for more cases of optic atrophy in the adult population than any other cause. In young patients with inherited optic atrophy, Leber hereditary optic neuropathy is often the cause; it is usually bilateral. *Foster-Kennedy* syndrome is optic atrophy in one eye with papilledema in the other eye, secondary to a tumor compressing the atrophied optic nerve and causing raised intracranial pressure to produce papilledema in the opposite eye.

Ischemic optic neuropathy occurs in two forms. The *atherosclerotic* variety occurs mostly between the ages of 50 and 70 years, and no evidence of systemic disease is present. The *arteritic* form is usually a manifestation of giant cell arteritis; there may be systemic manifestations of the disease, including headache, scalp tenderness, and generalized myalgias. Laboratory evaluation shows anemia and elevated erythrocyte sedimentation rate in almost every case. Patients with arteritis should be treated with high doses of corticosteroids to prevent permanent loss of vision.

Acute transient monocular blindness is usually the result of embolization to the central retinal artery from an atheromatous plaque in the carotid artery *(amaurosis fugax)*. Any complaint of transient visual loss constitutes an emergency, and steps must be taken to prevent permanent loss of vision by making a prompt diagnosis and initiating appropriate therapy. Examples of sight-saving procedures include corticosteroid therapy for cranial arteritis, reduction of intraocular pressure for acute glaucoma, and carotid surgery, anticoagulation, or antiplatelet therapy for embolic cerebrovascular disease.

Binocular Visual Loss

Gradual bilateral visual loss caused by optic nerve lesions is rare. Causes include Leber hereditary optic neuropathy and a toxic nutritional–deficiency state. Acute transient bilateral visual loss (visual obscuration) may be a symptom of raised intracranial pressure caused by a brain tumor or idiopathic intracranial hypertension (IIH); papilledema is often severe. IIH, formerly known as *pseudotumor cerebri,* requires prompt investigation and treatment to prevent potential bilateral visual failure. It is often associated with a high body mass index (BMI) and is more common in young females. Vitamin A and tetracycline ingestion have been associated with the condition. Unilateral or bilateral lateral rectus palsy may be present. It is one of the few situations in which, after imaging, performance of a lumbar puncture is safe in the setting of marked bilateral papilledema. Cerebral venous sinus thrombosis may mimic IIH and should be screened for with neuroimaging.

Bilateral damage to the optic radiations or visual cortex results in cortical blindness. The pupillary light reflex is normal, as are the funduscopic examination findings, and the patient may occasionally be unaware that he or she is blind *(Anton syndrome)*. Patients are often misdiagnosed as having a conversion reaction. Transient cortical blindness occurs most often in basilar artery insufficiency but is also seen in hypertensive encephalopathy. Positive visual phenomena (e.g., phosphenes, scintillating scotomas) are characteristic of migrainous aura and probably reflect oligemia to the occipital lobes from vasoconstriction. Arteriovenous malformations, tumors, and seizures may produce similar symptoms and should be distinguished from migraine with aura by a careful history and examination as well as by imaging in appropriate cases.

Visual hallucinations are visual sensations independent of external light stimulation; they may be either simple or complex, may be localized or generalized, and may occur in patients with a clear or clouded sensorium. Visual illusions are alterations of a perceived external stimulus in which some features are distorted. The simplest visual phenomena consist of flashes of light (photopsias), blue lights (phosphenes), or scintillating zigzag lines, which last a fraction of a second and recur frequently or which appear to be in constant motion. These can arise from dysfunction within the optic pathways at any point from the eye to the cortex. Glaucoma, incipient retinal detachment, retinal ischemia, or macular degeneration can cause simple visual hallucinations based on dysfunction in the eye. Lesions of the occipital lobe are often associated with simple hallucinations; classic migraine is by far the most common condition of this type. Complex visual hallucinations such as seeing objects as people, animals, landscapes, or various indescribable scenes occur most frequently with temporal lobe lesions or parieto-occipital association areas. Visual hallucinations of epileptogenic origin are typically stereotyped.

HEARING AND ITS IMPAIRMENTS

Symptoms of Auditory Dysfunction

The main symptoms of lesions within the auditory system are hearing loss and tinnitus. Hearing loss can be classified as conductive, sensorineural, mixed, or central, based on the anatomic site of pathology (Figs. 112-7 and 112-8). Tinnitus can be either subjective or objective. Conductive hearing loss results from lesions involving the external or middle ear. Patients with a conductive hearing loss can hear speech in a noisy background as well as in a quiet background, because they can understand loud speech as well as anyone. The ear often feels full, as if it is blocked. The Weber test localizes to the deaf ear, if the deafness is unilateral.

Sensorineural hearing loss usually results from lesions of the cochlea or the auditory division of the vestibulocochlear (eighth cranial) nerve. Patients with sensorineural hearing loss often have difficulty hearing speech that is mixed with background noise and may be annoyed by loud speech. They usually hear low tones better than high-frequency ones. Distortion of sounds is common with sensorineural hearing loss. Central (retrocochlear) hearing disorders are rare and result from bilateral lesions of the central auditory pathways, including the cochlear and dorsal olivary nuclear complexes, inferior colliculi, medial geniculate bodies, and auditory cortex in the temporal lobes. Damage to both auditory cortices may result in pure word deafness, in which patients are selectively unable to discriminate language but may be able to hear nonverbal sounds.

Tinnitus is the perception of a noise or ringing in the ear that is usually audible only to the patient (subjective), although, rarely, an examiner can hear the sound as well. The latter, so-called *objective tinnitus,* can be heard when the examining physician places a stethoscope against the patient's external auditory canal. Tinnitus that is pulsatory and synchronous with the heartbeat suggests a vascular abnormality within the head or neck (see Fig. 120-7). Aneurysms, arteriovenous malformations, and vascular tumors can produce this type of tinnitus.