3.6 Developmental and Acquired Anomalies

3.6.1 Hydrocephalus in Adults

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3.6.1.1 Basics

Adult hydrocephalus is a syndrome that is not yet fully understood. For almost a century Dandy’s observations in dogs paved the way to hydrocephalus being referred to as obstructive or communicating. Our knowledge of hydrocephalus has developed stepwise, based on fundamental inventions in diagnostic procedures such as pneumoencephalography, cisternography, computed tomography (CT), and magnetic resonance imaging (MRI).

While childhood hydrocephalus acts on a developing brain with soft and growing borders, in adults space within the skull is fixed, and the relationship among brain tissue, cerebrospinal fluid (CSF), and blood changes physiologically as a reaction to changing brain volume caused by ischemic or traumatic damage, toxic or degenerative atrophy. The adult brain may be more vulnerable due to co-morbidities such as hypertonic small vessel disease, multi-infarction syndrome, white matter lesions, and gliotic fibrosis. Many degenerative states and diseases of the brain can result in hydrocephalus. Thus, adult hydrocephalus may not be a disease per se, but a multi-etiological clinical entity as a result of chronic changes in an aging or damaged brain.

Hydrocephalus is one of the few almost completely reversible causes of dementia and gait disturbance in the elderly. Adult hydrocephalus markedly impairs mobility of affected persons. Falls and other problems associated with gait disorders, and the enforced immobility that ensues, are a major source of preventable morbidity and mortality.

Adult hydrocephalus is usually characterized by enlarged ventricles, compared with normal or compressed external subarachnoid spaces. However, normal ventricular size is no guarantee that hydrocephalus will not occur. CSF pressure in adult hydrocephalus may be elevated, but can be within normal ranges or even low. Ventricular enlargement in the presence of “normal” CSF pressure seems to be related to periodic wave-like changes in CSF pressure.

In occlusive or obstructive hydrocephalus CSF flow is impaired, thus producing an increase in ventricular pressure and ventricular size. In communicating hydrocephalus the relationship between CSF production and CSF resorption is impaired. While CSF production is almost constant, CSF resorption seems to be subject to change, owing to many states, resulting in elevated resistance to CSF outflow.

Normal pressure hydrocephalus (NPH) is usually referred to as being first described by Hakim and Adams in 1965. NPH can be idiopathic (iNPH), typically beginning in the sixth decade of life or secondary (sNPH), which may be a result of subarachnoid hemorrhage, brain injury, meningitis, posterior fossa surgery or tumors, including carcinomatous meningitis at any age.

Synonyms are Hakim–Adams syndrome, low-pressure hydrocephalus, malresorptive hydrocephalus, aresorptive hydrocephalus, and adult-onset hydrocephalus.

3.6.1.2 Diagnostic Procedures

Proper diagnosis is both fundamental and difficult, for treatment is not without risk. Hydrocephalus due to pure atrophy (hydrocephalus ex vacuo) will not respond to a shunt. After the intervention of shunting procedures in NPH patients, many patients with dementia of other origin were shunted, resulting in severe complications. Therefore, the aim of diagnostic studies is to find the optimal surgical procedures.

There is no commonly accepted standard for diagnosis at the moment. Usually, NPH is diagnosed by a combination of clinical signs and radiological findings.

3.6.1.3 Clinical Signs

The most important clinical signs are the so-called Adams–Hakim Triad: gait disturbance (shuffling), which usually precedes the other symptoms; cognitive deficit; and urinary incontinence. As these are common symptoms of many age- and degeneration-related diseases of the brain, it is often not possible to differentiate hydrocephalus from senility, Alzheimer’s disease, and other
forms of dementia. The combination of NPH and other diseases is common. Co-morbidity may complicate diagnosis, but it is not a negative prognostic factor per se.

Gait disturbance is the most important sign and should preferably be the first one to appear. Gait disturbance is referred to as the stretching of the fibers of the pyramidal tract caused by ventricular enlargement. Reduced gait velocity, owing to a diminished and highly variable stride length, may be found in both Parkinson's disease and NPH. It may be distinguished by a broad-based gait pattern with outward rotated feed and diminished height of the steps, which is typical of NPH. External cues will only mildly improve gait in NPH as opposed to good effects being obtained in Parkinson's disease. Unsteadiness and multiple steps on turning is another typical pattern in NPH.

Differential diagnosis in cognitive deficits and urinary incontinence may be even more difficult. Many patients with NPH are at an age at which they suffer from diseases that may result in the same symptoms.

Dementia in NPH usually starts with memory impairment, followed by impaired wakefulness, bradypnea (mental slowness), and bradykinesia.

Urinary incontinence takes place often unwittingly and may be exacerbated by gait disturbance, preventing the patient from reaching a toilet in time. Fecal incontinence is rare.

Headache, dizziness, vertigo, and psychiatric disturbances may also be symptoms of NPH.

### 3.6.1.4 Radiological Findings

Computed tomography and MRI allow for the evaluation of ventricular enlargement and brain atrophy, but diagnosis should not be based on these procedures alone.

Typical signs of hydrocephalus are said to be symmetrical communicating quadri-ventricular enlargement and flow void in the aqueduct of Sylvius. Ventricular enlargement may be detected as rounding of the frontal horns, enlargement of temporal horns ≥2 mm on both sides and upward bulging of the corpus callosum in MRI. A discrepancy between an enlarged third and a normal fourth ventricle is often seen. Cortical sulci are usually compressed apically and in the interhemispheric fissure, whereas the Sylvian fissure may be enlarged. Ventricular enlargement is calculated by various indices, e.g., Evan’s ratio >0.3. Evan’s ratio means the ratio of distances between the frontal horns of the lateral ventricles to the maximum biparietal diameter measured on axial slices in CT or MRI at the level of the foramen of Monro. Periventricular lucency (low density on CT, high intensity on T2-weighted MRI) adjacent to the frontal and temporal horns is often said to be a sign of hydrocephalus as transependymal CSF absorption or bulk flow, but has no significant value. Neither cortical or subcortical atrophy nor signs of cerebrovascular disease per se does not preclude NPH. Multiple white matter lesions as result of cerebrovascular disease, on the other hand, may be a negative prognostic sign.

Magnetic resonance imaging cine flow studies show unspecific CSF flow phenomena in the aqueduct of Sylvius, which some authors believe to be of prognostic value for treatment. Most important in CT and MRI findings is the exclusion of severe brain atrophy. When clinical signs and radiological findings alone do not allow for diagnosis, CSF pressure and dynamics should be evaluated.

#### 3.6.1.5 Spinal Tap

Withdrawal of CSF is most often used to further evaluate the patient.

Normal CSF pressure in NPH is 5–15 cm H_2O in a lying position. Higher values are usually found in hydrocephalus of other origins. Withdrawal of CSF (spinal tap) of 40–50 ml of CSF is performed via lumbar puncture. The patient is examined before and several hours after spinal tap. Improvement in walking ability and consciousness is thought to give proof of CSF absorption disturbance-related hydrocephalus. If the result of this test is not clear, either it can be repeated after some time (days), or a temporary spinal catheter may be inserted and CSF drained at 500–1,000 ml/72 h, thus simulating the effect of shunting. The effect of CSF drainage has to be evaluated clinically for at least 3 days in order to achieve reliable results.

#### 3.6.1.6 Continuous CSF Pressure Monitoring

Invasive methods to give objective criteria for CSF pressure are continuous intracranial pressure (ICP) monitoring via ventricular drainage or intraparenchymal sensors. Epidural pressure recording is less invasive, but gives less reliable values, which is especially true of absolute pressure values. The pressure monitoring has to be recorded for at least 72 h to be able to compare ICP values and pressure curves for at least two nights, as the first night after anesthesia is often not representative. During day time there may be too many artifacts due to activities of daily life and nursing procedures. The records are evaluated for absolute values of ICP and for specific breath- and pulse-dependent changes in ICP. Sometimes, pressure peaks at ≥20 cm H_2O and typical wave forms can be recorded. The time span of B-waves is recorded and set in relation to the recorded time. As B-waves seem to be associated with REM phases during sleep, up to 20% of the time is regarded as normal; if they are present more than 50% of the time, hydrocephalus is presumed.