Clinical Manifestations

The factors that influence the clinical picture in hydrocephalus are the time of onset, acuity of onset, and associated structural malformations. In infancy, before closure of the cranial sutures, head enlargement (increasing occipitofrontal circumference) is the predominant sign, but in older infants and children, the lesions responsible for hydrocephalus produce other neurologic signs through pressure on adjacent structures.

In infants with hydrocephalus, the head grows at an abnormal rate, although the first signs may be bulging fontanels. The anterior fontanel is tense, often bulging and non-pulsatile. Scalp veins are dilated, especially when the infant cries. With the increase in intracranial volume, skull bones become thin and the sutures become palpably separated to produce a cracked-pot sound (Macewen sign) on percussion of the skull. In severe cases, infants display frontal protrusion (frontal bossing) with depressed eyes and the eyes may be rotated downward (setting-sun sign). Pupils are sluggish, with unequal responses to light.

The signs and symptoms in early to late childhood are caused by increased ICP, and specific manifestations are related to the focal lesion. Most commonly resulting from posterior fossa neoplasms and aqueduct stenosis, the clinical manifestations are primarily those associated with space-occupying lesions (e.g., headaches on awakening with improvement after emesis or being in an upright position, strabismus, ataxia).

Diagnostic Evaluation

Hydrocephalus in infants is based on head circumference that crosses one or more percentile line on the head measurement chart within 2 to 4 weeks and on associated neurologic signs that are progressive. In evaluation of a preterm infant, specially adapted head circumference charts are consulted to distinguish abnormal head growth from normal rapid head growth. The primary diagnostic tools to detect hydrocephalus in older infants and children are CT and MRI. Diagnostic evaluation of children who have symptoms of hydrocephalus after infancy is similar to that used in those with suspected intracranial tumor. In neonates, echoencephalography is useful in comparing the ratio of lateral