show impairment in his gait, which gradually became spastic and ataxic (ataxia of gait or Bruns' frontal ataxia) and reflected a certain degree of high pyramidal dysfunction. At the beginning of his symptomatology it was limited to gait disturbances without involving the upper limbs. Gradually, a lack of sphincter control and great r gait incoordination were added to the patient's mental decline.

These symptoms, which simulated a confusional state or dementia, were the grounds for thinking that this was a degenerative or demyelinizing disease. The most important point of the present case is the coexistence of these symptoms with normal C.S.F. pressure, and for this reason we included him with the other cases reported here. In none of the four lumbar punctures did the C.S.F. pressure exceed 180 mm of water, and after each puncture the patient showed a temporary improvement. Finally, the almost full recovery of the patient was obtained when, by means of the ventriculo-atrial shunt, the C.S.F. pressure was reduced and sustained below normal.

Case 3. A 43-year-old patient suffered a cranial trauma with fracture and depression of the right parietotemporal region. There was immediate coma, pinpoint pupils and decerebrate rigidity. After the depressed fragment was corrected, the patient came out of his coma and remained the same for a month with fluctuations in consciousness, limb movements and lack of sphincter control. An arteriogram made two months after the accident showed hydrocephalus. The C.S.F. pressure never exceeded 170 mm of water. With each lumbar puncture and with the extraction of C.S.F. a progressive improvement was observed that lasted for several days.

A ventriculo-atrial shunt produced permanent improvement.

History

This 43-year-old patient was in good health when on July 7, 1961, while driving a jeep, he crashed into a bus. He was thrown out of the jeep and lost consciousness immediately. No medical attention was available until 4 hours later, when he arrived at the City Hospital by helicopter. On arrival at the hospital he was in deep coma, both pupils were miotic, and there was no response to painful stimuli. Decerebrate rigidity was present, and both plantar reflexes were extensor. Otorrhea was present bilaterally and, in addition, blood was oozing from the right ear. There were a bruise of the right scalp and a depressed skull fracture in the right temporoparietal region. Blood pressure was only 40 systolic, pulse was extremely rapid, and respirations were fast and shallow. He was treated with I.V. fluids, blood, nirethamide (Coramine), steroids and antibiotics, and a tracheotomy. After this his general condition improved, and his blood pressure gradually rose to 80/40.

On the following day, July 8, a right parietal burr hole was performed. There was no blood overlying the dura when the depressed bone fragment was elevated. There was immediate improvement postoperatively. His pupils reacted to light, and he moved all of his extremities. A right upper and lower peripheral facial paresis and a mild left hemiparesis were present. When the suction tube was introduced into the patient's trachea, he coughed. Blood pressure was 10 systolic, 7 diastolic; pulse 120; temperature 35.5°C. To prevent seizures, 10 centigrams of diphenylhydantoin sodium (Epamin) were given through the feeding tube every 12 hours.

During the following days (July 9 through 20), he remained in a fluctuating semicomatose state with inequality of pupils (the left larger than right), left hemiparesis, weakness of the right leg, and speechlessness. Bilateral Babinskis were present all of the time. He was treated with oxygen and antibiotics and was tube fed. The tracheal tube was removed on July 15. During this period, temperature rose to as high as 41°C, and there were some transitory rises in blood pressure, systolic up to 160, diastolic to 90.

From July 20 on, his temperature was normal. He moved all four limbs, opened his eyes, looked around, but did not speak and did not follow any commands, no matter how simple they were. Most of the time he slept but would open his eyes when touched or stimulated with noise. When pinched he would open his mouth but would not complain or make any sound. He took food offered by his wife, but at times he refused it and would not open his mouth or would leave the food in his mouth without chewing it.

On July 25, 1961, the patient was in about the same state — afebrile and incontinent. He seemed to be conscious, would open his eyes and look around, and seemed to be rather alert, as if he were impressed, astonished, or perplexed about something or someone in front of him. He was completely mute. He did

not answer any question. If he were left alone, he immediately would fall into profound sleep. Eyegrounds were within normal limits. There was a right total facial paresis. There was slight nuchal rigidity. Deep tendon reflexes were 3 plus on the left and 2 plus on the right. There were bilateral Babinskis. He withdrew the limb when a pin was applied. For the first time during his illness since the accident, a spinal tap was done. Initial pressure was 160. 12 cc of clear and colorless C.S.F. were removed. Final pressure was 140. The fluid contained 40 lymphocytes, a total protein of 35 mg%, and 45 mg% sugar.

The day after this spinal tap, the patient began to speak a few words, pronouncing them slowly. He obeyed and followed such simple commands as "open your eyes" and "stick out your tongue." He still had to be fed and was incontinent of urine.

On July 28, 1961, he was much brighter. He remembered the name of some of his nearest relatives and, in reply to questions, correctly gave the name of the President of the United States, his own date of birth, and the sum of 8 plus 5 plus 4. He was in control of his sphincters.

On August 1, 1961, he began to talk more adequately and spontaneously. He sat up in a chair. He was still disoriented and confused, and his memory was poor. The right facial paresis was still present. The right eye deviated upward and to the side.

On August 9, 1961, he began to walk with a broad-based gait, leaning slightly toward the left. Otherwise his gait was well coordinated. Deep reflexes were about equal (two plus); the left were slightly more active than the right. Plantar response was flexor bilaterally. He ate very well and had gained some weight. Since the accident, he had received 10 centigrams of diphenylhydantoin sodium (Epamin) twice a day.

On August 11, 1961, his sense of orientation and his memory were improved, but on several occasions he was still rather incoherent. He walked about the room most of the time.

On August 15, 1961, he presented a crisis of anguish followed by tears. He was put on meprobamate (Equanil), 400 mg twice a day, for relief of this anxiety.

On August 24, 1961, eyegrounds, sensation, and coordination were normal. Deep tendon reflexes were active and equal, 3 plus. There was a questionable grasp-and-suck reflex. He was confused and slightly dull and apathetic. Again he was disoriented and did not know where he was and could not give the names of his children. Memory was markedly impaired but mainly for recent events. Dextroamphetamine sulfate (Dexedrine), 5 mg twice a day, was added to his other medications.

On August 26, 1961, his mental state was about the same. He did not initiate any conversation, was apathetic, and did not want to get out of bed. He was incontinent of urine. Eyegrounds were normal and the right side of his face was about the same. The patient refused to take medication.

On September 2, 1961, he was completely mute and showed very poor cooperation with the examiner. Attention span was very superficial and unstable. There was a global diminution of all intellectual functions. He didn't answer any questions. There was a total lack of initiative. Nevertheless, he followed objects with his eyes, showing nystagmoid jerks bilaterally on lateral gaze; pupils were equal and reacted normally to light; optic discs were normal. The rest of the cranial nerves were within normal limits. There were bilateral Babinskis;

deep tendon reflexes were symmetrical but more active in the lower than in the upper limbs. Grasp-and-suck reflexes were definitely present. There was a slight nuchal rigidity. Vital signs were normal. Blood pressure was 120/90, pulse 68, respirations 20.

The reduction of his consciousness, the presence of bilateral Babinskis with nystagmus, and incontinence of urine and feces were indicative of a worsening of his neurological condition as a consequence of some intracranial pathology. It was then decided to rule out a chronic subdural hematoma.

At 9 p.m. on September 7, 1961 (two months after the accident), bilateral carotid arteriograms were done under local anesthesia. Blood pressure was 110/70, pulse 90, respirations 20.

Arteriograms made in the A-P position showed the anterior cerebral artery to be in midline. There was no subdural hemorrhage. The lateral film showed a clear picture of hydrocephalus (Figure 3, A and B).

This finding, with normal optic discs, was followed by a lumbar tap. With the patient in a recumbent lateral position, 20 cc of clear, colorless C.S.F. were removed with a 20-gauge needle under an initial pressure of 170; final pressure after the removal of the fluid was 100. The fluid contained 3 lymphocytes, 60 mg of total protein, and 52 mg of glucose; the Pandy was 1 plus; serology was negative.

On the day after this procedure (September 8) there was a remarkable improvement in the patient's state of consciousness. He had breakfast, was again talkative and answered questions, but was markedly confused. He continued to be incontinent of urine and remained in bed.

On September 9, 1961, another spinal tap was done and 15 cc of normal-appearing C.S.F. were removed with a number 20 needle. Initial pressure was 180, final pressure 110. The patient again showed improvement after this tap, and, because of this, another tap was done with an 18-gauge needle to create a fistula between the subarachnoid space and the epidural space and to keep the spinal fluid pressure down. Laboratory examination of this fluid showed 3 lymphocytes, 44 mg% protein, and 50 mg% glucose; Pandy was negative.

On September 10, 1961, the patient continued to improve, ate very well, and fed himself. During the night he was incontinent twice for urine and once for feces. Blood pressure was 110/80, pulse 90, and temperature 36.4°C.

A spinal tap showed an initial pressure of 120. The column of the manometer moved very nicely with his pulse rate and breathing.

Twenty cc of clear C.S.F. were removed. Final pressure was 70. At this time a ventriculo-atrial shunt was proposed in order to maintain the C.S.F. pressure at a lower level, but the family did not consent to this procedure, and we continued to treat the patient for a month with repeated lumbar taps.

September 11, 12, and 13, 1961. The patient was walking all around the ward. His gait was normal but slightly shifted toward the left. Plantars were bilaterally flexor. Pupils were normal. There was no nystagmus. The right facial paresis remained unchanged. His memory was poor and he was confused.

September 14, 1961. Another spinal tap showed initial pressure of 70, and after the removal of 13 cc of normal-appearing C.S.F. the pressure fell to 60.

On September 19, 1961, the patient was alert and communicative. His speech
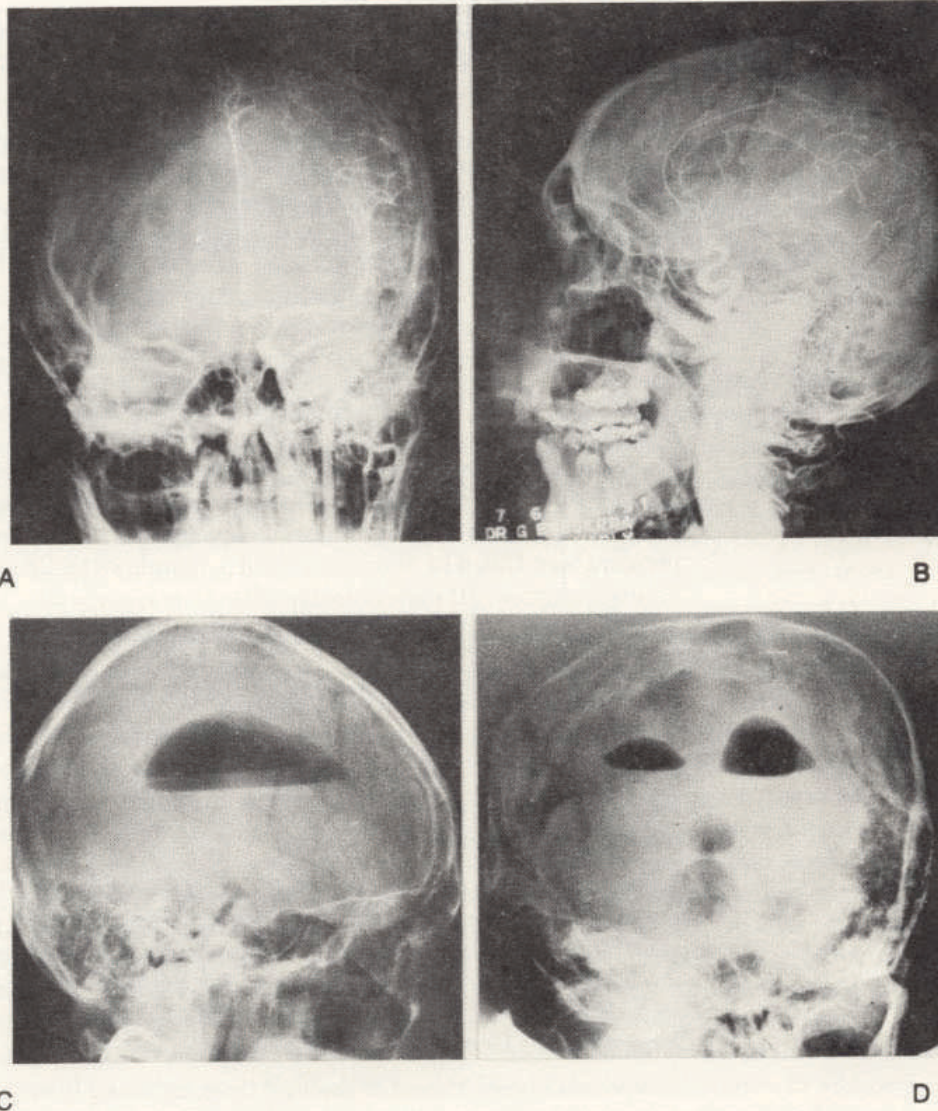
Figure 3. Case 3. A — Carotid arteriogram, anteroposterior view, showing normal vessels that rule out the presence of subdural hematomas. B — Arteriogram, lateral view, showing the elevation and stretching of the anterior cerebral artery, consistent with hydrocephalus. C and D — Pneumoencephalograms done two weeks after the arteriogram, showing the dilatation of the whole ventricular system including the fourth ventricle.

had improved in speed and pronounciation. He was able to give the names of 10 objects and to read and write without mistakes. He was excited and in general euphoric and pleasant.

On September 22, 1961, a progressive improvement was noted in his mental and neurological symptoms. His gait was adequate, well balanced, and well coordinated. His language was normal, he correctly wrote what was dictated to him, and he read satisfactorily with some pauses after each word. He started each paragraph by repeating the first word. He obeyed rather complicated orders and was able to do simple calculations and to explain the meaning of some of the popular proverbs. He named more objects than during the previous examination. However, he was loquacious, somewhat agitated, and inhibited.

On October 5, 1961, he did not leave his bed, was somewhat indifferent, and did not attempt to speak or move. During the previous night he had lost control of his urine on two occasions. Blood pressure was 120/80, pulse 80, respirations 20, temperature 36.5°C. Eyegrounds were normal. A spinal tap showed an initial pressure of 170, and 12 cc of normal-appearing C.S.F. were removed. Final pressure was 120. The fluid contained 1 lymphocyte, 28 mg% total protein, 73 mg% glucose, 10 mg% chlorides; Pandy was negative.

October 6, 1961. The patient got out of bed by himself.

October 8, 1961. Lumbar puncture revealed an initial pressure of 130.

October 12, 1961. The patient spent this holiday at home with his family.

October 19, 1961. The gait was again broad based, and he tended to fall to the left. He walked with his head turned to the right as if he had a right-sided nuchal contraction.

Since there was no doubt about the patient's improvement when his C.S.F. pressure was lowered below normal levels, it was believed necessary to perform a ventriculo-atrial shunt in order to maintain the C.S.F. pressure at subnormal levels. His family refused to authorize the performance of this procedure by us, so he was referred early in November 1961 to the Neurology Department of Massachusetts General Hospital in Boston.

When he was seen there for the first time, he could speak slowly but appeared vague and uncertain of himself, was not precise about the details of his personal history, walked with a broad base, and showed a tendency to turn to the left. He had bilateral Babinskis. On November 10, a pneumoencephalogram showed a marked hydrocephalus with the septum pellucidum in the midline. The right temporal horn was larger than the left. The aqueduct of Sylvius plus the third and fourth ventricles were dilated (Figure 3, C and D). Following this procedure, the patient became worse, spoke less, again was incontinent, and had bilateral Babinskis with hyperreflexia on the left side. For several days he did not attempt to rise from bed.

On November 21, 1961, Doctor W. Sweet performed a ventriculo-atrial shunt using a McPherson valve. The procedure was well tolerated, but two days later the patient had chills and a temperature of 39°C. The C.S.F. contained 40 lymphocytes per cubic millimeter. On November 25, he was more active, faster in answering questions, and less drowsy. A lumbar puncture showed a pressure of 170 mm. The patient was still confused, incoherent, and vague in his speech. It was decided to change the valve, and this was done by Dr. R. G. Ojemann.

In January 1962, the C.S.F. pressure was 90 mm and the valve was operating

normally. During the five following months the patient's attention span, mental capacity, initiative, and speech continued to improve. He no longer showed sphincteric incontinency.

In June, he was able to lead an independent life. The neurologic examination was almost normal. The plantar reflexes were flexor. His memory had not recovered fully. Although the patient leads an-almost-normal life, he is not the same imaginative and intelligent man he was before the accident.

Comment

Obviously this patient suffered a severe trauma with contusions and lacerations of the right temporal region that caused unconsciousness and probably subarachnoid bleeding and cerebral edema. As he improved, he entered a state of akinetic mutism and mental change. These symptoms were attributed to irreversible cerebral injury. The patient remained in this condition until the first lumbar puncture, when he began to improve progressively for 15 days. Then he started to relapse to his previous conditions of akinetic mutism and stupor. Arteriography showed hydrocephalus, and the lumbar puncture showed normal C.S.F. pressure. Each time the patient became worse, a lumbar puncture was all that was required to reduce his C.S.F. pressure to subnormal values. It is interesting that in this patient it was possible to observe the syndrome progressing and also regressing. When the first puncture was performed, the patient was in akinetic mutism with paresis of his four limbs. After the lumbar puncture the mutism disappeared, his memory gradually improved, he began to leave his bed and to walk unsteadily with a broad base, his gait improved, and sphincteric control appeared. If some time passed without performing another lumbar puncture, the patient would again show signs of cerebral dysfunction, manifested by gait disorders and lack of sphincteric control; later on, the patient would not leave his bed, his memory would become poorer, his initiative was reduced, and again he entered into akinetic mutism with quadriparesis, most noticeably in the lower limbs. If this condition was allowed to progress, the patient became stuporous and semicomatose. The ventriculo-atrial shunt made his improvement more stable. However, the results are not easy to interpret because, in addition to the hydrocephalus, there probably are other factors of irreversible damage in the cerebral parenchyma, and an infection probably was added with the first ventriculo-atrial shunt. But it seems evident that the secondary symptoms of hydrocephalus were permanently relieved by reducing the C.S.F. pressure below its normal value.

Cases 1 and 3 were the direct results of trauma, and, as will be pointed out later, the appearance of hydrocephalus in this type of condition is well known. It is worth noting that in these cases, despite the presence of normal C.S.F. pressure, the neurological and mental condition of the patients continued to deteriorate. The return to a normal C.S.F. pressure probably is due to the establishment of an equilibrium between the production and absorption of cerebrospinal fluid.

In Case 2, the etiology of the hydrocephalus is not clear. However, as in the other two cases, symptomatic hydrocephalus was combined with normal C.S.F. pressure.

The three cases described were marked not only by the same hydrodynamic characteristics of the C.S.F. but also by gratifying clinical improvement after shunting. It seems, therefore, that in this type of hydrocephalus a drop in C.S.F. pressure below normal is associated with clinical improvement.

Although C.S.F. pressure returned to normal, the condition of each patient continued to deteriorate as though a vicious cycle had been established by the increase in the area of the enlarged ventricles. As the ventricular area increased, the symptoms of C.N.S. dysfunction were perpetuated even though C.S.F. pressure was normal.

Pneumoencephalography in general is associated with an increase in C.S.F. pressure (Cronqvist, Lundberg, and Pontén, 1963), probably because of blockage of the subarachnoid space secondary to expansion of the volume of air (Cushing, 1926; Foley, 1957; Johnson, 1958). These events could explain the clinical deterioration that took place in Cases 1 and 3 following the completion of air studies and the absence of deterioration in Case 2 after ventricular drainage was established with a catheter (Poppen, 1950).

In addition to the three cases described in detail, we have observed others with the same combination of normal C.S.F. pressure and enlarged ventricles. Some of these will be described briefly.

A 16-year-old female exhibited ataxic gait, poor memory, amenorrhea for one year, and hydrocephalus. These symptoms remitted entirely after a ventriculo-atrial shunt was installed.

There were other cases in which only a slight gait disturbance and minor mental changes were found.

Another patient was hospitalized for three months after suffering a subarachnoid hemorrhage. Her symptoms included akinetic mutism, apathy, loss of sphincter control, and absence of initiative.

A patient who had been diagnosed as having Parkinson's disease, but in whom hydrocephalus was found, showed complete remission of symptoms following insertion of a V-A shunt.

A 63-year-old female was seen in March 1959 with unstable gait, loss of memory, slight mental confusion, occasional urinary incontinence, and transient episodes of giddiness. C.S.F. findings were normal with a pressure of 170 mm of water. Fractional pneumoencephalograms revealed hydrocephalus. A V-A shunt was suggested in Bogotá at that time, and it was subsequently performed at Massachusetts General Hospital in Boston. Recovery was complete, and the patient had remained asymptomatic up to the time this report was written, a period of five years.

The clinical picture seen in these patients is well known to anyone who has had

experience with hydrocephalic cases. The production of these syndromes may well be due to the dilatation of the ventricular system. The varied combinations of symptoms are the result of the different degrees of enlargement undergone by the various portions of the ventricular system. In those cases where only early signs of gait disturbance are present, these signs might be the consequence of greater dilatation of the frontal horns (Bruns' frontal lobe ataxia). As the hydrocephalic condition progresses, the structures surrounding the third and fourth ventricles are gradually impaired to the point where the patient begins to show signs of pyramidal tract involvement, endocrine disturbances, memory loss, psychomotor retardation, etc. Thus, the clinical picture of drowsiness, coma, decerebrate rigidity, and ophthalmoplegia, which can be a part of severe hydrocephalus, would be a direct result of force being applied to those regions where these symptoms originate. For instance, drowsiness could relate to the third ventricle, and nystagmus and slight incoordination to the fourth ventricle. Cushing (1912) described a case of acromegaly that was due not to a pituitary gland lesion but to a cerebellar tumor, which in turn produced hydrocephalus and dilatation of the third ventricle.

The points we wish to emphasize in this monograph are:

The occurrence of the hydrocephalic syndrome in the presence of normal C.S.F. pressure, and the remarkable mental and neurological remissions obtained as a result of lowering the C.S.F. pressure below normal levels.

The three cases reported here in detail were chosen in order to illustrate the working hypothesis of how hydrocephalus can be symptomatic in the presence of normal C.S.F. pressure. Cases 1 and 3 had a traumatic origin, while in Case 2 the cause of hydrocephalus was unknown; however, the second case also showed a complete cure after shunting. It was this latter case that suggested the possibility of the existence of similar cases of hydrocephalus of undetermined etiology that had been placed in the hopeless categories of brain atrophy with irreversible encephalopathies. It was our impression that once hydrocephalus had been established in these cases it was maintained by the same basic mechanism in all three patients since shunting, which was performed although C.S.F. pressure was normal, brought on clinical improvement.

These observations indicate that it is vitally important to be able to recognize such cases since proper treatment can provide a complete cure.

In our experience with cases during the past seven years, a group of patients has emerged with a syndrome consisting of hydrocephalus and normal C.S.F. pressure, which produce a progressive dementia characterized by mild memory impairment, inattentiveness, lack of spontaneity, slowness and paucity of thought and action, sporadic urinary incontinence, and slight uncertainty of gait. More advanced cases show severe memory impairment, pronounced unsteadiness of gait (Bruns' frontal lobe ataxia), lack of sphincter control, episodes resembling catatonia or early Parkinsonism, akinetic mutism, apathy, drowsiness, and lethargy. Terminal cases present stupor with grasping and sucking reflexes, positive Babinski's signs, and coma.

The pneumonencephalogram shows dilatation of the ventricular system beyond that seen in organic brain disease. No air is visualized over the convexity of the cerebral hemispheres. It was observed that following this procedure there was a noticeable worsening in the clinical condition of most of these patients.

C.S.F. pressure readings taken in the lumbar region with the patient lying on his

side usually were below 180 mm of water. Intraventricular pressure was measured in a few of these patients at the time the shunt was installed, and it was found to correspond to that obtained by lumbar puncture.

An electroencephalographic study was made in only one patient. It showed an abnormally diffuse record due to nonspecific, generalized slowness with some alpha waves. That is to say, most of the waves that are found in the tracing are of the theta or delta type with very few alpha-type waves. This electroencephalogram is similar to those described by Gastaut (1950) in hydrocephalus cases.

IV Review of the Literature

The development of hydrocephalus after an inflammatory process or subarachnoid bleeding from a ruptured aneurysm, trauma, or head surgery has already been recognized (Bagley, 1928, 1929; King, 1938; Strain and Perlmutter, 1954; Foltz and Ward, 1956; Kibler *et al.*, 1961). It is believed that absorption of cerebrospinal fluid may be impaired by occlusion of the villi by thrombophlebitis of a vein or sinus, or impairment may be due to blockage of the subarachnoid|spaces by an inflammatory reaction that has produced adhesions (Russell, 1949; Aronson, Volk and Epstein, 1955). In some cases, autopsy has failed to reveal the cause of hydrocephalus (King, 1938).

Enlarged ventricles occurring after subarachnoid hemorrhage have been ascribed to the reaction of the meninges to the red cells with consequent fibrosis (Adams and Prawirohardjo, 1959). However, Jackson (1949) believed that the most irritating component of the spilled blood was probably bilirubin or a similar pigment. Blood in the C.S.F. was not always associated with development of hydrocephalus in experimented animals. Thus, in 6 out of 19 young and in only 1 out of 14 adult dogs, moderate dilatation of the ventricular system was observed after repeated injections of homologous blood into the cisterna magna. In adult patients, complete recovery sometimes took place after subarachnoid hemorrhage, but infants were more liable to progressive symptoms, paralleling animal experiments (Bagley, 1929).

Conflicting statements have been made on the basis of autopsy studies. Thus, Strauss, Globus, and Ginsburg (1932) found external hydrocephalus in only 4 of 13 cases and thought this to be an incidental finding. On the other hand, Monitz and Wartman (1938) reported generalized ventricular enlargement due to hemosiderin-containing leptomeningeal adhesions in two cases after head injury. In 53 cases studied at autopsy after subarachnoid hemorrhage, only half of those surviving ten days or longer had fusion of the pia-arachnoid. Hydrocephalus was not a feature in any of these cases (Hammes, 1944).

The occurrence of ventricular dilatation after subarachnoid hemorrhage was also mentioned by Penfield and Cone (1943), Russell (1949), Stehbens (1954), and Walton (1956).

The association of subarachnoid hemorrhage and intracranial hypertension has been noted by Wertheimer and Dechaume (1950). They concluded that subarachnoid bleeding at times produced total blockage of C.S.F. pathways, causing an acute rise in C.S.F. pressure and death. In other cases it caused a partial blockage and hydrocephalus.

Progressive ventricular enlargement has been reported to occur after head trauma without associated subarachnoid bleeding. Thus, King (1938) reported a case in which four consecutive air studies over a period of a year showed progressive dilatation of the ventricular system with deterioration of the patient. Autopsy failed to reveal the cause of the hydrocephalus.

The treatment of hydrocephalus secondary to subarachnoid hemorrhage by means of a shunting procedure (ventriculomastoidotomy) was first reported in 1954 by Strain and Perlmutter. Their patient had a C.S.F. pressure of 440 mm of water at surgery and later showed clinical improvement.

It is not surprising for a shunting procedure to be effective in cases of hydrocephalus associated with increased intracranial pressure. Foltz and Ward (1956) reported on the beneficial effects obtained from ventriculomastoidotomy in 10 cases of hydrocephalus due to subarachnoid hemorrhage. All of these

cases had hydrocephalus with increased C.S.F. pressure except Case 6, in which the C.S.F. pressure was normal and clinical improvement followed ventriculomastoidotomy. This remarkable finding was not elaborated upon and remained unexplained until now. Kibler *et al.* (1961) reported five similar cases, all of which had either increased C.S.F. pressure or increased proteins. In none of these patients were permanent shunt procedures carried out. In three of the cases studied at autopsy, fibrous adhesions were found in the pia-arachnoid of the upper basal cisterns.

Several cases of gait disorders and progressive mental deterioration have been reported by L'Hermitte and Mouzon (1942), Roger, Paillas, and Tamalet (1950), and Lafon, Gros, and Enjalbert (1950). In none of these cases were C.S.F. pressures reported. Studies made at autopsy showed dilatation of the ventricular system and, in some of them, thickening of the arachnoid membrane.

Wertheimer and Dechaume (1950) reported their experience with hydrocephalus in adults. They divided their cases into three groups: (1) hydrocephalus secondary to tumor; (2) hydrocephalus without tumor; and (3) compensated hydrocephalus associated with atrophy. They concluded that there is no difference between the second and the third groups.

From the review of this literature, it is evident that two main categories of hydrocephalus have been reported: one in which increased intracranial pressure was found, and a second, reported in a large heterogenous group, in which the exact C.S.F. pressure was unknown. (Probably many of these cases were patients with hydrocephalus and normal C.S.F. pressure.) *We believe that our cases form a definitely different group not previously classified, with the following three characteristics:*

1. Mental and neurological symptoms related to hydrocephalus.

2. C.S.F. pressure within normal or upper limits of normal.

3. Clinical improvement after reduction and maintenance of C.S.F. pressure below normal levels.

We wish to emphasize the fact that in a group of cases with normal C.S.F. pressure and hydrocephalus, an improvement is obtained by reducing this "normal" pressure by means of repeated lumbar punctures or the performance of a ventriculo-atrial shunt.

Relationship Between Neurological Symptoms and C.S.F. Pressure

The relationship between C.S.F. pressure and C.N.S. disturbances is not delineated clearly in the pertinent literature. Furthermore, many contradictory statements have been made in the past; from these, three groups of opinions can be summarized:

First Group: There are those who believe that an increase in C.S.F. pressure is directly related to the clinical condition of the patient. Magendie (1842) pointed out that pressure exerted over a meningocele sac was transmitted to the fontanelle in children with spina bifida, and that increasing the pressure over this sac resulted in unconsciousness.

Adson and Lillie (1927) recorded C.S.F. pressure in a patient with a malignant glioma and took manometer readings every 15 minutes for five days. They observed that as the C.S.F. pressure increased the patient became restless, confused, and irrational, and sometimes convulsed. Ethelberg and Jensen (1952),

with a single measurement performed by ventricular puncture, found a direct relationship between ventricular pressure and the degree of papilledema and state of alertness of the patient.

Second Group: There are those who believe there is no relationship between C.S.F. pressure and the clinical condition of the patient. Browder and Meyers (1938) — by inducing artificial changes in intracranial pressure and observing the resulting changes in blood pressure, pulse rate, and state of consciousness — came to the conclusion that none of these factors, either together or separately, bore any relationship to the level of C.S.F. pressure. Ryder *et al.* (1953) performed "lumbography", thereby inducing rapid changes in C.S.F. pressure, by adding and withdrawing fluid to and from the lumbar subarachnoid space; they concluded that high C.S.F. pressure did not interfere with cerebral function and that no specific syndrome resulted from either high or low intracranial pressure.

Third Group: A third group of workers believes that C.S.F. pressure and clinical condition are related in some patients but not in others. Guillaume and Janny (1951) recorded intraventricular pressures in patients with various intracranial lesions. Their results indicated a relationship between symptoms and C.S.F. pressure in some adult patients and in children with hydrocephalus. They concluded, however, that clinical science could not be relied upon to determine the presence of high or low C.S.F. pressure.

Lundberg (1960), while monitoring intraventricular pressures, observed clinical signs of increased intracranial pressure in patients with normal intraventricular pressures. He also found patients with severe intracranial hypertension who did not present clinical signs of it. He reported still another group of patients in which there was a correlation between the amount of C.S.F. pressure and the severity of neurological manifestations. He stated that these observations strongly suggested but did not prove the existence of the causal relationship between C.S.F. pressure and cerebral dysfunction symptoms. He also pointed out that this suggestion could be objected to on the grounds that high intraventricular pressures were not always symptomatic and that minor increments in intracranial pressure were sometimes followed by severe disturbances and even respiratory arrest.

It seems clear from the above review of the literature that there is a correlation between C.S.F. pressure and C.N.S. dysfunction in some cases but not in others.

And the pertinent question is: Why? Obviously, many factors may play a role, and they are difficult to evaluate. They include the thickness and elasticity of the cortex cerebri, state of oxygenation, capillary blood pressure, systemic conditions, age, and — most important — a factor that had never been considered before: the size of the ventricles (that is, the area upon which the C.S.F. pressure is acting). As we will see later, it is the size of the ventricles that determines the effect of the C.S.F. pressure upon the central nervous system.

The reports of Browder and Meyers (1938) did not include information regarding ventricular size. Ryder *et al.* (1953), in their studies with "lumbography", produced only rapid variations in pressure rather than the slow changes seen in hydrocephalus. Furthermore, it is possible that the pressure produced by some of the fluid they injected through the lumbar route may not have been transmitted into the ventricular cavities because of a valve mechanism.

It is noteworthy that those cases reported by Guillaume and Janny (1951) in

which symptoms correlated with pressure were mostly in children with hydrocephalus. Some of Lundberg's observations are of great significance. For example, in Case 133 (cerebellar metastasis) slight (30 to 50 mm) increments in pressure caused severe headache, restlessness, and even respiratory arrest. At autopsy, symmetrical dilatation of the ventricles was found. In Case 119 (cerebellar abscess), cessation of ventricular drainage with a pressure increase of 30 to 40 mm was followed by gradual loss of consciousness and then coma, which promptly reversed when drainage was re-established. Case 134 (malignant glioma), on the other hand, presents a striking contrast in that great increments in pressure of up to 100 mm of mercury did not disturb consciousness.

These examples from the literature substantiate our contention that the size (area) of the ventricles, as we have previously stated, is the important factor in determining whether a change in C.S.F. pressure will result in C.N.S. dysfunction. Any given pressure will produce different effects, depending on whether it is applied to a normal or to an enlarged ventricle.

All of these apparently contradictory findings regarding C.S.F. pressure and the production of symptoms can be clarified by analyzing the cases reported herein on the basis of the theory to be submitted in the following chapter.

V Theory and Mechanism of the Hydrocephalic Syndrome with "Normal" C.S.F. Pressure

The clinical improvement shown by our patients paralleled the drop in C.S.F. pressure produced by the shunting procedure. This suggests a direct relationship between the neurological disorder and the C.S.F. pressure since the only effect of the surgical procedure was a lowering of C.S.F. pressure below its previously normal level.

We believe that brain impairment in hydrocephalus with normal C.S.F. pressure can be explained by Pascal's law. First stated 300 years ago, this law says that:

When a pressure is applied on a certain area of a confined liquid, the pressure is transmitted without gain or loss to each similar area within the vessel.

Pascal's law applies to the C.S.F. system because the system is enclosed and the flow (production and absorption) is rather slow. The C.S.F. is contained within the ventricular cavity under a pressure higher than atmospheric. The difference between the two constitutes the C.S.F. pressure, which is maintained by the balance between production and absorption.

C.S.F. pressure can also be defined as the pressure that must be exerted in order to counteract the flow of C.S.F. through a needle placed in the subarachnoid space or the ventricular cavity (Davson, 1956). Therefore, a C.S.F. pressure of 120 mm of water means that each square mm of ventricular surface is subjected to the weight of a column of water 120 mm high. Pressure is force per unit area ($P=F/A$), and force is pressure times area ($F=PxA$). If ventricles of two different sizes, A_1 and A_2 (Figure 4), are under the same pressure, increasing the area of A_2 will obviously result in a greater force (F_2) being applied to this ventricle ($F_1 < F_2$). *The increase in force resulting from the enlargement of the ventricles could be designated the "hydraulic press" effect of hydrocephalus.*

This greater force (F_2) is the one responsible for the maintenance of hydrocephalus. Thus the hydraulic press effect is responsible for the perpetuation of hydrocephalus in the presence of normal C.S.F. pressure, but not for its pathogenesis. The development of hydrocephalus is based on different principles.

To illustrate the application of Pascal's law, the ventricular system can be represented as a rubber balloon filled with water, inasmuch as the ventricular cavity can expand and enlarge (see Figure 5). The C.S.F. is contained, at least in part, within an elastic vessel that can have different sizes. When a liquid is confined in an elastic and spherical container, the Pascal principle also applies; but when there is an additional factor (in this case, the elasticity of the rubber), it has to be expressed in terms of the relation established by LaPlace: $P=2T/r$. In other words, pressure, P, is directly related to the elastic tension of the sphere and inversely proportional to its radius. If this equation ($P=2T/r$) is carefully analyzed, we can see that it is Pascal's law applied to special conditions: an elastic and spherical container in which the force generated within the sphere by the components —*pressure and area* — must be balanced by another opposing force, *the elastic tension.*

To facilitate the understanding of C.S.F. hydrodynamics when hydrocephalus occurs, we have tried to establish an analogy with the physics involved when a rubber balloon is inflated.

A series of photographs were taken while a rubber balloon was inflated with a manometric capsule similar to the one used in the blood pressure cuff apparatus. A photo was taken every time the pressure changed by 10 mm of Hg. It must be observed that as the balloon increases in size, less pressure is required to keep it enlarged in spite of the increased force in its walls due to
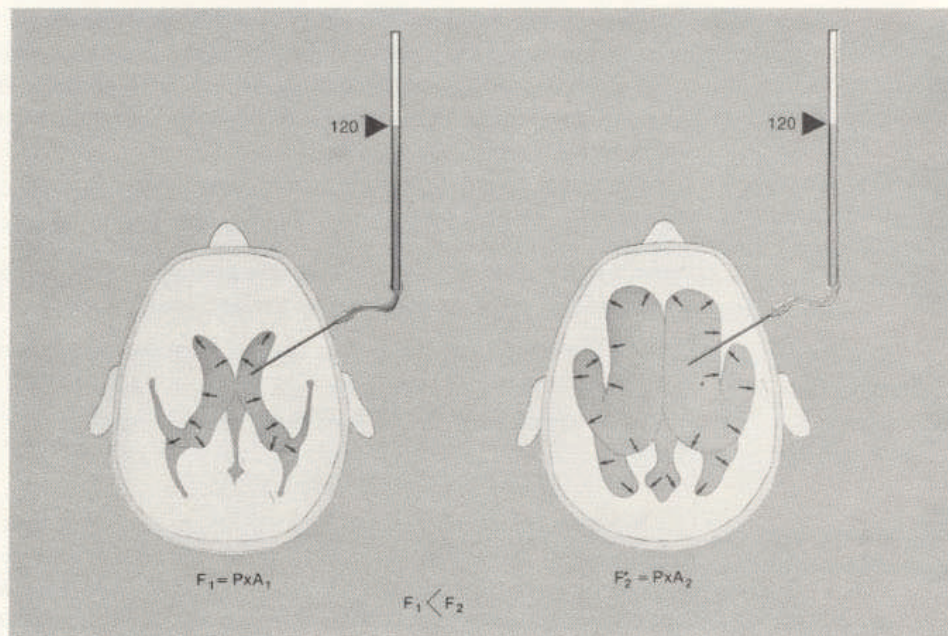
Figure 4. Ventricle enlargement and the "hydraulic press" effect.

In the left drawing, the ventricular system is of normal size and the C.S.F. contained therein is circulating with a pressure estimated to be normal. In this case the hydrodynamics of the C.S.F. are normal.

In the right drawing, the pressure is exactly the same but the ventricle is enormously enlarged. According to Pascal's law — and without considering for the moment the cause for its enlargement — the ventricle, for the simple reason that it has a larger surface, will exert (at "normal" pressure) a force proportional to the area, thus causing an impairment in cerebral function. (See text.)
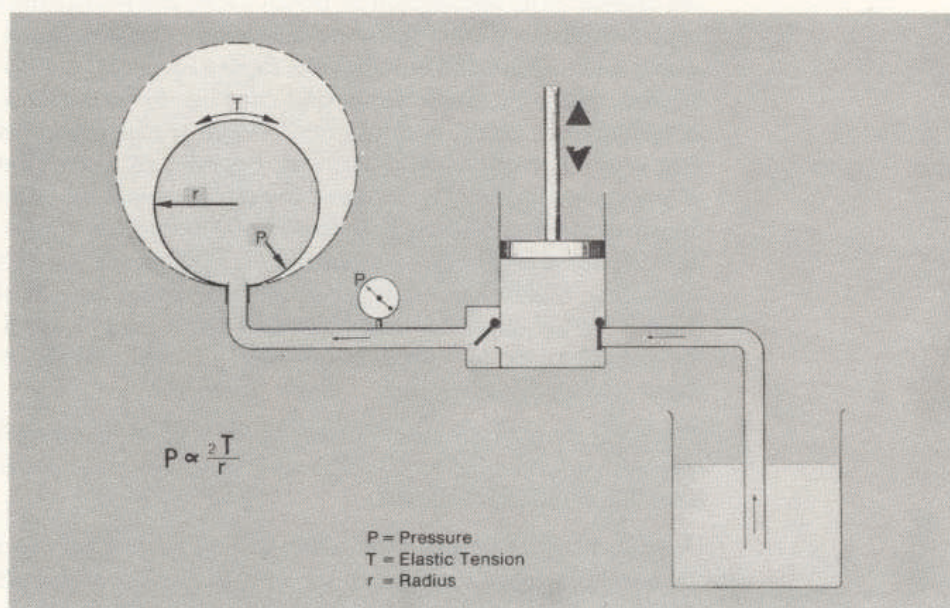
Figure 5. This sketch illustrates the application of Pascal's law to an elastic and spherical container, or the LaPlace ratio: P=2T/r. As the balloon expands, pressure drops.

stretching. (See photos and remarks in Figure 6.)

To show the increase in the elastic tension of the rubber as it is stretched, another set of photos was made of a band of the same rubber in a dynamometer. A picture was taken every time the force applied to the rubber band increased by ½ pound. The different values of elastic tension and length of stretch of the rubber have been plotted in a graph (see Figure 7.) It can be seen that as the rubber is stretched there is more elastic tension. Therefore, the larger the balloon becomes, the more elastic tension there is.

In Figure 8, the two curves obtained from the above two experiments have been superimposed (the pressure on the balloon and the elastic tension in the balloon); also shown are two photographs in which the manometer shows the same pressure but the balloon is of different sizes.

The analogies that we have just submitted may be criticized because the elasticity of rubber is different from that of the brain. In rubber, the elastic tension increases with stretching, and in the brain — where the elasticity is much less — the elastic tension decreases as the ventricular cavity expands. Besides that, the brain is — to a certain extent — encased within its membranes and by the skull, while the balloon is only subject to atmospheric pressure. However, since the cerebral tissue and the ventricles possess a certain elasticity (later on we will see where the elasticity of the brain comes from), these comparisons are acceptable and pertinent.

We are applying the analogy of the balloon only to the ventricular cavity and not to the whole brain. The brain cannot be visualized as a globe with thick walls whose pressure gradients would be between the ventricular system and the subarachnoid space; the reason is that these two pressures (ventricular and subarachnoid) may be equal and may produce hydrocephalus. The pressure gradient is from the ventricular system toward the intracerebral capillary venous system. In other words, the ventricles increase in size by occupying space given up not by the subarachnoid space but by the venous system of the cerebral parenchyma. After the veins have collapsed, the cerebral tissue itself also contributes to the enlargement of the ventricles with the loss of lipids from the white substance (Fishman and Greer, 1963). The elasticity (and compressibility) of the encased brain is derived mainly from its capillary venous bed, which is the first to lose volume and furnish space so that the ventricles can enlarge when there is an increase in the C.S.F. pressure.

It seems that the tensile properties of the brain depend not only on the white substance but also especially on the intraparenchymatous venous bed that gives away first.

These ideas can perhaps be supported by several observations: Masserman (1934) showed that a rapid drainage of a quantity of C.S.F. causes a reduction in the size of the ventricles, which starts immediately after the drainage and lasts for eight hours. We believe that this is possibly due to the engorgement of the veins and cerebral venous capillaries with the lowering of the C.S.F. pressure. That is, the venous system engorges within the cerebral tissue to take over the space given up by the extraction of C.S.F. Foltz and Ward (1956) observed a reduction in the size of the ventricles after a lowering of the C.S.F. pressure in some patients.

We have observed that in cases of acute hydrocephalus, where air studies have shown large ventricular cavities, the corresponding autopsies showed ventricles

Figure 6.
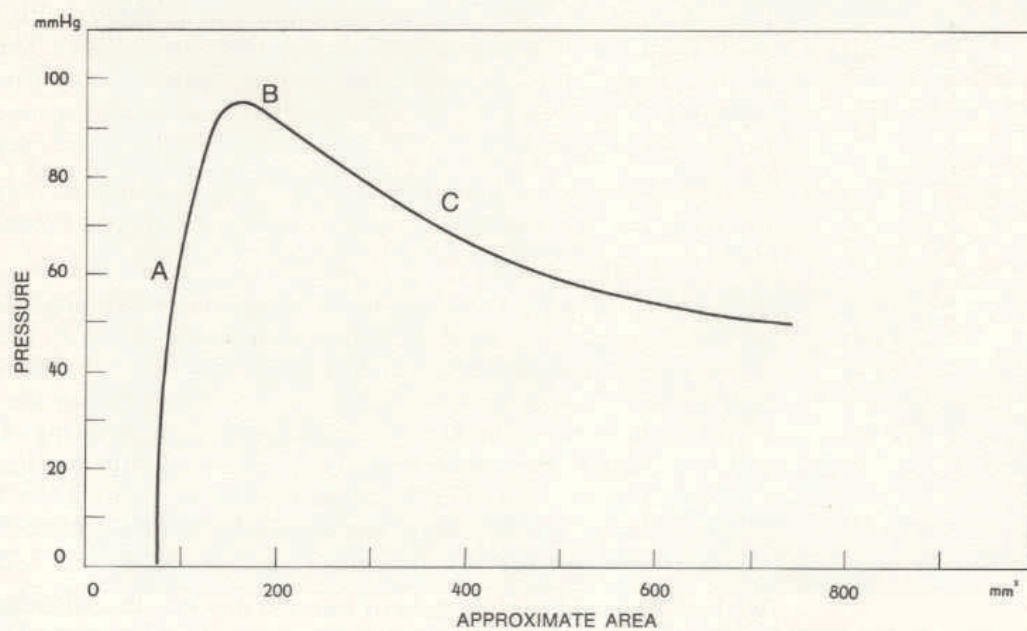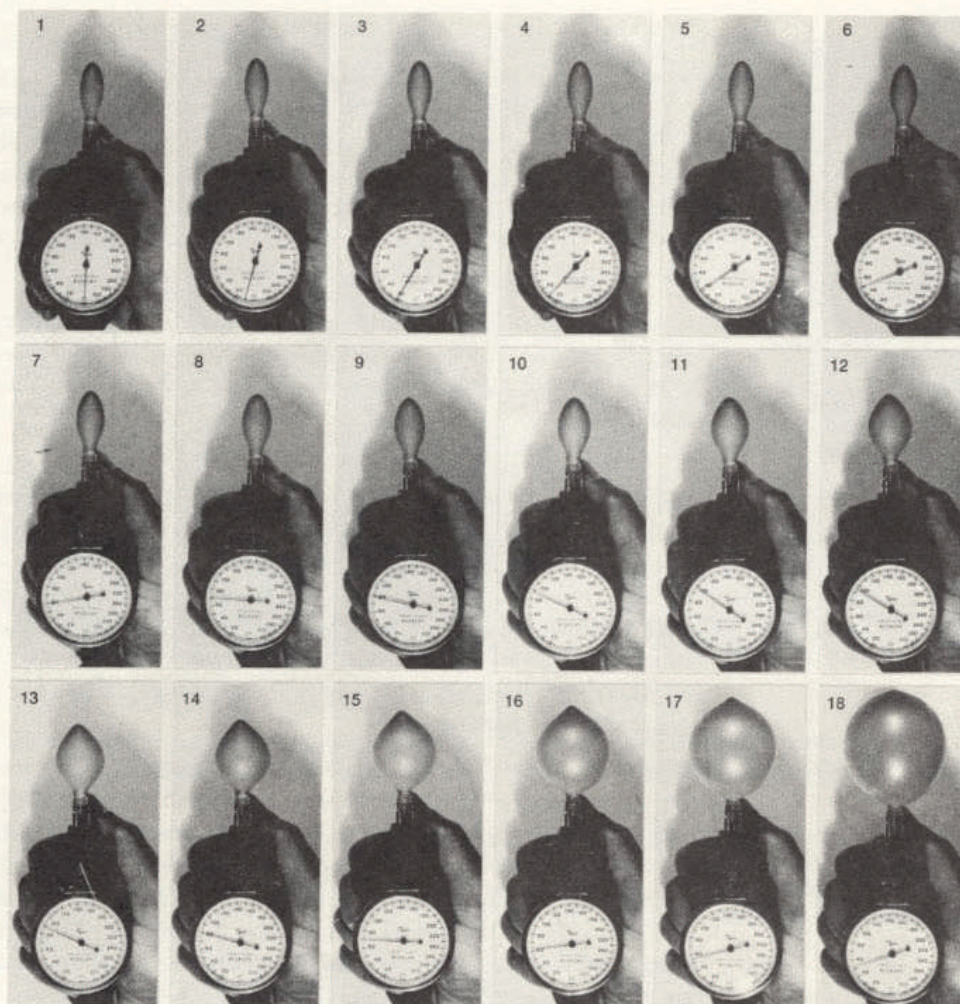
Figure 6. The rubber balloon in this series of photographs presents a rather close analogy of what happens in the cerebral ventricles when hydrocephalus develops. This simple experiment is another illustration of the Pascal principle combined with the LaPlace equation.

To inflate a rubber balloon requires only that the pressure be increased until the initial elastic tension in the balloon walls is overcome; after that, the pressure drops. Pressure does not have to be increased to keep inflating the balloon, even though the elastic tension increases. These relationships of pressure and area, which are grossly similar to those observed in hydrocephalus, are illustrated in this experiment, the results of which have been plotted in the pressure-area curve.

When inflation of the rubber balloon begins, the pressure increases rapidly until the balloon begins to expand; then the pressure drops as the balloon increases in size, in spite of the increase in elastic tension in the balloon walls. This paradox occurs because there are two forces that are maintained in equilibrium and are exerted on all of the balloon surface: externally the elastic tension of the rubber, and internally the total force applied to the balloon inner surface. When the balloon is inflated, the elastic tension and the area increase. The increase in elastic tension is proportional to the stretching of the rubber, and the total force within the balloon is proportional to its area ($F=PA$). Different values of pressure ($P=F/A$) are obtained as the balloon is inflated because of the adjustment made by these two forces in order to maintain the equilibrium and also because their components (area and elastic tension) vary with the size of the balloon. Consequently, the curve corresponds in:

A — to the part in which, because there is no increase in area, pressure rises to overcome the elastic tension.

B — as the balloon starts to enlarge, the pressure remains constant because the area increases at approximately the same proportional rate as the elastic tension increases.

C — as the balloon continues to increase in size, the area increases more rapidly than the elastic tension; therefore, the pressure is reduced in accordance with the LaPlace ratio ($P=2T/r$).

If we compare photos 7 and 16, we see that in the latter the balloon is larger and, therefore, has a greater elastic force; however, the manometer shows the same pressure in the two photos. In photos 9 and 18, the differences in pressure and size are even greater.
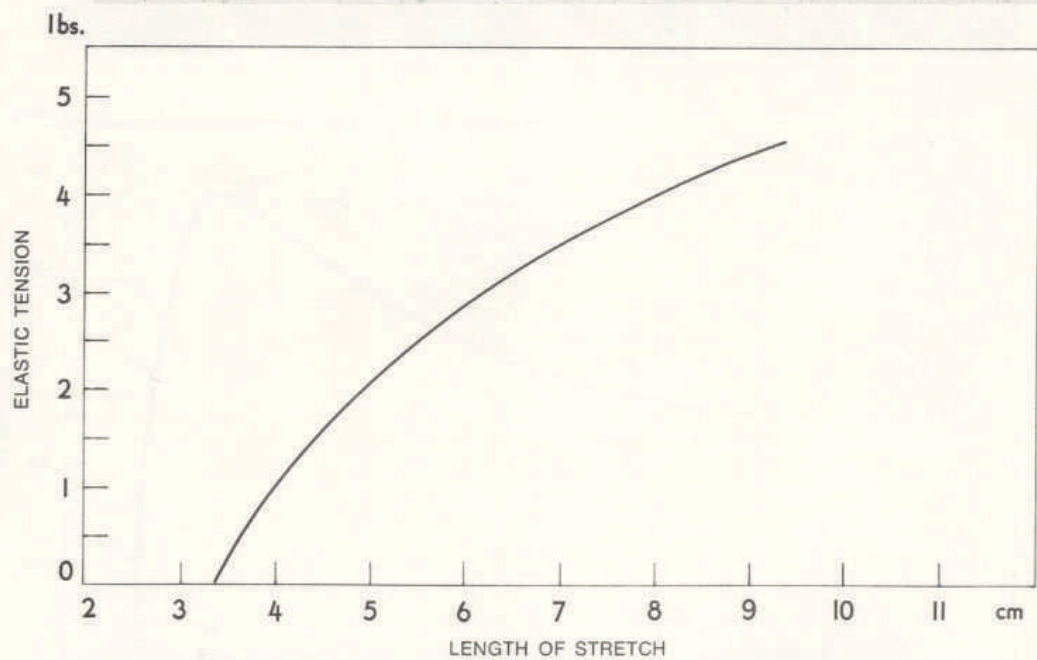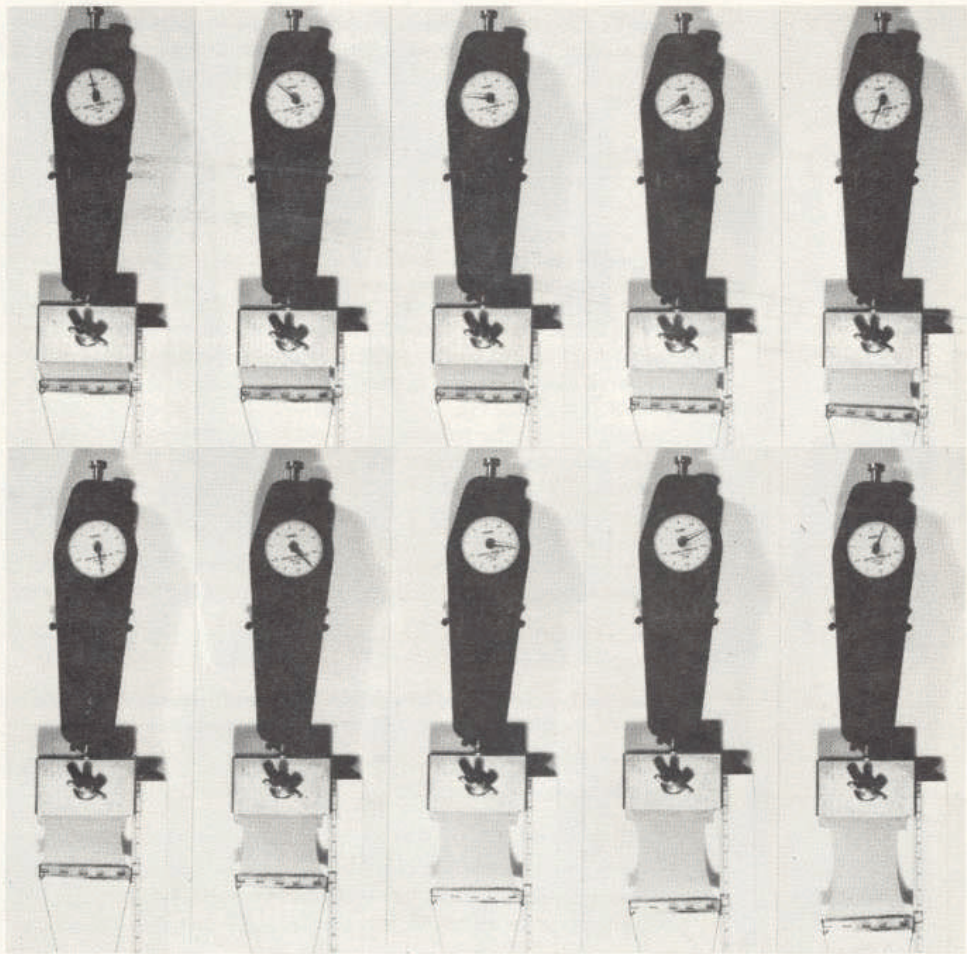
Figure 7. These photographs illustrate the increase in elastic tension of a rubber band during stretching. The graph shows that elastic tension increases as the rubber is stretched.
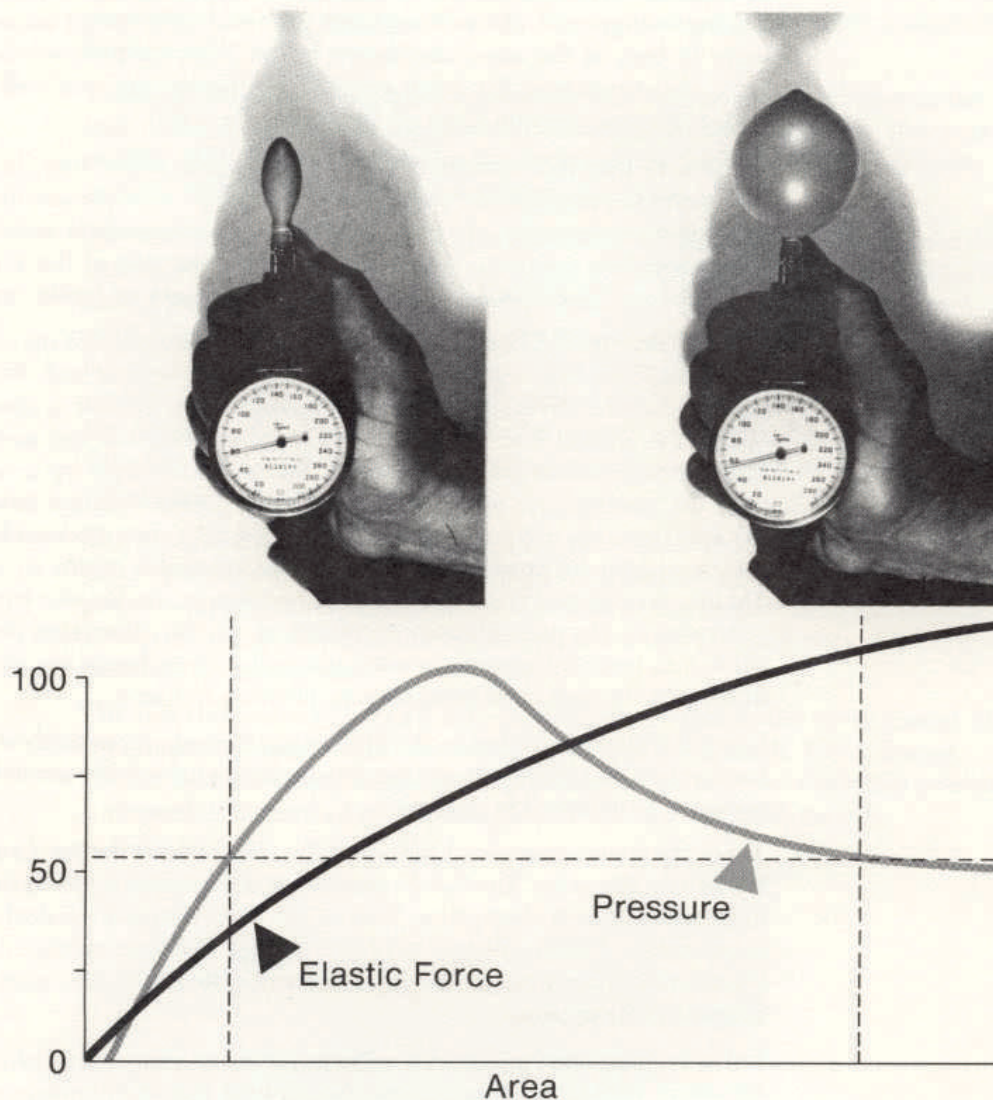
Figure 8. No additional pressure is required to keep the balloon enlarged, in spite of the increased elastic tension. The left photo represents normal C.S.F. hydrodynamics. The right photo shows the analagous hydrodynamics of the hydrocephalic syndrome with "normal" C.S.F. pressure.

Although the pressure is the same in the two rubber balloons, the balloon on the right remains enlarged. The same thing happens in the brain. Even though C.S.F. pressure is normal, it is still too great to allow the ventricular system to return to its normal size.

of a smaller size than seen in the pneumoencephalogram. On the other hand, in long-lasting cases of hydrocephalus, the ventricles found on autopsy are, more or less, of the same size shown in the X-rays. All these observations show that there is in the brain a system or mechanism, not well understood, that produces a certain form of "elasticity."

We believe that the immediate changes produced, either way, in the size of the ventricles through the variation in the C.S.F. pressure are due to the compensating capacity of the reservoir of the intracerebral venous system, which acts like a sponge. The slow changes in the size of the ventricles are produced in the cerebral tissue itself by loss or gain of lipids, water, etc.

The cerebral tissue should be considered as a sponge made up of interconnected cells represented by the capillary venous bed. When the C.S.F. pressure increases, the venous blood of this system is slowly squeezed out and is diluted into the rest of the venous system of the body. Thus, it is only through this venous system that the intracranial cavity is connected with the atmospheric pressure. If the C.S.F. pressure comes down again and the ventricles do not have a greatly enlarged area, the intracerebral venous system regains its previous volume and the ventricles return to normal size. That is, the cerebral tissue is subject to two forces: on the one hand, the force generated by the parenchymatous veins and, on the other, that produced by the C.S.F. pressure against the ventricle walls. These forces are also maintained in balance through their components: pressure and area.

Using the ideas expressed in the previous paragraph, let us analyze the mechanism that leads to symptomatic hydrocephalus with normal C.S.F. pressure, as occurred in our cases.

There are many causes for hydrocephalus, but in each the mechanism is essentially the same. The C.S.F. pressure is determined by the balance between its production and absorption. Two of the cases selected for study in this paper suffered from subarachnoid hemorrhage as a consequence of head injury. Presumably there was a blockage in the absorption of C.S.F. with a consequent increase in its pressure.

Let us suppose that a patient is suffering from an intracranial process that interferes with C.S.F. circulation; also suppose that the conditions are favorable for the development of hydrocephalus. As the intraventricular pressure increases, the ventricles increase in size by occupying space given up by the venous capillary bed of the cerebral parenchyma, particularly that of the white substance. Blood is being squeezed out by the venous capillaries that are being compressed slowly to make room for the increased volume of the ventricular system. If the blockage of the C.S.F. circulation is complete, the intraventricular pressure increases until, after compressing the venous capillary system, it compresses the cerebral parenchyma itself until its functions are paralyzed; it also compresses the brain stem, causing respiratory standstill. Fortunately, this happens rather seldom.

After the interference of the C.S.F. circulation has disappeared (for instance, by reabsorption of the blood), the intraventricular pressure returns to normal but the ventricles remain enlarged and some neurological symptoms persist because, once the ventricles have expanded, they are kept in the same condition by lower pressures than those that produced the initial expansion. They do not return to their previous condition because the venous bed cannot decompress itself since there is more force on the ventricular side due to the increase in area. In order to

balance these systems again, it is necessary to lower the C.S.F. pressure below the normal level by means of a shunt. The sequence of these events is illustrated in Figure 9.

From the review of the literature available on hydrocephalus and also on the role of C.S.F. pressure in the production of neurological symptoms, it becomes evident that, so far, no attention has been paid to the area of the ventricular system, and its importance has not been recognized.

The measurement of the C.S.F. pressure alone, in the presence of a ventricular system with abnormal size, does not supply exact information about the force that is pressing on the ventricular walls. Therefore, it is not possible to state what is the representative figure for normal pressure in C.S.F. because that which is normal for one person may not be for another individual. The three cases described in this paper have illustrated this point. A "normal" pressure in the presence of an expanded ventricular system was responsible for a continuous deterioration of the patients, until this pressure was reduced by means of the shunt procedure.

The acceptance of the importance of the ventricular area and its relation to C.S.F. pressure in generating abnormal forces that act upon nervous tissues may serve to explain the hydrodynamics of C.S.F. in the formation of subarachnoid cysts, porencephalia, infantile hydrocephalus where the skull is expanded, and, possibly, syringomyelia.

The fact that pulse and high C.S.F. pressure have a role in the development of certain pathological conditions cannot be denied (O'Connell, 1953; Bering, 1955). We wish to emphasize the importance of the area on which this pressure is exerted; it is another contributing factor in C.S.F. hydrodynamics.
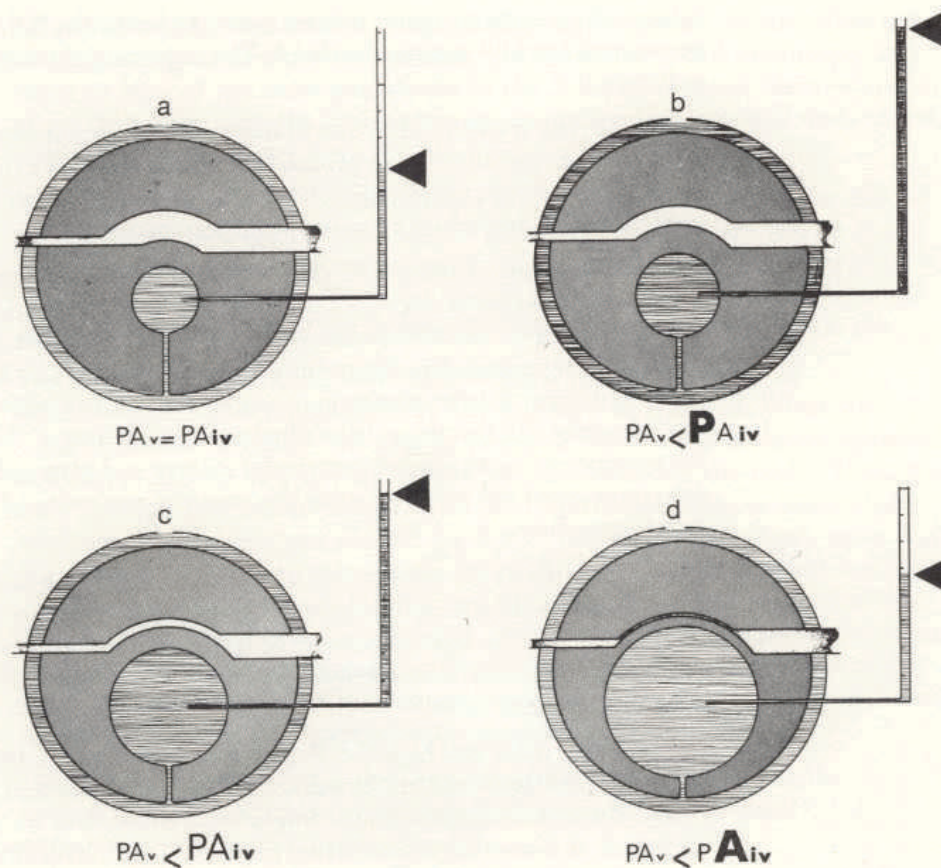
Figure 9. These drawings represent the three factors that affect hydrocephalus:

(1) The C.S.F.; (2) the ventriculo-subarachnoid system; and (3) the venous system, represented by a single large vein. (The arteries are not modified because they are under much higher pressure and are not affected in the same manner.)

a — Under normal conditions, the intraventricular pressure (Piv) and the size of the ventricle (Aiv) are normal. The total force produced by this pressure when acting upon the normal ventricle area is in balance with another equal force generated by the venous pressure (Pv) and the total area of the venous capillaries (Av). The ventricular system neither increases nor decreases in size.

b — When hemorrhage occurs, the C.S.F. pressure increases considerably due to diminished absorption (partial blocking of the arachnoid villi). The ventricle expands and presses against the cerebral tissue. Since this tissue is semi-solid, it is more resistant than the veins, which are the first to yield and collapse, causing symptoms of cerebral dysfunction. The brain also gives way in turn, causing the loss of lipids from the white substance and greater expansion of the ventricle. Due to the increased C.S.F. pressure, an unbalanced condition now exists between the forces of the ventricle and those of the venous side and cerebral tissue.

c — The cerebral ventricle continues to expand because of the imbalance. The C.S.F. pressure begins to come down as the hemorrhage is absorbed.

d — The production and absorption of C.S.F. are balanced again and the pressure has dropped to the "normal" value. This normal pressure, when acting upon the area of an enlarged ventricle, in accordance with Pascal's law, generates enough force against the brain and its venous system to prevent their returning to the conditions that existed prior to the hemorrhage. That is, a new mechanism is now operating to maintain the cerebral dysfunction in the presence of normal C.S.F. pressure. From this it is deduced that the only way to offset the hydraulic press effect of hydrocephalus is to lower the pressure below its normal level.

VI Conclusions

1. Several cases of a hydrocephalic syndrome with normal C.S.F. pressure have been observed and three of them have been reported in detail. The patients showed psychomotor retardation, lack of attention, diminished memory, akinetic mutism, gait disturbances, and urinary incontinence. This picture may be confused with a degenerative or vascular disease or irreversible sequelae of craneoencephalic trauma. A ventriculo-atrial shunt corrected this condition, with improvement of the patients, by lowering the C.S.F. pressure below the "normal" value.

2. A review is made of the literature pertaining to: (a) development of hydrocephalus after a subarachnoid hemorrhage; (b) the relation between C.S.F. pressure and dysfunction manifestations of the C.N.S.

3. The importance of the area of the ventricular system in the production and maintenance of a hydrocephalus syndrome with normal pressure is emphasized. It is concluded that the force exerted on the ventricular system walls is equal to the C.S.F. pressure multiplied by the area of this system. A normal C.S.F. pressure will apply greater force upon an expanded ventricle than upon a normal one. Example: a pressure of 180 mm of H_2O upon an expanded ventricular system may produce symptoms, while this same pressure is innocuous in one of normal size.

4. A theory is submitted for consideration regarding these cases, and new points of view are developed on the physiopathology of the hydrodynamics of the C.S.F. circulation, based on Pascal's law. (See Figure 10.)

5. The hypothesis of the "hydraulic press" should be applied in all hydrocephalus cases, both in adults and children, in which the skull increases in size. This principle probably plays an important role in porencephalia, in postoperative meningeal and subarachnoid cysts, and perhaps in syringomyelia.

Bogotá, March 1964.

Normal

**Hydrocephalus
with Normal C.S.F. Pressure**
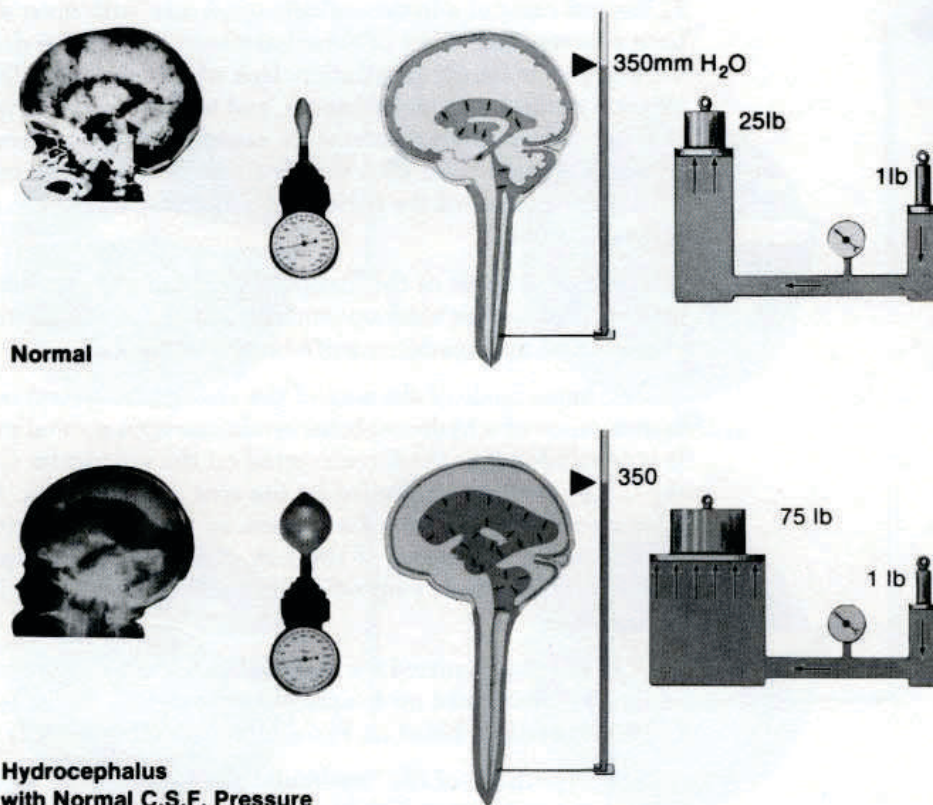
Figure 10. These drawings summarize the theory proposed to explain the hydrocephalus syndrome with "normal" C.S.F. pressure. Shown are C.S.F. hydrodynamics in the normal and hydrocephalic conditions and the principles of physics that apply.

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