Chapter 113

High Cerebrospinal Fluid Pressure

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IDIOPATHIC INTRACRANIAL HYPERTENSION (OR PSEUDOTUMOR CEREBRI)

International Headache Society (IHS) code and diagnosis: 7.1.1 Headache attributed to idiopathic intracranial hypertension

ICD-10 code and diagnosis: G44.82 Headache associated with other intracranial disorders

Short description: Idiopathic intracranial hypertension (IIH), or pseudotumor cerebri, is defined as increased intracranial pressure (ICP) in the absence of an intracranial mass or hydrocephalus. The syndrome is characterized by headache, papilledema, no localizing neurologic signs, and normal cerebrospinal fluid (CSF) composition.

Other terms: Serous meningitis, pseudotumor cerebri, meningeal hydrops, otitic hydrocephalus, toxic hydrocephalus, intracranial hypertension without ventriculomegaly, and benign intracranial hypertension. Because the condition does not always run a self-limiting course, but is often a chronic disorder or leaves the patient with variably severe visual loss, the term benign is certainly a misnomer, and the term *idiopathic intracranial hypertension* is preferred.

Epidemiology

Idiopathic intracranial hypertension (IIH) is a relatively common neurologic disease that has been reported from most parts of the world with an annual incidence of 1 to 2 per 100,000. In the 1980s, the annual incidence in the general population was reported to be 0.9 per 100,000 persons in Iowa and Louisiana in the United States (19) and 1.7 per 100,000 in northeastern Libya (51). The syndrome occurs in both children and adults. No sex difference is seen in children, whereas the female:male ratio in most series of adult patients is between 3:1 and 10:1 (16,19,51,71). The condition occurs most commonly in obese women of childbearing age, in whom the incidence may be at least 19 per 100,000 (19). Familial occurrence has been encountered in siblings and in mother and son (7,54,68). The incidence of IIH seems to parallel that of obesity in a given population (6) and is thus increasing in Western countries. Recent studies in the United States suggest that the incidence of IIH has doubled over the past decade (27,39).

Pathophysiology and Etiology

The pathophysiology of intracranial hypertension is controversial. In the absence of a space-occupying lesion or hydrocephalus, a number of mechanisms have been suggested as possible explanations of increased ICP: increased brain volume caused by increased brain water content (30), increased blood volume (15), increased ICP transmitted from elevated intra-abdominal and intrathoracic pressure in obese persons (65), increased rate of CSF formation (17), or a decreased rate of CSF absorption at the site of the arachnoid villi (42). Only insignificant increments in cerebral blood volume have been reported by some investigators studying IIH (1), and a normal CSF production rate was found using magnetic resonance (MR) techniques for indirect calculations of CSF production (29).

The two prevailing hypotheses of the pathophysiologic mechanisms of IIH are increased brain water content or increased resistance to CSF outflow. While intracellular or interstitial edema is an attractive hypothesis to explain IIH phenomenology, no confirmatory evidence of brain edema exists by either MR imaging (MRI) or autopsy studies (3,70). Increased resistance to CSF outflow across the arachnoid villi is proposed as the underlying cause of IIH (42), and many investigators have convincingly demonstrated an increased resistance to CSF outflow using spinal infusion or perfusion tests (28,61). Unfortunately, abnormal spinal infusion tests do not differentiate clearly between impairment of CSF absorption and decreased intracranial compliance resulting from an increase in brain volume. These two hypotheses are not