University of Toronto Neurosurgical Rounds No. 3.

Normal Pressure Hydrocephalus — High Pressure Normocephalus

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INTRODUCTION

The case history to be presented spans most of the lifetime of a young lady who is now twenty-two years old, and it reflects the stages of development of our knowledge of several important neurosurgical concepts.

Subarachnoid hemorrhage is rare in children, and although successful repair of intracranial aneurysms was common by 1961, it was unusual in a three year old child. Hydrocephalus following subarachnoid hemorrhage had been produced in the laboratory (Bagley, 1928) and reported in sporadic autopsy cases (Strauss, Globus et al., 1932), but it was not until the year before our patient was born, that Foltz and Ward (1956) described ten patients with symptomatic hydrocephalus, several weeks following subarachnoid bleeding. Some of those patients had normal CSF pressure and yet improved dramatically after the insertion of a treacherous but effective ventriculomastoid shunt. The rest of the story of "symptomatic normal pressure hydrocephalus" is well known, told first by Hakim (1964), and named by Adams and Fisher (1965). However, three years before Hakim's paper appeared, our patient developed symptomatic normal pressure hydrocephalus. Her problem was correctly diagnosed and she was treated successfully by a shunting procedure that had been pioneered by Dr. William Keith at Toronto's Hospital for Sick Children since 1952, that is, a lumboperitoneal shunt.

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During the 60's, while our patient was attending school and achieving good marks, Lundberg (1960) pioneered the use of long term ventricular pressure recording, and described various wave forms including plateau or 'A' waves. Shortly afterwards, Langfitt, (1964) further clarified our understanding of raised intracranial pressure (ICP) and related cerebrovascular responses. Dr. Hart Schutz studied with both of these investigators, and subsequently established the first intracranial pressure laboratory in Canada, at Toronto Western Hospital (Schutz, 1976). When our patient developed some rather bizarre symptoms in 1977, it was not a CT scan, but ventricular pressure recording that helped to define the nature of her problem and lead to its successful treatment. Perhaps an analysis of the shape and height of her CSF waves, respiratory waves, or A or B waves, revealed abnormalities in the viscoelastic property of her brain, or abnormalities in her craniospinal compliance, and unlocked the secret of why her brain and spinal fluid behaved as they did.

Measurement of the CSF absorptive capacity by infusing fluid into the sub-arachnoid space was described by Katzman and Hussey (1970). Hope that this test might predict shunt benefit in hydrocephalic patients was not quickly realized. However, recent enthusiasm for measuring what is now called "conductance to outflow" (Borgeson, Gjerris et al., 1979) has produced a promising variety of infusion tests, including not only constant rate infusion tests but also a bolus infusion test, and a variable rate infusion (servo-controlled) constant ICP test (Sklar, Beyer et al., 1978). Dr. Schutz has carefully studied a group of patients suspected of having normal pressure hydrocephalus, by means of a constant rate infusion test, and has been able to draw some useful and important conclusions. It was an abnormal infusion test that shed real light on our patient's problems.

The case raises more questions than it answers. Why do ventricles dilate when CSF absorption fails? Why does the ICP sometimes rise and sometimes not? Is there really such an entity as normal pressure hydrocephalus, or is it intermittent high pressure hydrocephalus, the episodic highs to be found only by 24 or 48 hour monitoring (Symon and Hinzpeter, 1977)?

What about pseudotumor cerebri? Some features of today's case are reminiscent of pseudotumor. Does a CSF absorption defect play a part in its genesis, as has been recently suggested by Sklar, Beyer et al. (1979), or is the abnormality primarily in the brain parenchyma and/or vascular bed?

Ladies and gentlemen, as we listen to the case history, and as we think about compliance of the craniospinal space, CSF outflow conductance, and thevisco-elastic properties of brain tissue, it is fitting that in these University of Toronto Neurosurgical Rounds, we pay tribute to the surgeon who repaired our patient's aneurysm in 1961, and who successfully treated her normal pressure hydrocephalus before that syndrome was ever named, using a shunting technique which he pioneered: innovator, scholar, master technician, teacher and friend, Bill Keith.

CASE HISTORY AND DISCUSSION

On April 15th 1961, a 3½ year old girl presented with headache, vomiting and photophobia. Initially the symptoms were attributed to an upper respiratory infection. Vomiting persisted, and on April 18th she complained of a painful neck and back, and sore

arms. At night she wakened with crying spells. On April 19th she suddenly fell forward and lost consciousness.

She was admitted to the Hospital for Sick Children. She was drowsy, but could be roused. A subhyaloid hemorrhage was seen in the left optic fundus. Bruits were heard over both orbits. The opening pressure at lumbar puncture was 360 mm of CSF, and the fluid was bloody. Angiography demonstrated an aneurysm of the anterior communicating artery, filling from the left side only; the aneurysm was elongated and pointed down and to the right (fig. 1).

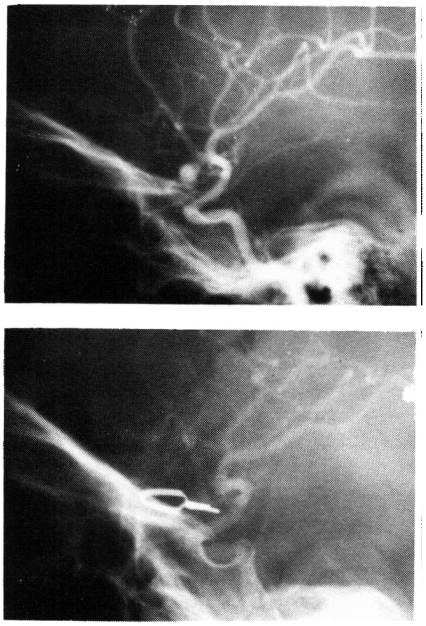

Figure 1 a & b — Cerebral arteriograms showing anterior communicating artery aneurysm before and after surgical repair.

She was operated upon seven days following her first symptoms. Surgical adjuncts included hypothermia, hyperventilation, lumbar CSF drainage, osmotic diuresis with 30% urea, and dissection of both carotid arteries in the neck for proximal control. The aneurysm was approached by the interhemispheric route, and clipped with both a MacKenzie and Mayfield clip. The patient awakened immediately without neurological deficit.

She was well for the first two weeks, but then became irritable, drowsy and developed a stiff neck. Repeated lumbar punctures were done to clear the CSF of blood products. The fluid gradually cleared, CSF protein was 16 mgm % and the pressure was 120 mm of CSF. She continued to vomit, remained listless, and showed lid retraction. Hydrocephalus was suspected. A pneumoencephalogram was attempted, but the air escaped into the subdural space. Ventriculography showed enlargement of the ventricles and a block to the passage of air at the tentorium, confirming the diagnosis of communicating hydrocephalus. A lumboperitoneal shunt was inserted, and she improved dramatically and was discharged home.

Revisions of the shunt were required seven months later and two years later. In January 1964, the lumboperitoneal shunt was removed and a ventriculoatrial shunt was inserted. From then on she developed normally and was at the top of her class throughout her years in school.

She came to medical attention again in July 1977 at age 20. She was admitted to Toronto Western Hospital for investigation of headaches. The headaches were episodic, lasting three to five minutes, and were frequently associated with right sacroiliac pain. They were not related to posture, straining or coughing, but were aggravated by stress and anxiety. They varied from occipital to frontal, and from throbbing to stabbing. She had some emotional and personal problems which raised the possibility that the headaches were functional. The neurological examination was normal. There was no papilledema and the retinal veins were pulsating. The ventriculoatrial shunt assembly was palpable behind the right ear, but there was no pump.

Lumbar puncture showed a pressure of 210 mm of CSF, normal protein concentration, and no cells. CT scan showed normal sized ventricles, minor ventricular asymmetry and a well placed catheter tip (fig. 2). CSF infusion test was done using mock CSF (Elliots "B" solution) infused at a constant rate; although the baseline pressure was normal, infusion at the slowest rate (1.5 ml/min) caused an abnormally high rise in pressure, indicating an abnormality in CSF absorption (fig. 3). A higher infusion rate (2 ml/min) was discontinued because of a dangerously steep rise in pressure. The test result was interpreted with caution because of the possibility of arachnoiditis due to a previous lumboperitoneal shunt and two shunt revisions. Because of the normal sized ventricles on CT scan, and because of the rather vague and possible functional nature of her complaints, it was concluded that she was no longer shunt dependent and she was discharged home from hospital.

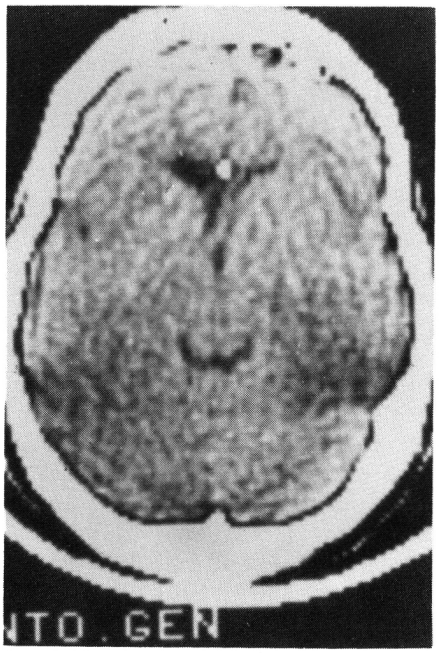

Figure 2 — CT scan showing small ventricles, minor ventricular asymmetry and a well placed ventricular shunt tip.

Her headache diminished and she remained unchanged for three months. In November 1977 her symptoms became more bizarre. Headache was then a minor complaint. She experienced episodes of tingling in all four extremities, arthralgia of various joints, flatulence, sacroiliac pain, abdominal cramps, and periods of anxiety with flushing of the face. During one night she had four episodes, each about five minutes long, during which she would awaken from sleep, become restless and agitated, cry out with pain in the shoulder, knee or hip, complain of double vision, and quickly go back to sleep. As before, we were perplexed by her symptoms. However, her optic disc margins were now blurred. A blocked shunt was now a distinct possibility even though a CT scan again showed small ventricles. It

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was decided to institute continuous ventricular pressure monitoring. When the right frontal burr hole was made, the dura was slack, the subdural space was roomy and the ventricular pressure was subatmospheric. When she returned to the ward, there was a rise in pressure, and high plateau waves appeared frequently, rising almost to systemic arterial pressures, but without producing a Cushing response (fig. 4). Her symptoms were reproduced with each pressure wave. She was able to abort her symptoms and the plateau waves by hyperventilation. There were long periods when the pressure was normal, only to be followed by recurrent pressure waves. As it was then clear that she was shunt dependent, the ventriculoatrial shunt was converted to a ventriculoperitoneal shunt. The lower end of the old shunt was too short and had pulled out of the jugular vein. Since the revision two years ago, she has been well and free of all symptoms.

DISCUSSION (Hart Schutz)

The development of communicating hydrocephalus due to obstruction of the subarachnoid spaces by breakdown products of blood was recognized by Bagley (1928). What was not fully realized at the time of our patient's original illness in 1961 was the paradox that communicating hydrocephalus could be symptomatic in the presence of normal or low intracranial pressure. She deteriorated three weeks after subarachnoid hemorrhage because of communicating hydrocephalus, yet her lumbar pressure was only 120 mm of CSF. Normal pressure hydrocephalus (NPH) as a clinical entity was not described until three years later by Hakim (1964). Hakim and Adams (1965) pointed out that the mechanism of NPH was in accord with Pascal's law for enclosed fluids, namely that the force on the walls of a container is equal to the product of pressure and surface area. They reasoned that the ICP was probably high in the early stages, but once raised pressure had enlarged the ventricles, the enlargement presisted at a lower pressure. Thus, a pressure of 180 mm of CSF is well tolerated in normal sized ventricles but may cause symptoms if the ventricles are enlarged. The mechanisms and pathophysiology of NPH have been controversial, and have been the subject of much research (Adams, Fisher et al. 1965; Appenzeller and Salmon, 1967; McCullough, Harbert et al. 1970; Philippon, Ancri et al. 1971; Salmon, 1972; Salmon and Armittage, 1968). It is agreed that NPH is a fairly common sequel to subarachnoid hemorrhage and infection but idiopathic NPH is rare.

Until recently, radioisotopic cisternography (DiChiro, 1964) was the primary investigation used in selecting patients who might benefit by shunting. It was pioneered and developed by Dr. Ross Fleming at this institution (Fleming, Sheppard et al., 1972). Fleming described both normal and abnormal patterns of isotope circulation. In communicating hydrocephalus, the characteristic finding is ventricular filling and retention of the tracer for 48 hours or more, with little or no subarachnoid filling. Unfortunately, isotope cisternography has limited value in predicting shunt benefit, and the success rate of shunting patients with NPH is only about 60% when isotope cisternography is used for case selection (Black, 1980; Crockard, Hanlon et al., 1976; Symon and Hinzpeter, 1977). Other methods of predicting shunt benefit are the clinical picture (Belloni, Di Rocco et al., 1976; Heinz, Davis et al., 1970;

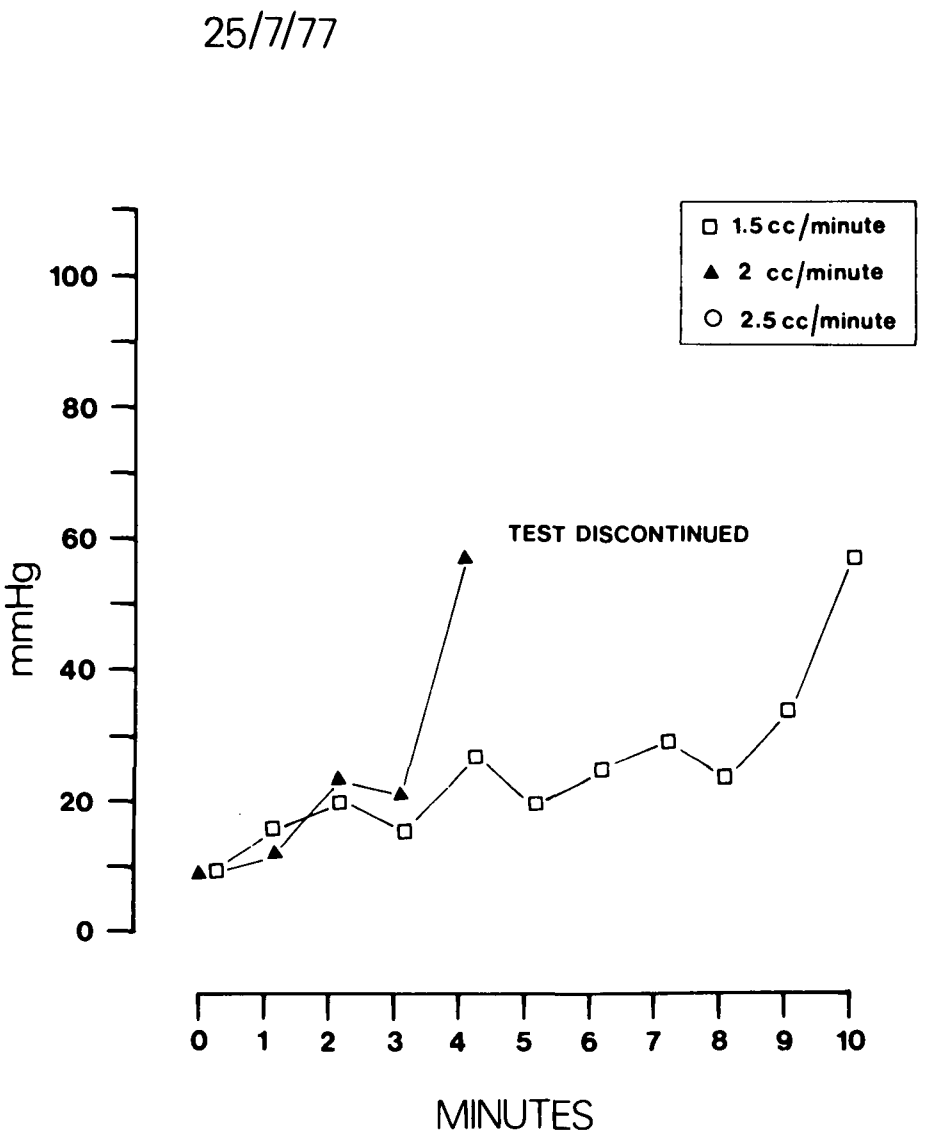

Figure 3 — Abnormal CSF infusion test showing significant rise in CSF pressure with initial infusion. Test discontinued with second highest infusion rate because of excessive rise in pressure.

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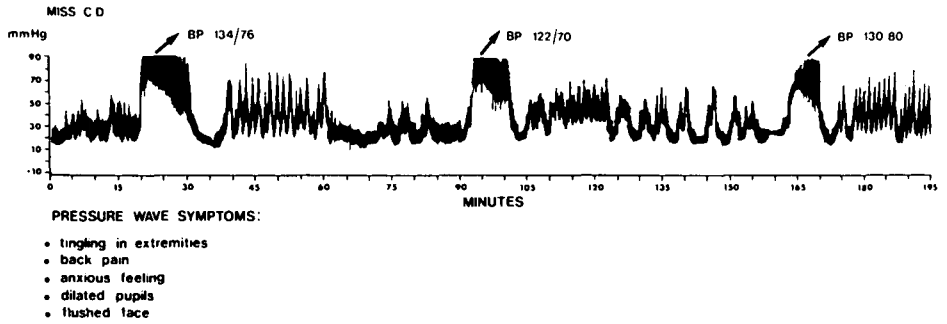

Figure 4 — Series of plateau waves (Lundberg "A" waves), coinciding with patient's symptoms.

Jacobs, Conti et al., 1976; Ojemann, Fisher et al., 1969; Philippon, Ancri et al., 1971; and Shenkin, Greenberg et al., 1973), pneumoencephalography (Adams, Fisher et al., 1965; Greitz and Grepe, 1971; Lemay and New, 1970; Rovit, Schecter, et al., 1972), cerebral blood flow determinations (Greitz, Grepe et al., 1969; Matthew, Meyer et al., 1975; and Hartman, Alberti et al., 1976), response to repeated lumbar punctures (Hakim and Adams, 1965) and CT scanning (Black, 1980; Crockard, Hanlon et al., 1976; Greitz and Hindmarsh, 1975; Hindmarsh and Greitz, 1975; and Jacobs and Kinkel, 1976). Unfortunately none of these methods accurately and consistently predict the effect of shunting.

Continuous ICP monitoring (Chawla, Hume et al., 1974; Cooper and Hulme, 1966; Hartman, Alberti et al., 1976; Matthew, Meyer et al., 1975; Symon, Dorsch et al., 1972; and Symon and Hinzpeter, 1977) appears to be the most reliable predictor, but even though its risks are few, it probably should not be used in the investigation of every patient being considered for shunting. Also, some doubt has been raised regarding the significance of B waves, previously considered an indicator for shunting, since B waves are regularly associated with REM sleep (Gucer and Viernstein, 1979).

The role of the constant rate infusion test in assessing CSF dynamics is becoming more clearly defined and refined by a number of investigators (Braham, Sarova-Pinhas et al., 1971; Foldes and Arrowood, 1948;Hartman, Alberti et al., 1976; Katzman and Hussey, 1970; Martins, 1973; Matthew, Meyer et al., 1975; and Nelson and Goodman, 1971). Contrary to some (Stein and Langfitt, 1974; Wolinsky, Barnes et al., 1973) but in agreement with others (Katzman and Hussey, 1970; Nelson and Goodman, 1971), we have shown that it is of considerable value in predicting shunt benefit. An abnormal response consists of a rapid rise of pressure at various infusion rates, and a slow return to baseline levels. An increase in lumbar pressure of more than 1.5 mm Hg per minute at an infusion rate of 1.5 ml/min is abnormal. The CSF infusion test is also useful in assessing the patency of shunts (Symon and Hinzpeter, 1977; Woodford, Saunders et al., 1976). Patients with a normally functioning shunt show a completely flat pressure response, conspicuously different from the rapidly rising pressure if the shunt is blocked.

We have recently reviewed 87 CSF infusion tests in 81 adult patients, with the following findings: 1) Of thirty-seven hydrocephalic patients with an abnormal infusion test, thirty-two were shunted; twenty-eight of the thirty-two improved. 2) In six patients, the infusion test correlated extremely well with continuous intraventricular pressure recording in predicting shunt benefit. 3) The infusion test correctly predicted shunt patency or blockage in all five patients studied by this test. 4) Six patients with pseudotumor cerebri have been followed by repeated infusion tests and their response to treatment correlated well with the infusion test result. In selecting patients who are suspected of having NPH for shunting, our current approach is a CT scan to show anatomical detail, followed by a CSF infusion test.

Although an acute increase in CSF pressure may not interfere with cerebral function (as is the case in pseudotumor cerebri) (Ryder, Rosenauer et al., 1953), there is often a direct relationship between an acute transient rise in intracranial pressure and the occurrence of acute episodic symptoms. Lower pressure waves are often accompanied by less severe symptoms, and higher pressure waves by more severe symptoms, although the correlation is not absolute. Pressure or "A" waves are related to altered vasomotor control of cerebral circulation. The initial rise in ICP may be triggered by CO_2 retention; it results in decreased cerebral vascular resistance and vasodilation, which in turn causes increased cerebral blood volume and a consequent further rise in ICP. Vasomotor control is later regained, often due to hyperventilation, and the ICP returns to normal. "A" waves occur exclusively at the steep part of the volume pressure curve (Miller and Garibi, 1972).

Our patient's periodic bizarre symptoms might have been attributed to a seizure disorder. Indeed, the relationship of such symptoms to epileptic phenomena in patients with brain tumors has been considered by many workers (Cairns and Mosberg, 1951; Grinker, 1976; Jackson, 1871; Penfield and Erikson, 1941; Penfield and Jasper, 1954). Such symptoms have been variously described as "tetanus-like seizures", "cerebellar or tonic seizures", "minor epilepsy", "mesencephalic seizures", and "diencephalic autonomic seizures". Lundberg (1960) suggested that these "episodes" had a close relationship with "A" waves. This patient's symptoms were clearly related to plateau waves, and it is unlikely that her symptoms were epileptic. EEG recordings during plateau waves characteristically show no signs of cortical epileptic activity (Lundberg, 1960). Our patient's EEG showed "transient abnormalities of cerebral activity, not epileptiform, superimposed on a normal pattern, suggesting a brain stem dysfunction". Even though her symptoms were related to plateau waves, it is difficult to explain such bizarre symptoms as hip pain, shoulder pain, and flatulence. An interesting variety of similar

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symptoms have also been recorded by Lundberg (1960). Perhaps distortion of spinal nerve roots from distention of the dural sleeves during periods of high pressure is a cause of limb pain. Distention of the periventricular area may explain some of the autonomic symptoms. Interference with cerebral blood flow when the pressure is extremely high is probably a contributing factor in the genesis of such symptoms.

"Slit ventricles" are not uncommon in children whose shunts are blocked. One possible explanation is subependymal gliosis described by Russel (1949). The gliosis is due to repeated distention and inflammation and stiffens the ventricular walls, discouraging their enlargement in spite of high ventricular pressure. Engel, Carmel and Chutorian (1979) have recently described five children with blocked shunts, high pressure and small ventricles. They found increased cerebral elastance in four of their patients. They have suggested the ingenious term "Normal Volume Hydrocephalus". In adults the ventricles usually dilate when intraventricular pressure rises. The presence of small ventricles in the face of waves of high ventricular pressure in our adult patient was rather surprising. We suspected that the cerebral elastance, although not measured, was high and played at least some role in preventing ventricular dilatation. Pascal's law, by which the force on ventricular walls has been used to explain the symptoms in patients with normal pressure and large ventricles (NPH), may also explain the symptoms in our patient with high pressure and small ventricles, or HPN ("high pressure normocephalus"). One wonders, however, whether the ventricles might have enlarged in time.

It is important to heed the complaint of episodic or bizarre symptoms and to recognize the significance of raised pressure in shunted patients even though the ventricles are of normal size. Since the preparation of this report, three more adult patients have come under our care with a similar situation.

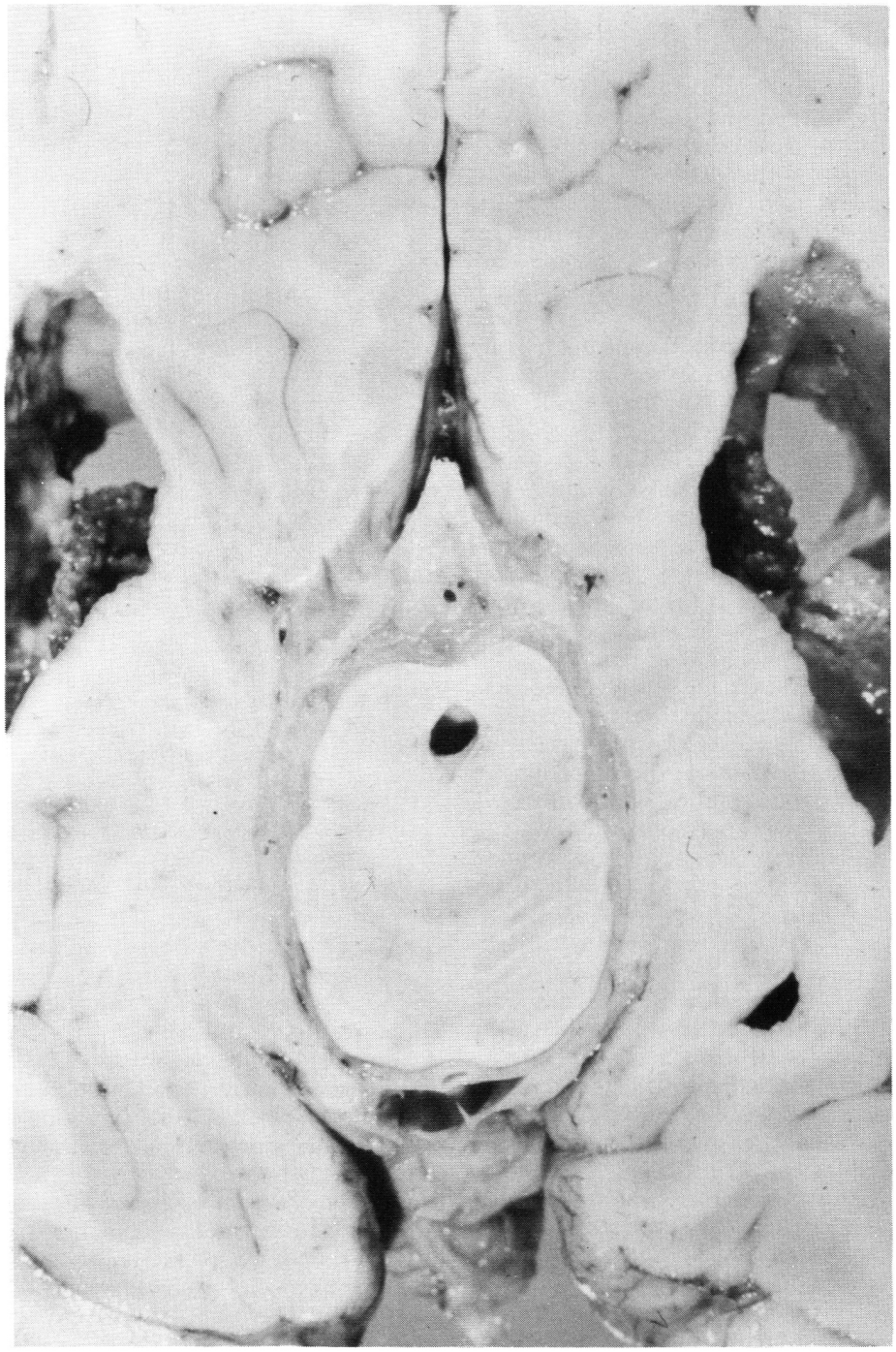

Figure 5 — A case of subarachnoid hemorrhage complicated by the delayed onset of communicating hydrocephalus. The brain stem has been cut at right angles to its axis at the level of the junction of midbrain and pons. CSF flow has been obstructed at the level of the incisura of the tentorium and the cisterna ambiens is seen to be obliterated by dense fibrous tissue. Ventriculoperitoneal shunting has been only partially successful in reversing the hydrocephalus, and the lateral ventricles and aqueduct are seen to have remained dilated.

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PATHOLOGICAL PERSPECTIVES

The effects of obstruction of CSF pathways distal to the 4th ventricular foramina following subarachnoid hemorrhage depend to some extent on the site or sites of obstruction to CSF flow.

Hemorrhage from rupture of a berry aneurysm is predominantly basal, and post hemorrhagic fibrous obliteration of the subarachnoid space is frequently severe at the base and may obliterate the cisterna ambiens (fig. 5). The result of obstruction at this site is a pressure gradient between the ventricular system and the subarachnoid space over the cerebral convexities with the development of classical communicating hydrocephalus. Compression of the cerebral convexities against the dura mater of the cranial vault may be sufficiently severe to produce herniation of brain parenchyma through trabeculae of the lateral lacunae into the arachnoid villi (fig. 6).

If, however, the subarachnoid blood passes beyond the basal cisterns (as it is likely to do in less severe subarachnoid hemorrhage when the blood fails to clot due to dilution with CSF) it will reach the arachnoid villi, most of which are parasagittal in location. Absorption of CSF may then be impaired by fibrous obliteration of the arachnoid villi, thus leading to altered hydrodynamics demonstrable by an abnormal CSF infusion test.

If obliteration of CSF pathways at the level of the cisterna ambiens has not occurred, there will be no pressure gradient between the ventricular system and the subarachnoid space over the cerebral convexities. Under these circumstances there will be little or no tendency for ventricular enlargement at the expense of the external subarachnoid space.

In the case presented here there was radiographic evidence of obstruction of CSF flow, shortly after her subarachnoid hemorrhage, and some evidence to suggest that the obstruction was at the level of the cisterna ambiens. We did not know if the arachnoid villi were also normal. The effects of subarachnoid hemorrhage are quite unpredictable. In the majority of cases the blood undergoes complete absorption without fibrosis, while in other cases the blood incites a variable and sometimes vigorous fibroblastic reaction at either or both of the sites of CSF flow mentioned, with fibrous obliteration of the CSF pathways. It is also possible that obstruction in the post-hemorrhagic period may subsequently resolve due to condensation of scar tissue, re-establishing continuity of CSF pathways.

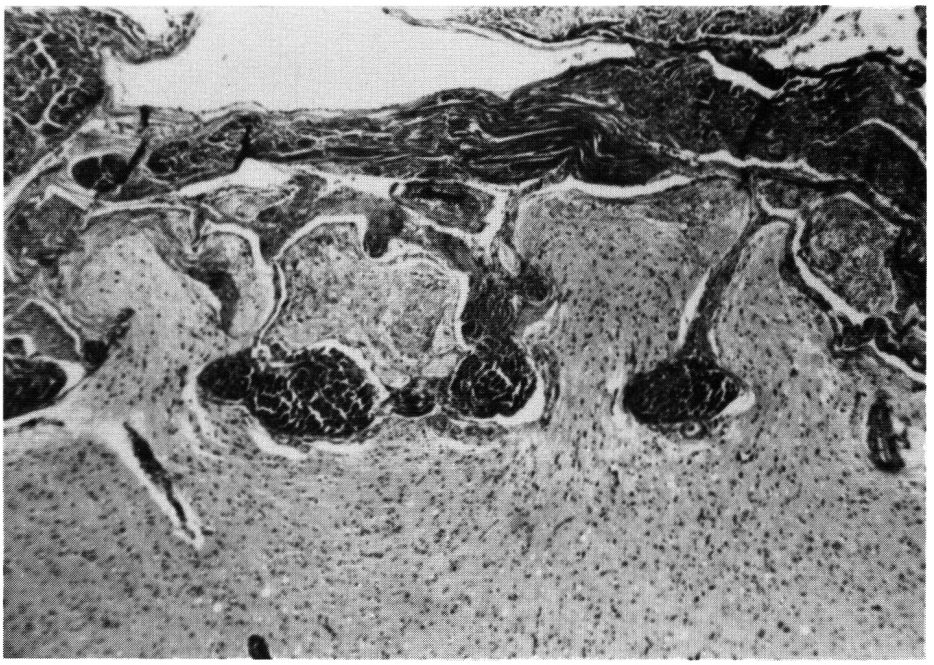

Figure 6 — Parasagittal lateral lacunae (top) and associated trabeculated dura mater and arachnoid villi. (Masson trichrome stain: original magnification x 40). Compression of brain tissue against the inner table of the skull due to hydrocephalus from obstruction of CSF flow at the level of the cisterna ambiens has resulted in herniation of brain parenchyma between dural trabeculae into the partially fibrosed arachnoid villi. Subsequent resolution of blockage of CSF flow at the level of the cisterna ambiens would not improve CSF absorption, although the pressure gradient between the ventricular system and the subarachnoid space over the cerebral hemispheres would disappear permitting the return of ventricular size to normal. Such a mechanism would account for cases of "high pressure normocephalus".

The subsequent course of our case included a pattern of variable adequacy of CSF drainage (by normal routes or by shunt) requiring a number of shunt revisions. There may well have been transient periods of ventricular dilation, ependymal and subependymal gliosis. However, the gliosis seen in such cases is usually no greater than that frequently seen with aging where hydrocephalus occurs ex-vacuo, and therefore is not likely to be a significant restrictive factor in determining ventricular size. This is particularly the case in portions of the ventricular wall such as the head of the caudate nucleus where the ventricular cavity is concave in configuration.

Unfortunately, there is no reliable morphological technique available at present for assessing the degree of obstruction to CSF flow produced by either subarachnoid space fibrosis or by fibrosis of arachnoid villi. Both undoubtedly occur, unpredictably in any single case, and the two sites of obstruction probably have different effects on ventricular size. The hypothesis that obstruction of arachnoid villi is the principal process in the recent course of events of our patient, seems to best fit the available data.

COMMENTS OF PEDIATRIC NEUROSURGEON

A child with spontaneous subarachnoid hemorrhage (SAH) is more than twice as likely to have suffered from rupture of an intracranial arteriovenous malformation (AVM) than rupture of an aneurysm. Studies at the Hospital for Sick Children for the period 1950 - 1979, revealed seventy

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children with subarachnoid hemorrhage due to arteriovenous malformations and twenty-seven with ruptured aneurysms. The young lady whose case is presented here is thus unusual not only as to the cause of her subarachnoid hemorrhage, but also the age of presentation, location and size of her aneurysm.

For a condition which is supposedly of congenital origin, it is curious that intracranial arterial aneurysms tend not to rupture during childhood and adolescence. In large published series of aneurysms, the percentage of patients who were children varied from one to four percent of all groups surveyed (Humphreys, Hendrick et al., 1972). Moreover, intracranial aneurysms rarely show up as incidental findings in pediatric cerebral angiographic studies. Harwood-Nash and Fitz (1976) upon reviewing several thousand cerebral angiograms report that "no clinically latent cerebral aneurysms have been discovered at angiography or serendipitously at autopsy in infants and children".

Pediatric aneurysms differ in site and size from adult aneurysms. The carotid bifurcation is the commonest location in children, followed by anterior cerebral-anterior communicating complex, and posterior circulation aneurysms account for 25% of our series (Table 1). Thus we agree with Amacher and Drake (1979), who state, with regard to aneurysms in the young, that "they occur in unusual locations relative to the adult population and many are of giant size". Eight of the twenty-seven aneurysms in our series were giant.

Hence, the case reported here is somewhat unique in that the aneurysm presented at age three years, was located at the anterior communicating — anterior cerebral complex, but was of usual size. The child responded well to clipping of the aneurysm, but developed communicating hydrocephalus. She was well served by a shunt until she presented with the symptoms which prompted the current review. In her many symptom-free years, the young lady may have achieved the state of "arrested hydrocephalus", in which there is stabilization of the clinical picture and of the size of intracranial spaces containing CSF. DiRocco, Caldarelli et al. (1977) however, exort us to pursue an "accurate longitudinal evaluation of the clinical status and size of intracranial CSF spaces (which) should allow differentiation between an 'arrested' and a slowly progressive hydrocephalus". They also recommend intracranial pressure (ICP) monitoring as a useful adjunct in such situations, and this is the pivotal point of Dr. Schutz's presentation.

TABLE 1

Distribution of 27 Intracranial Aneurysms, Hospital for Sick Children: 1950 - 1979

Anterior Cerebral — Anterior Communicating	4
Pericallosal	2
Posterior Communicating	1
Internal Carotid	8
Middle Cerebral	4
Basilar-Posterior Communicating	6
Vertebral-Posterior Inferior Cerebellar	2

It is indisputable that the patient had episodic high pressure waves. But how could her ventricles remain unexpanded in the face of these pressure escalations? We might theorize that ependymal and periventricular gliosis results in a ventricular "stiffening" after longstanding hydrocephalus. Such changes are not unusual, and not surprising when one considers that the structural changes in hydrocephalus are ependymal flattening and tearing, an increase in periventricular white matter water content and demyelination (DiRocco, Ditripani et al., 1979). One might argue that the "extent of ventriculomegaly is dependent on the restraining forces exerted on the brain by the covering dura and skull. As the skull and dura become thicker and more rigid and finally cease to enlarge, they tend to oppose and finally check the increase in ventricular volume. Thus the elevation of intraventricular pressure may be both more prolonged and increased by the properties of these structures" (Sahar, 1979). Perhaps the latter is the explanation for the phenomenon of the slit ventricle syndrome and for the success of subtemporal decompression and dural opening (Epstein, Fleischer et al., 1974; Holness, Hoffman et al, 1979).

Whatever the solution to this perplexing problem, Dr. Schutz has drawn our attention to the value of ICP monitoring in the symptomatic treated hydrocephalic patient with normal size ventricles. If the pediatric neurosurgeon may deign to advise his "adult" colleagues about the "graduates" of pediatric hydrocephalus treatment, the following are applicable:

1. Communicating hydrocephalus carries a better prognosis in terms of future shunt independence than does obstructive hydrocephalus. The corollary of this is that those patients with communicating hydrocephalus that do become shunt dependent, tend to become ill very rapidly with shunt obstruction and require immediate care.
2. "Acquired" forms of obstructive hydrocephalus (e.g. tumor obstruction of CSF pathways) carry a better prognosis with regard to future shunt independence than "congenital" obstructive hydrocephalus. However, current figures suggest that 50% of myelodysplastic children with obstructive hydrocephalus will become shunt independent by age five to six years.
3. When a shunt obstructs, ventriculomegaly may be recognized on computed tomography. But beware, especially in patients with communicating hydrocephalus, of the slit ventricle syndrome and, as we appreciate from the current case presentation, the state of "high pressure normal ventricles".

CONCLUDING REMARKS

It is now clear that a blocked shunt may result in severely raised intracra-

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nial pressure, either intermittent or constant, without ventricular enlargement, both in children and in adults. We have been given four posssible hypotheses for failure of the ventricles to enlarge: 1) subependymal gliosis, 2) site of CSF pathway obstruction, 3) increased cerebral elastance, and 4) a corollary of Pascal's law. We have three possible names for the condition, "slit ventricle syndrome", "normal volume hydrocephalus" and "high pressure normocephalus". Whatever the pathogenesis and whatever the name, the condition demands treatment by shunt revision.

Dr. Schutz has shown that the pressure response to carefully quantitated constant rate CSF infusion, at various rates of infusion, can provide a reliable prediction of shunt benefit in patients with normal pressure hydrocephalus. He has also shown the importance of ventricular pressure monitoring in demonstrating episodic pressure increases even though the classical signs of intracranial hypertension are absent and lumbar pressure is normal.

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https://doi.org/10.1017/S0317167100023222 Published online by Cambridge University Press