INTERNAL HYDROCEPHALUS

AN EXPERIMENTAL, CLINICAL AND PATHOLOGICAL STUDY*†

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Part 1.—Experimental Studies

1. INTRODUCTION

The term “hydrocephalus” is merely a symptomatic designation for an idiopathic disease. The subdivisions into acute and chronic, internal and external, congenital and acquired, are made according to no one standard, but according to several—pathological, clinical and embryological. Such subdivisions do not clarify the pathogenesis, but serve to obscure it. Chronic internal hydrocephalus, whether congenital or acquired, is the most important and frequent form encountered.

Internal hydrocephalus is characterized by a progressive accumulation of cerebrospinal fluid in the ventricles, causing their dilatation and a consequent cortical atrophy and, when possible, enlargement of the head. The disease is usually fatal; spontaneous recovery, however, does occur in a small percentage of cases.

Numerous forms of treatment have been suggested and tried, but, as the number of methods indicates, they have been almost uniformly unsuccessful. The etiology being so obscure, any treatment is necessarily empirical and consequently unsatisfactory. Successful therapy must depend on the identification and the treatment of the cause of the disease.

It is evident that internal hydrocephalus is due to an abnormality either in the formation or in the absorption of cerebrospinal fluid or possibly in both. Our studies—experimental and clinical—have been concerned with the development, the pathology and the diagnosis of internal hydrocephalus.

2. HISTORICAL

Reference to hydrocephalus is made by the earliest medical writers. Hippocrates is credited with suggesting surgical treatment by trephining the anterior part of the skull. He evidently thought that the accumulation of fluid was extracerebral.

Galen was the first to give special consideration to this disease, which played a conspicuous part in his theory of the “animal spirit.” Galen was really advanced in his knowledge of the anatomy of the ventricles of the brain. He thought they were in free communication with one another and that they formed a closed system. He knew of the aqueduct of Sylvius and of the foramina of Monro. Galen, however, believed that the soul or “animal spirit” was contained in the ventricles and that here it underwent a process of purification; the purified products were supposed to pass into the pores of the brain and the waste products found their way through the pituitary body and were discharged into the nose as “pituita.” He considered hydrocephalus due to some defect in this process of elaboration of the “animal spirit.”

The teachings of Galen were accepted without question until Vesalius, in 1543, denied the existence of the “animal spirit.” Following Vesalius a succession of distinguished anatomists have been interested in the study of hydrocephalus. Among them have been Willis, Sylvius, Rhazes, Celsius, Petit, Pacchionis, Brunner, Littre, Morgagni, Cotugno, Monro, Haller, Robert Whytt, and in the nineteenth century Magendie, John Hilton, Luschka and Key and Retzius.

Many theories regarding the content of the ventricles have been considered since the overthrow of Galen’s theory of the “animal spirit.” It has at various periods been regarded as water, air, vacuum, vapor, until finally it was proved to be a fluid. Verduc, about 1700, insisted that fluid was never present in the normal ventricles, and this agitation led to Haller’s vapor theory. Haller had the advantage of a correct knowledge of the circulation of the blood and supposed the vapor to be exhaled by the arteries and inhaled by the veins.

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Cotugno (1770) first proved the existence of the subarachnoid space and in addition found fluid in this space in living fishes and turtles, but was unable to demonstrate fluid in dogs because the spinal cord so closely filled the dural envelope. Though he was the real discoverer of the existence of cerebrospinal fluid in the living animal, his findings were not accepted because of the firm belief in Haller’s vapor theory. At this time all fluid was explained on the basis of some pathological process or as a post-mortem condensation of the vapor.

Galen’s teaching that the pituitary body was the portal of exit of the ventricular contents was held by many until the end of the eighteenth century. Haller denied this function to the pituitary body, but Petit (1718) and even Monro (1798) supposed that hydrocephalus was due to sclerosis of the pituitary body, which effectually closed the channels of exit from the ventricles.

Monro (1798), after whom the foramen of Monro is named, was also interested in the study of hydrocephalus. The presence of a foramen (the foramen of Magendie), leading from the fourth ventricle to the subarachnoid space as claimed by Haller and Cotugno, was denied by many, including Monro. He said:

The bottom of the fourth ventricle has no such communication with the cavity of the spinal marrow as Dr. Haller supposed, being completely shut off by its choroid plexus and pia mater. As further proof that the four ventricles communicate with each other and that they do not communicate with the cavity of the spinal marrow, I have observed in the bodies of every one of fifteen children who died from internal hydrocephalus that all the ventricles were distended; that on cutting into one of the lateral ventricles, all the ventricles were emptied, that in these cases, the passages above described were greatly enlarged, and that in none of them was water contained in the cavity of the spinal marrow or between its pia and the dura mater.

So near to the cause of hydrocephalus, in his zeal to prove a closed foramen, Monro, unfortunately, mistook it for the normal condition and left the discovery of the communication between the ventricles and the subarachnoid space to Magendie.

Without doubt, Magendie’s contribution is the most important that has been made to the subject of hydrocephalus. He demonstrated by experiments on animals (1) that fluid normally fills the ventricles and the subarachnoid space; (2) that free communication exists between the ventricles and the subarachnoid space by means of a foramen which now bears his name; (3) that the central and spinal subarachnoid cavities form a single freely communicating space, and (4) that the aqueduct of Sylvius or the foramen of Magendie was obstructed in several cases of hydrocephalus.

Magendie, however, did not understand why hydrocephalus should result from an obstruction, for he thought the pia secreted the cerebrospinal fluid. He was led to believe that in some way the fluid could readily make its way upward through these membranous obstructions, but for some reason which he did not understand, its return was impeded and accumulation in the ventricles resulted.

The existence of cerebrospinal fluid has since been admitted, but the other observations of Magendie have been opposed. The controversy over Magendie’s various claims and the views of more recent workers will be considered later.

3. INTERNAL HYDROCEPHALUS EXPERIMENTALLY PRODUCED

Flexner has noted that internal hydrocephalus sometimes follows the injection of the meningococcus into the subarachnoid space of monkeys. With this exception, we have been able to find no instance of hydrocephalus experimentally produced. The more common pathological processes producing internal hydrocephalus are usually so large (tumors) or so diffuse (inflammations) that it has been difficult to determine their exact part in the production of this disease. It is obvious that if hydrocephalus can be produced by experimental means, it will be possible to obtain definite information regarding its cause.

We conducted two series of experiments. In one, the aqueduct of Sylvius was occluded and in the other, the vein of Galen or the straight sinus or both were ligated. In each series the experiment was such that the function of either the aqueduct or of the vein was not disturbed.

I. EFFECT OF OCCLUSION OF THE AQUEDUCT OF SYLVUS

In this series of experiments, in which an obstructing body was placed in the aqueduct of Sylvius, an internal hydrocephalus invariably resulted. It should be emphasized that the obstructing body was so placed in the aqueduct that the topographical relations were undisturbed and the lumen of the vein of Galen unaffected. The resulting hydrocephalus was therefore due solely to the mechanical occlusion of this channel. It is preferable to use a small obstructing body and depend on the formation of adhesions gradually to produce total occlusion. When this is done, practically no postoperative irritative effects result. A small pledget of cotton proved most efficient as the obstructing body.

These experiments were performed most successfully on dogs. Cats and monkeys were tried, but without success. When carefully done, the operative mortality in dogs was negligible. The animals at the time of operation were from 2 to 6 months of age. At this age the sutures of the skull are united so