Historical vignette

Third ventriculostomy for internal hydrocephalus complicated by unrecognized subdural hygroma and hematoma: a case report of a patient treated by Dr. Walter Dandy

ROBERT B. KING, M.D., RICHARD L. DAVIS, M.D., PH.D., AND GEORGE H. COLLINS, M.D.

Departments of Neurosurgery and Pathology, State University of New York Upstate Medical University, Syracuse, New York

The authors review the case of a patient treated by Dr. Walter Dandy. When the patient was a young child he underwent two right transtemporal third ventriculostomies during which he sustained an unrecognized contralateral subdural hygroma and a chronic subdural hematoma with a mild infantile hemiparesis. He was able to complete high school, albeit at a slower pace than usual. As an adult he held several limited employment positions, lived at home for several decades, and was later cared for at a nursing home for a short time. The patient died when he was 66 years of age.

KEY WORDS • infantile hydrocephalus • subdural hygroma • hematoma • ventriculostomy • children

Efforts to control infantile hydrocephalus have been recorded for centuries, but were largely ineffective until the development, early in the 20th century, of a better understanding of the normal and pathological anatomy of CSF pathways and circulation. Among those responsible at that time for advancing our conceptions of normal and compromised CSF circulation were Drs. Walter E. Dandy and K. D. Blackfan. As a result of their studies, they were able to develop new surgical approaches to reduce the problems associated with the management of infantile hydrocephalus.

An early derivative of their studies was a third ventriculostomy introduced in 1920. It involved an anterior approach to the third ventricle through the lamina terminalis and, frequently, required the sacrifice of one optic nerve. Outcomes of this procedure were frequently compromised by the postoperative accumulation of CSF in the subdural space. Nevertheless, this anterior approach remained a preferred one for several years. In 1966 the authors of a review of outcomes of patients who had undergone the procedure during the preceding 30 years reported an operative mortality rate of 15% and an arrest-of-hydrocephalus rate of 70%. Other operative techniques included catheterization of the sylvian aqueduct and a third ventriculostomy performed transcerebrally through the foramen of Monro.

The disadvantages of these procedures led Dr. Dandy to consider yet another approach in 1930, which was reported in 1933. He suggested using a more posterior epidural approach from the right side across the floor of the middle fossa to enter the ballooned third ventricle and establish a fistula into the cisterna interpeduncularis. In addition to preservation of the optic nerve, a major advantage of this approach was the likely avoidance of a subdural spinal fluid accumulation "because the dura lining the middle fossa . . . ends medially in a shelf that lies along side the cisterna laterally." The temporal lobe, Dandy suggested, would therefore reestablish itself on the floor of the middle fossa and seal off that junction line (Fig. 1).

This case report concerns a patient who underwent a third ventriculostomy performed by Dr. Dandy through a right subtemporal approach to the third ventricle on February 18, 1930, when the patient was 27 months old, and a second similar operative procedure on February 6, 1931. Apparently, it was not uncommon for a secondary procedure to be performed if the fistula became sealed off.

This case is of historical interest because it shows the early treatment of infantile hydrocephalus and also because of the patient’s long survival with unrecognized complications due to the early surgical procedures.

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance.
Case Report

Clinical Summary. The patient, the product of an unremarkable pregnancy and delivery, appeared healthy with respect to growth, development, and behavior until July 1929. At the time it was noted that his head had enlarged from 58 to 61.5 cm during a 6-month period. He was referred to The Johns Hopkins University Hospital where a right temporal craniotomy was performed by Dr. Walter Dandy on February 18, 1930. Six months following surgery the patient’s mother began to notice a slight swelling at the incision site and some bulging of the anterior fontanelle. The boy’s head circumference had increased approximately 1.5 cm in the intervening period. In January 1931, he experienced a brief episode of unconsciousness followed by a continual occipital headache. This prompted a second surgical procedure on February 6, 1931, to reestablish the ventriculostomy; this repeated surgery was not an unusual requirement with such a procedure. When the patient returned home the second time he was substantially more irritable, vomited easily, and complained of increased headaches. His fontanelle remained flat, although the swelling at the temporal incision site varied in size.

The patient progressed steadily through his developmental stages, although his walking was somewhat delayed and he remained clumsy. He maintained passing grades in elementary school, but required 6 years to complete high school. An attempt at college, which had not been recommended, resulted in his withdrawal during the first semester. As an adult he worked as a self-employed salesman and, for several years, as a maintenance worker in a department store. He lived with his mother for many years until he was placed in a nursing home.

When the patient was in his mid-50s, despite his ability to work part-time, his family recognized that he could no longer be as independent as he had been in the past. He was referred for further medical evaluation.

Physical examination showed that the man’s cranium was enlarged and his head was tilted to the left side. His gait was slow and uncertain. On neurological examination, he was reported to have a mild left infantile hemiparesis and a mild diplegic gait. His visual perception and organizational skills were inadequate due, in part perhaps, to failing vision caused by severe primary optic atrophy, which was more severe on the left side than on the right. He also had mild vertical nystagmus. Fine motor coordination was limited on the left side. His memory was impaired and his mental processing was slow.

Detailed neuropsychological testing indicated a verbal intelligence quotient of 106 (66%) and a performance intelligence quotient of 78 (4%). The patient was no longer able to write, but could use a computer. His overall verbal ability was quite good; however, he did have significantly compromised cognitive and perceptual functions, which are generally controlled by the nondominant cortical regions.

The remainder of this patient’s life was uneventful, except that he was hospitalized briefly on one occasion while he was treated for seizures. In his later years he gradually became increasingly dependent on institutional care. The cause of his death at age 66 years is not known.

Imaging Reports. Current modes of neuroimaging were not available prior to 1981, when the patient was first examined. After they became available these studies were undertaken in this case and they were repeated periodically; however, no substantial changes developed during the time of...
observation. Magnetic resonance images (Fig. 2) were obtained 3 years before the patient’s death. They demonstrated a temporal skull defect on the right side and multiple cystlike fluid collections on the left side, which severely compressed the left hemisphere and shifted the falx and other midline structures far to the right. The left occipital horn and the right lateral ventricle were both substantially dilated (Fig. 2).

**Postmortem Pathological Report.** The formalin-fixed brain without the cyst and dura mater weighed 925 g. A large multilocular cyst on the left side contained bright yellow fluid, had walls that were 1- to 2-mm thick, measured 9 × 15 cm, and compressed the left hemisphere so that the latter was only 2 cm in width at the vertex (Fig. 3A and B). Ventrally, the cyst crossed the left temporal tip and extended to the interpeduncular fossa and the floor of the third ventricle. The left optic nerve was adherent to the cyst wall and was reduced in size. The cyst contents consisted of a somewhat turbid, mahogany-colored fluid with a viscosity slightly greater than water. On the coronal section (Fig. 3C and D), there was a reduction in the central white matter on the left side with normal-appearing cortical gray matter. The left lateral ventricle was of average size anteriorly, but the occipital horn was enlarged. The right lateral ventricle was enlarged throughout. The basal ganglia, thalamus, hypothalamus, brainstem, and cerebellum appeared to be normal, except for atrophy of the left cerebral peduncle, basis pontis, and pyramid.

Microscopically, the cyst walls consisted of two layers of tissue. The outer one was a thick, hypocellular fibrillar membrane with areas of mineralization. Immediately subjacent to this was a loose fibrillar membrane containing blood vessels (Fig. 4A). The former varied greatly in thickness and contained large deposits of calcium in areas where it was thickest. This membrane lay in apposition to the inner table of the skull and appeared to be coextensive with normal dura mater (Fig. 4A). The more loosely structured membranes, which frequently contained deposits of arachnoid cells on the outer surface, were found overlying the cerebrum and, in places, were coextensive with normal-appearing arachnoid (Fig. 4B). In other areas it was found to be in apposition to the greatly thickened subdural membrane and contained foci of calcium and hemosiderin, as well as hyperplastic arachnoid cells (Fig. 4A).

Between the floor of the third ventricle and the tissue comprising the cyst wall (Fig. 5), there was a ventral extension of an ependymal-lined outpouching of the third ventricle that appeared to communicate with the adjacent subarachnoid space on the left side, through a small aperture in the membrane wall (Fig. 5 arrow). Immediately adjacent to this subarachnoid space lay a dense collection of disorganized arachnoid and dura (Fig. 5D).

Other histological sections of the left cerebrum displayed no perceptible loss of cortical gray matter. The central white matter, however, was reduced to approximately 50% of its normal size. The putamen showed no cell loss, and diencephalic sections exhibited loss of thalamic and hypothalamic tissue only in subependymal areas. Brainstem sections showed a marked loss of descending fibers in the basis pontis on the left side, with an associated reduction in the size and number of neurons in the ipsilateral pontine nuclei, as well as a loss of pontocerebellar fibers.
Infantile hydrocephalus with subdural hygroma and hematoma

Fig. 5. Montage of several photomicrographs showing a tissue from the interpeduncular fossa and adjacent structures. Structures shown include the third ventricle (III), an extension of an ependymal-lined membrane from the floor of the third ventricle (IIIex), the subarachnoid space (SAS), the presumed site of communication between third ventricular fluid and the subarachnoid space; the arachnoid (A); the third cranial nerve (asterisk); the dura mater (D); and the thalamus (Th). H & E, original magnification × 4.

In the cerebellum there was a moderate reduction in the number of Purkinje cells.

Discussion

Although the nature of the surgical procedures performed when the patient was a young child was not certain from the historical information available to us, it was clear from neuroimaging and autopsy records that a third ventriculostomy had been performed through a subtemporal approach on the right side, opposite to the abnormalities demonstrated on the left. It would appear that this was one of the earliest cases treated using this approach, which was intended to avoid disadvantages of the anterior approach such as accumulation of subdural spinal fluid. This method was the only one used by Dandy after 1932, and in 1945 he published a summary of his experience with this method. There were 29 patients older than 1 year of age at the time of surgery with one operative death and four additional postoperative deaths within 6 months. Among 63 patients younger than 1 year of age at surgery, 10 died while in the hospital, 21 within the 1st year, and one after 12 years. The incidence of postoperative subdural spinal fluid collection in those cases is not known. There are no available autopsy reports pertaining to these patients.

Experience with recently developed endoscopic third ventriculostomies has shown very few postoperative subdural hygromas. Two such cases were effectively treated during the immediate postoperative period. Subdural hematomas, which occur occasionally after ventricular shunt placement, rarely occur in association with subdural hygromas.

In the case under discussion gross examination of the brain at autopsy revealed an obvious relationship between the cyst and the region of the interpeduncular cistern. Microscopic evaluation of this area demonstrated an ependymal-lined outpouching from the floor of the third ventricle, which communicated with the arachnoid and subarachnoid space on the left side. This extended into an area consisting of redundant arachnoid and thickened dura mater. A pathway for the fluid beyond this point could not be determined, but the histopathological findings suggest that the diversion of CSF to the subdural space could have occurred in this region. Nevertheless, diversion of spinal fluid through other channels in the basal cistern cannot be excluded. It is also uncertain as to when the spinal fluid may have accumulated in the subdural space. A pathway for adequate absorption of CSF was presumably established shortly after the second operative procedure; however, there was no abrupt or apparent progression of the patient’s symptoms until late in his sixth decade of life. It would appear that absorption of fluid was primarily through the subarachnoid space over the right hemisphere, where the sulci were well preserved.

The area of membranous thickening adjacent to the outflow track from the third ventricle, through which CSF may have entered the subarachnoid space, may also have been the site of a defect in the arachnoid leading to a subdural hygroma. This delicate membrane lying adjacent to the dura mater and over the surface of the brain appeared to envelop the entire subdural space on the left side. Some of the membranes were remarkably thickened and heavily calcified, and on neuroimaging examination were thought to have been associated with very long-standing subdural hematomas. The occurrence of such an association with pre-existing subdural hygromas has been suggested, but the mechanism relating hygromas to subdural hematomas is not clear. A history of head trauma can sometimes be found, but in this case the only known head trauma occurred in early adulthood and it was not associated with altered consciousness, neurological deficit, or change in the patient’s activities of daily living. It has also been suggested that blood vessels within a cyst membrane may give rise to spontaneous bleeding. Hemosiderin deposits were identified within the cyst membranes.

In summary, this autopsy study has documented many of the pathological conditions associated with the outcome of a transtemporal third ventriculostomy performed approximately 64 years before the death of the patient. The major neuroimaging and postmortem findings identified the skull defect on the right side with the accumulation of major abnormalities in the left hemicranium, portions of which were coextensive with the interpeduncular fossa at the site of the surgery.

On the basis of the material we reviewed, we were able to demonstrate a direct communication between the third ventricle and the subarachnoid space; however, no direct communication between the subarachnoid space and subdural spaces has been identified. Nonetheless, the area of abnormal relationships of arachnoid to dura adjacent to the interpeduncular cistern may have been the site of an abnormal communication, which gave rise to a subdural hygroma and subsequent subdural hematoma.

Acknowledgment

We are grateful to Dr. Arthur Rosenbaum for providing the postmortem MR images shown in Fig. 3.
References


Manuscript received September 9, 2002.
Accepted in final form December 20, 2002.
Address reprint requests to: Robert B. King, M.D., Department of Neurosurgery, Upstate Medical University, 750 East Adams Street, Syracuse, New York 13210. email: kingr@upstate.edu.