HYDROCEPHALUS AND THE DURAL VENOUS SINUSES

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In a classic monograph on the pathology of hydrocephalus, Russell\textsuperscript{21} recorded admirably all known causes of internal or obstructive hydrocephalus. At the same time she speculated about the relationship of thrombosis in the dural venous sinuses to hydrocephalus. The clinical and pathological material suggested that sinus thrombosis could be followed by a limited degree of hydrocephalus after a time and that the size of the ventricles could revert to normal with canalization of the thrombus. Further, roentgenographic evidence of antemortem hydrocephalus could not be corroborated at necropsy, because collapse of the ventricles was thought to be the cause of a normal ventricular pattern. Deliberate attempts to occlude the superior sagittal sinus in a series of animals produced only slight and equivocal degrees of hydrocephalus.\textsuperscript{3} The over-all results were attributed to compensation of circulation by collateral veins.

It is the purpose of this presentation to relate obstructive phenomena in the dural venous sinuses to states of intracranial hypertension and hydrocephalus. Aside from the customary diagnostic measures, the diagnostic value of measurements of intrasinal venous pressure and dural sinus venography will be emphasized.

The physiological studies of Becht\textsuperscript{5} indicated that pressure of the intracranial fluid was the result of the influence of venous and arterial pressure. Venous and fluid pressures varied in the same direction and to some degree proportionately in nearly every case. Raising the venous pressure raised the fluid pressure, while lowering the venous pressure lowered the fluid pressure. Increasing or decreasing the fluid pressure moderately did not alter the venous pressure unless the arterial pressure was affected. Weed and Flexner\textsuperscript{21} corroborated these studies.

In a series of dogs Bedford\textsuperscript{4,5} showed that with prolonged occlusion of the external jugular veins or the lateral sinuses, the venous pressure rose higher than the fluid pressure. Within minutes the fluid pressure gradually fell to its normal level; the venous pressure maintained itself above that of the spinal fluid until release of the occlusion. Bedford then ligated the external jugular veins and concluded that the procedure did not cause hydrocephalus after 6 weeks of observation. In this series the internal jugular veins, usually small accessory channels, were dilated considerably.

Stewart\textsuperscript{26} was able to produce a state of chronically increased intracranial pressure in cats by ligating both posterior sinuses of the vertebral column, both internal jugular veins and often one external jugular vein. The fluid pressure fell slowly as remaining venous channels grew larger and new venous channels developed. To maintain the raised pressure for 4 to 5 weeks required second and third operations, but no hydrocephalus resulted.

Bering and Salibi\textsuperscript{6} were able to produce increased cerebral venous pressure, cerebrospinal fluid pressure and hydrocephalus in 74 per cent of 21 dogs as a result of occlusion of the cephalic venous drainage. Ventricular enlargement reached its peak 2 or 3 weeks after the second dissection of the neck and then remained stabilized. This effect may have been the result of development and enlargement of remaining venous collateral drainage. They felt that the mechanisms involved in the ventricular enlargement were a combination of two factors: the possible failure of cerebrospinal fluid absorption in the face of increased venous pressure in the sagittal sinus and increased intraventricular pulse pressure from the choroid plexus.

In the human, deliberate ligation of the
jugular veins has been followed by varied effects. Rohrbach and Linser ligated the internal jugular vein unilaterally during dissection of the neck for cancer. In each case death resulted from acute passive congestion of the brain, because of the ligation of the major draining venous sinus. Guillain reported a case of ligation of the internal jugular vein followed by neurological symptoms and increased intracranial pressure relieved by repeated spinal punctures. Creyssel and Douillet reported 1 death and 2 cases in which neurologic complications followed ligation of a jugular vein. More recent reports of unilateral and bilateral ligation of the jugular vein indicate that the procedure may be innocuous or followed by mild neurological complications, chronic neurological symptoms and states of intracranial hypertension. The latter may be evanescent or prolonged and have not been evaluated with pneumoencephalography and dural sinus venography. It remains undetermined how many of these patients would show the ultimate effects of increased intracranial pressure, that is hydrocephalus.

An analysis of clinical entities that affect the dural venous sinuses, provides a correlation of obstructive phenomena in the dural venous sinuses with increased intracranial fluid pressure and communicating hydrocephalus. Symonds evolved the clinical picture of "otic hydrocephalus," and demonstrated the relationship of otitis media to thrombophlebitis of the dural venous sinuses and the picture of benign intracranial hypertension. Gardner corroborated Symonds' clinical observations and stated that encephalography showed no evidence of hydrocephalus. The roentgenograms reproduced in 3 cases indicated a mild but definite hydrocephalus. Foley and Bradshaw corroborated the clinical picture described by Symonds with their findings at operation, autopsy, ventriculography and arteriography. In a small per cent of cases the ventricles were larger than normal. Frenckner was the first to demonstrate by dural sinus venography the relation of otitis media to thrombosis in the lateral sinus. Since then Ray et al. and Frenckner have shown that thrombosis of a major dural venous sinus invariably leads to intracranial hypertension. Kinal and Jaeger were able to demonstrate hydrocephalus in 1 patient out of 3 with thrombosis of a major dural venous sinus.

Cranial trauma has afforded further case material for the study of obstructive phenomena in the dural venous sinuses and their relation to intracranial hypertension and communicating hydrocephalus. Martin first described cases of head injury in which signs of increased intracranial pressure developed, without signs of severe intracranial bleeding and in which ultimate recovery occurred without operation. One syndrome was attributed to obstruction in the sagittal sinus itself but not infrequently in the lateral sinus. Many of his cases had points of similarity to those of "otic hydrocephalus" and he used the term "traumatic hydrocephalus" in referring to them. In a discussion of benign forms of intracranial hypertension, Foley related 4 cases of mild head injury to inexplicable intracranial hypertension. By means of dural sinus venography, Kinal was able to demonstrate that in his cases the "traumatic hydrocephalus" of Martin was indeed caused by thrombosis in the dural venous sinuses. In these cases, the spinal fluid pressure and the intrasinal venous pressure were increased invariably, although they might be normal. In these and subsequent cases, fluid dynamic studies indicated that recanalization of the obstructed sinus was possible. The patent venous sinus system could dilate to accommodate the additional burden of blood. Collateral channels of drainage into the vertebral-azygos plexuses of Batson could drain excess quantities of blood thereby averting a state of intracranial hypertension. That a communicating hydrocephalus may result from traumatic thrombosis of a major draining dural venous sinus has been shown in a small per cent of unpublished cases.

It has been shown in the preceding discussion that deliberate or clinical interference of drainage through the major dural venous sinuses may induce states of intracranial hypertension. In the clinical group of throm-
basis of the major draining dural venous sinuses a small number of cases has been observed associated with communicating hydrocephalus. The inference is that other forms of communicating hydrocephalus may be related to obstructive phenomena in the dural venous sinuses. The following case reports are presented as examples of a probable relationship of multiple congenital abnormalities of the dural venous sinuses with the presence of a communicating hydrocephalus in infants. The demonstrations of the congenital anomalies are founded on the anatomical studies of Padget and Woodhall, and the techniques of dural sinus venography were those evolved by Frenchner and Ray et al.

CASE REPORTS

Case 1. A 4-month-old infant with obvious hydrocephalus and elevation of the anterior fontanelle, was found to have a head 18\frac{1}{2} inches in circumference. The eyeballs were divergent and deviated inferiorly.

Roentgenograms of the skull showed a bulging anterior fontanelle and a hydrocephalic configuration. Bilateral subdural punctures showed no collections of fluid. No simultaneous ventricular or lumbar punctures, or ventriculograms were done.

The intrasinal venous pressure was 300 mm. of blood. Serial sagittal sinograms were taken during the injection of 10 cc. of 50 per cent Miokon. The sagittal sinus was slightly dilated and partially duplicated (Fig. 1). The torcular gave rise to a right lateral sinus of considerable volume, which terminated in a stenotic sigmoid sinus. The right jugular bulb was normal, and continuous with a right jugular vein containing a heavier concentration of contrast medium than the left jugular vein. The left lateral sinus was of smaller calibre than the right and its sigmoid sinus was markedly stenotic. It terminated in a normal left jugular bulb, but the left jugular vein was smaller in calibre than the right.

In summary the sinograms disclosed a small lateral sinus on the left side while both sigmoid sinuses were markedly stenotic. The left jugular vein was smaller in calibre than the right.

Case 2. A male infant, 5 weeks of age, had a dolichocephalic hydrocephalus. The suture lines were separated widely and both fontanelles were tense. The circumference of the head was 15\frac{1}{2} inches.

The veins of the scalp were numerous, and distended with dark blood. Compression of these veins resulted in excessive distension of the segment of vein cephalad to the compressing finger. After "wiping out" a segment of the vein between two compressing fingers, there was poor refilling from the cardiac direction on release of compression; more rapid refilling of the segment occurred from the cephalad direction. The cerebral cortex did not transilluminate but the dilated sagittal sinus was visible.

Simultaneous ventricular and lumbar punctures showed a pressure of 90 mm. of fluid at both sites. When the infant cried the fluid levels rose and fell equally in both manometers. A bubble ventriculogram showed marked enlargement of the ventricular system.

At the age of 8 weeks the circumference of the head was 17\frac{1}{2} inches. The exposed sagittal sinus appeared widened and convex from distension with blood. The intrasinal venous pressure was 80 mm. of blood.

Serial sinograms showed an irregular sagittal sinus (Fig. 9). A normal right lateral sinus arose from the torcular. It terminated in a tubular right sigmoid sinus stenotic throughout its entire length. The right jugular bulb was formed irregularly and the right jugular vein appeared normal, though dilated in its lower portion. The left lateral sinus was visible initially as a markedly attenuated channel, continuous with a
graphic appearance of the system of the left dural venous sinus. Cardiac arrest occurred when a ventriculoauricular shunt was attempted. Permission for postmortem examination was not granted.

Case 3. A 9-month-old infant was found to have a head measuring 22\(\frac{1}{2}\) inches in circumference. The anterior fontanelle was tense and there were no palpably separated sutures.

Roentgenograms of the skull showed a hydrocephalic configuration and separation of the suture lines. A simultaneous lumbar and ventricular puncture disclosed a pressure of 450 mm. in both manometers. The fluid levels rose and fell equally at both sites. Pneumoencephalograms revealed a communicating hydrocephalus.

After continuous treatment with Diamox in lieu of a ventriculoauricular shunt the head was 21\(\frac{1}{2}\) inches in circumference at the age of 12 months. At the age of 18 months the circumference of the head was 22 inches. An abrupt increase in the circumference of the head occurred at the age of 20 months. Further diagnostic studies were permitted at the age of 33 months when the circumference of the head was 24\(\frac{1}{2}\) inches. At this time the infant could stand but could not walk unsupported. The anterior fontanelle was closed. The veins of the scalp were not distended. All functions of the cranial nerves were normal. Speech was relatively well developed. Coordination in the upper extremities was good. The deep tendon reflexes were normal.

The venous pressure in the veins of the scalp was 120 mm. of blood. The intrasinal venous pressure was 370 mm. of blood. A simultaneous lumbar spinal fluid pressure varied from 380 mm. to 400 mm. of fluid.

Serial sinograms showed a sagittal sinus that was dilated and foreshortened (Figs. 3 and 4). The right lateral sinus arose from a torcular of normal pattern and coursed obliquely downward at an angle of 45\(^\circ\). It continued into a sigmoid sinus of larger calibre which rapidly became stenotic and joined a normal jugular bulb and vein. The left lateral sinus arose by three separate narrow channels which fused to form one stenotic channel coursing obliquely downward at an angle of 45\(^\circ\). The termination of the stenotic left lateral sinus was lost in the region of the petrous pyramid. No left sigmoid sinus, jugular bulb or vein could be visualized in the serial sinograms.

In summary the serial sagittal sinograms demonstrated an obliquely situated right lateral sinus and right stenotic sigmoid sinus and an obliquely situated stenotic left lateral sinus with atresia of the sigmoid sinus, jugular bulb and vein.

Case 4. A 10-month-old male infant with re-
tarded neurological development had a slightly asymmetrical head, with flattening of the right parieto-occipital region. The anterior fontanelle was tense and circumference of the head was 10 inches.

Roentgenograms of the skull showed a calvarium that was slightly large for the infant's age. The asymmetry of the skull was confirmed. The suture lines were separated.

In the horizontal position, the intrasinal venous pressure was 280 mm. Measured simultaneously, the spinal fluid pressure was 280 to 290 mm. In the sitting position the spinal fluid pressure was 470 to 480 mm. The intrasinal venous pressure was 60 mm. at the time. The patient's length from the site of the lumbar puncture to the crown was 410 mm.

The serial sinogram showed a normal sagittal sinus and torcular (Fig. 5). The right lateral sinus was narrowed moderately and emptied into a stenotic sigmoid sinus. The right jugular bulb and vein were slightly smaller in calibre than the left. The left lateral sinus was normal, while the left sigmoid sinus appeared as a flattened, attenuated channel. The left jugular bulb and vein were normal. Attempts to carry out a pneumoencephalogram were not successful and roentgenographic evidence of hydrocephalus was lacking.

SUMMARY

The normal intracranial fluid pressure is the result of the influence of venous and arterial pressure and varies in the same direction and to some degree proportionately to the venous pressure. While changes in venous pressure cause proportionate changes in the fluid pressure, moderate changes in fluid pressure do not appreciably alter the venous pressure. This physiological observa-

Figs. 3 and 4. Case 3. The sagittal sinus was foreshortened and dilated. The right lateral sinus coursed obliquely downward. The right sigmoid sinus became stenotic rapidly. The stenotic left lateral sinus arose from the torcular by three separate channels. The left sigmoid sinus, jugular bulb and vein were atretic.

Fig. 5. Case 4. The right lateral sinus was narrowed moderately and emptied into a stenotic sigmoid sinus. The left lateral sinus was normal. The left sigmoid sinus appeared as a flattened, attenuated channel.
tion suggests that pathologic processes of the dural venous sinuses that increase the venous pressure cause increased intracranial fluid pressure.

Though the ligation of cranial venous channels in animals has resulted in acute and chronic states of intracranial hypertension, only Bering and Salibi\(^6\) were able to produce more consistent hydrocephalus.

Deliberate ligation of the jugular veins in man has resulted in death, neurological deficits and states of acute and chronic intracranial hypertension. Infectious and traumatic thromboses of the dural venous sinuses may result in states of chronic intracranial hypertension and hydrocephalus in a small percentage of cases.

Dural sinus venography applied to 4 cases of congenital hydrocephalus has demonstrated anomalies of the dural venous sinuses which appear to be bilateral in the form of stenoses and/or atresias. The presence of intracranial hypertension and hydrocephalus with increased intrasinal venous pressures and congenital stenoses and/or atresias of the dural venous sinuses suggests a causal relationship.

REFERENCES

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