with apnea. The breathing waxes from breath to breath in a smooth crescendo and then, once a peak is reached, wanes in an equally smooth decrescendo. CSR implies bilateral dysfunction of neurologic structures usually lying deep in the cerebral hemispheres or diencephalon, but rarely as low as the upper pons (414).

During CSR, the size of the pupils fluctuates. The pupils dilate in the hyperpneic phase unless there is a concomitant sympathetic nerve paralysis, and they constrict during the apneic phase unless there is a concomitant oculomotor nerve paralysis (415). The cyclic breathing and pupillary movement also are associated with cyclic changes in the level of consciousness. During the apneic phase, the patient slips into a deeper coma; in the hyperpneic phase, the patient may become agitated. In humans, the mydriasis of the hyperpneic phase is almost, but not entirely, blocked by topical sympathetic drugs (e.g., 5% guanethidine) and does not occur when there is an associated or unrelated Horner syndrome. This suggests that neural activity, mediated by the peripheral sympathetic pathway to the eye, plays an important role in the dilation. This suggests, in turn, that the mechanism of the mydriasis of the agitated phase of CSR may be similar to that of reflex pupillary dilation and related to the intermittent partial arousal of a semicomatose patient.

**DISORDERS OF ACCOMMODATION**

Abnormalities of accommodation usually are acquired and occur most frequently as part of the normal aging process (presbyopia). However, disturbances of accommodation also may occur in otherwise healthy persons, in persons with generalized systemic and neurologic disorders, and in persons with lesions that produce a focal interruption of the parasympathetic (and rarely the sympathetic) innervation of the ciliary body. Also, accommodative function can be voluntarily disrupted.

**ACCOMMODATION INSUFFICIENCY AND PARALYSIS**

**Congenital and Hereditary Accommodation Insufficiency and Paralysis**

Congenital defects are a rare cause of isolated accommodation insufficiency. The ciliary body is defective in a number of congenital ocular anomalies, but in most cases vision is so defective that an inability to accommodate is never noted by either the patient or the physician. Aniridia and choroidal coloboma cause obvious defects of the ciliary body. Ciliary aplasia can occur in well-formed eyes in which the iris is intact and reacts normally to light.

Sédan and Roux described three brothers who could see normally for distance without glasses but required +4.00 spherical lenses for reading (416). None of the children had pupillary constriction during near viewing, although their other ocular functions were normal. The children’s retinoscopic findings were not reported, but neither atropine nor physostigmine influenced the state of their accommodation. Aplasia of the ciliary body was the presumed congenital defect. In another family of 10 affected members, an accommodative defect was present in infancy and thereafter nonprogressive by history (417). Pharmacologic assessment with various topical agents suggested a difficulty with either the ciliary musculature or the lens of the affected eyes.

Congenital absence of accommodation has been noted in combination with congenital mydriasis. In three such cases, patent ductus arteriosus was an associated anomaly (43). Defective accommodation was noted in 21 of 78 (27%) dyslexic children, suggesting an association between the two disorders (418).

**Acquired Accommodation Paresis**

**Isolated Accommodation Insufficiency**

Accommodation insufficiency refers to an accommodative ability that measures below the minimum for the age of the patient. Most clinicians use the near point of accommodation as their diagnostic criterion for accommodation insufficiency (accommodative amplitude that is 2 diopters or more below the age-appropriate minimum). Isolated accommodation insufficiency occurring in otherwise healthy eyes can be divided into two groups: (a) static insufficiency and (b) dynamic insufficiency (419).

Static accommodation insufficiency is an inadequate response of either the lens or the ciliary muscle, despite normal ciliary body innervation and neural function. It usually occurs gradually from changes occurring in either the lens or the ciliary body. The most common cause of isolated static insufficiency is presbyopia. In some patients, however, there is sudden loss of accommodation that does not recover. Treatment consists of appropriate spectacle correction.

Dynamic accommodation insufficiency occurs in patients who have inadequate parasympathetic impulses required to stimulate the ciliary musculature but have normal pupil size and reactivity. Such patients usually are asthenopic persons who become ill, often hospitalized, with some unrelated condition. Dynamic accommodation insufficiency also may occur in otherwise healthy young individuals, particularly in children with nonspecific viral illnesses (420,421). The transient loss of accommodation that can occur just before or after childbirth may be another example of this phenomenon (422). Raskind listed various systemic disorders associated with an acquired accommodation insufficiency (423). In all these cases, it is likely that the accommodation insufficiency represents a nonspecific manifestation of the systemic disorder.

The symptoms of dynamic accommodation insufficiency are asthenopia, tiring of the eyes sometimes associated with brow ache, irritation and burning of the eyes, blurred vision particularly for near work, inability to concentrate, and photophobia. As a general rule, symptoms resolve and accommodation recovers following treatment of the underlying illness and restitution of the patient’s former state of health. If accommodation insufficiency remains, the prescription of convex (plus) lenses is indicated, regardless of the patient’s...
age. In patients with an associated convergence insufficiency, convergence exercises or base-out prisms added to the near correction may be of benefit.

**Accommodation Insufficiency Associated with Primary Ocular Disease**

Iridocyclitis may cause profound dysfunction of the ciliary body. In the acute stage, there may be ciliary spasm and loss of accommodation. In the chronic stage, atrophy of the ciliary body results in accommodation insufficiency. The more severe the uveitis, the more commonly mydriasis and cyclopia (internal ophthalmoplegia) are associated with it. In addition, viruses such as herpes zoster may produce a uveitis associated with a ciliary ganglionitis, resulting in a tonic pupil syndrome.

Glaucoma in children or young adults causes accommodation insufficiency from secondary atrophy of the ciliary body. The drugs used in the management of glaucoma affect the ciliary body as well as the iris. In patients who still are able to accommodate, miotic drugs frequently produce ciliary spasm with symptoms of blurred vision.

Metastases to the suprachoroidal space may produce cyclopia and pupillary dilation from damage to the ciliary neural plexus. Lymphoma and carcinoma of the breast, lung, or colon are the most common tumors that produce this condition.

Internal ophthalmoplegia associated with contusion of the globe was discussed in the section above on the pupil. In most cases, when accommodation is paralyzed, the pupil is dilated and fixed. Recovery of accommodation is common, but full recovery of pupillary function is less likely. On rare occasions, the pupil is spared or recovers fully but the ciliary muscle remains paralyzed. Patients who “accidentally” notice the inability to accommodate in one eye should be questioned specifically for previous trauma to that eye (11).

Immediately following a concussion injury to the globe, the pupil is small and accommodation is spastic. Subsequently, the pupil dilates and the ciliary muscle becomes paretic. Accommodation usually returns in 1–2 months. The traumatic etiology of the accommodation paresis may be suspected by slit-lamp biomicroscopic observation of tears in the iris sphincter, tears at the root of the iris, or recession of the angle of the anterior chamber with posterior displacement of the ciliary attachment. There also may be associated ocular hypotension or glaucoma. Following trauma to the globe, rupture of zonular fibers with partial subluxation of the lens also may produce loss of accommodation.

Iatrogenic trauma to the eye, such as that which occurs during retinal reattachment surgery, cryotherapy, or panretinal photocoagulation, may injure the ciliary nerves, producing accommodation paresis and mydriasis (424–426). Laser applications at or anterior to the equator and long exposure times are important factors in the development of accommodation paresis following photocoagulation (427). Optic nerve sheath fenestration performed from the lateral approach can damage the short ciliary nerves that penetrate the temporal sclera, causing a postoperative tonic pupil (245). Sector palsy of the iris sphincter has been reported after argon laser trabeculectomy (244). Transient internal ophthalmoplegia also can result from local anesthesia injected into the lids or gums that inadvertently enters into the orbit (see also the section above on Local Tonic Pupils).

**Accommodation Insufficiency Associated with Neuromuscular Disorders**

Some diseases produce myopathic changes in the smooth muscle fibers of the ciliary body, but isolated ocular involvement of this type is rare.

Myasthenia gravis may cause defective accommodation (428,429). Manson and Stern studied patients with MG and patients with unexplained accommodation disturbances (430). All patients were questioned about diplopia, ptosis, and the effects of sustained reading or close work upon their vision. Binocular and uniocular accommodation amplitudes were measured. Of nine patients with MG (six with generalized and three with ocular disease), eight had abnormal fatigue of visual accommodation. The accommodation defect improved rapidly after an intravenous injection of edrophonium hydrochloride (Tensilon).

Botulism nearly always causes an acute accommodation paralysis. The toxins produced by *C. botulinum* interfere with the release of acetylcholine at the neuromuscular junction and in the cholinergic autonomic nervous system. Although several different subtypes of toxin exist and routes of human infection can vary (food-borne, wound, gastrointestinal tract colonization in infants), clinical manifestations are similar. These include progressive descending skeletal muscle weakness, ocular motor palsies, bulbar paralysis, and cardiovascular lability (431). Food-borne botulism produces prominent gastrointestinal symptoms as well.

Almost 90% of patients with botulism of any type complain of blurred vision. Paralysis of accommodation and impairment of the pupillary light reflex are common and early signs of botulism, often appearing suddenly about the 4th or 5th day of the illness. Accommodation is usually more severely affected than pupil function, for unclear reasons (Fig. 16.31). In some cases, accommodative failure is the initial and sole sign of nervous system involvement. In a series of nine patients with food-borne botulism, all patients examined acutely had marked or complete accommodative loss but full mydriasis was present in only one patient, and another patient had sluggish pupillary reactions (328). Recognition of the clinical syndrome of botulism is critical, as respiratory failure can result in mortality rates up to 20% (431).

Tetanus can produce accommodation paralysis. In most cases, the accommodation paralysis occurs in the setting of generalized ophthalmoplegia; however, one patient described had normal eye movements and normally reactive pupils to light stimulation (432).

Myotonic dystrophy frequently produces degenerative changes in the lens, the region of the ora serrata, and the anterior chamber angle. It also may be associated with ocular hypotension. It is reasonable to assume that the ciliary muscle also is affected, because other smooth muscle dysfunction occurs in such patients (433).
The effect of botulism on the elements of the near reflex. The patient was a 33-year-old man with acute food-borne botulism. His symptoms were 4 days of blurred vision, abdominal pain, nausea, and sweating. Top, The patient’s pupils are large and unreactive to light. Middle, Simultaneous recordings of pupil size, accommodation, and convergence to near effort confirm complete paralysis of pupil responsiveness and accommodation, whereas convergence is intact. Bottom, Both pupils constrict following instillation of 0.1% pilocarpine. (Courtesy of Dr. Helmut Wilhelm.)

Accommodation Insufficiency Associated with Focal or Generalized Neurologic Disease

Accommodation paresis may be caused by both focal and generalized neurologic disorders that interrupt the innervation of the ciliary body. Supranuclear lesions can influence the signal inputs to the parasympathetic midbrain nuclei for accommodation, resulting in paresis or even paralysis of accommodation. Such lesions include damage to the cerebral cortex, rostral midbrain, superior colliculus, and possibly the cerebellum (434,435). In these cases, convergence insufficiency usually accompanies the accommodation insufficiency because of cross-coupling between these two systems.

Acute infectious or epidemic encephalitis as well as postinfectious acute disseminated encephalomyelitis (ADEM) such as that which is associated with or follows measles, chickenpox, or other viral infections can cause accommodative paralysis (436). Typically, patients have loss of the near triad (convergence palsy, absence of pupillary near response, failure of accommodation) and normal pupil responses to light. Histopathologic examination in fatal cases shows diffuse perivenous white-matter lesions throughout the cerebral hemispheres and brain stem.

An acute focal lesion of the hemispheres may cause acute bilateral accommodation insufficiency. Specifically, this has been reported with an acute ischemic stroke in the territory of the left middle cerebral artery and an acute hematoma in the left parieto-occipital region (437,438). The issue is whether the accommodation insufficiency is related to a direct effect of these lesions on supranuclear pathways for accommodation or is a nonspecific sequela of a significant focal cerebral lesion. If the former, it remains to be determined whether a lesion in either hemisphere can cause accommodation insufficiency or just a lesion in the left hemisphere.

Acute problems with near vision resulting from an abnormality of accommodation can be one of the earliest symptoms of pressure on the dorsal mesencephalon such as from obstructive hydrocephalus or a pineal tumor. These complaints may appear weeks before the pupil light reaction or ocular motility becomes clinically abnormal. Particularly in previously healthy children and young adults who suddenly lose accommodation, a careful evaluation should be undertaken to exclude lesions in the area of the rostral dorsal midbrain, including the superior colliculus (439,440). Some of these patients also demonstrate a sudden increase of myopia (pseudomyopia) related to accommodation spasm with blurring of distant vision.

Wilson disease is an hereditary disorder of copper metabolism characterized by progressive degeneration of the CNS associated with hepatic cirrhosis. The neurologic syndrome frequently includes rigidity, difficulty speaking and swallowing, and a characteristic tremor of the wrists and shoulders. Ocular findings include a peripheral corneal ring of copper deposition involving Descemet’s membrane (Kayser-Fleischer ring), copper pigment under the lens capsule, and various ocular motor disturbances, including jerky oscillations of the eyes, involuntary upgaze, paresis of upgaze, and slowed saccadic movements. Paresis of accommodation is common (441,442), but the location of the lesion responsible for the accommodation paresis is controversial. Some investigators favor a supranuclear lesion (441,443), whereas others postulate a lesion in the region of the oculomotor nucleus that serves the near response (442).

In contrast to the aforementioned supranuclear lesions that result in bilateral paralysis of accommodation with sparing of the pupil light reflex, lesions of the peripheral oculoparasym pathetic pathway typically result in unilateral paralysis of accommodation and paralysis of the pupillary light reflex in the same eye. This is because the peripheral impulses for pupil constriction and accommodation originate in the same visceral (Edinger-Westphal) nuclei and follow the same peripheral pathway to the eye. Thus, the patient with an acute lesion in the peripheral pathway subserving accommodation will more likely seek medical consultation for the associated mydriasis than the blurred near vision. The evaluation of such an anisocoria was outlined in an earlier section of this chapter. Common types of injury along this oculoparasym pathetic pathway are infection, ischemia, and compression,
resulting in an oculomotor nerve palsy or a tonic pupil syndrome.

Primary and secondary aberrant reinnervation of the oculomotor nerve also can involve the ciliary muscle. Herzau and Foerster described three young patients who had increased myopia during attempted adduction of the affected eye, presumably from aberrant reinnervation (444).

An isolated accommodation paralysis—accommodation paralysis without mydriasis—can be caused by a lesion of the ciliary ganglion or short ciliary nerves. We are unaware of any well-documented cases of this phenomenon.

**Accommodation Insufficiency Associated with Systemic Disease**

Children and adults may develop transient accommodation paresis following various systemic illnesses. In such cases, the accommodation paresis often appears to occur as an indirect complication of the systemic disorder rather than from direct damage to the ciliary body or its innervation. There are, however, certain systemic diseases that produce accommodation insufficiency through direct effects on the ciliary body and lens or on their innervation.

In patients with diphtheria, accommodative paralysis usually is bilateral and occurs during or after the 3rd week following the onset of infection. Recovery is the rule but may take several years (445). Because of regular vaccination, the infection from *Corynebacteria diphtheriae* is now rare in most developed countries. Accommodation paralysis has been reported following injection of diphtheria antitoxin. The mechanism of diphtheritic accommodation paralysis appears to be related to toxin-induced segmental demyelination of peripheral nerves with preservation of axons (446).

Loss of accommodation may occur in patients with diabetes mellitus from several mechanisms (447). For example, accommodation paresis may develop in young patients with previously uncontrolled diabetes who have just begun treatment. Hyperopia and accommodation weakness develop concurrently within a few days after the patient’s blood glucose has been lowered and then gradually return to normal over 2–6 weeks. The mechanisms for the refractive and accommodative changes in diabetes are poorly understood. Sorbitol accumulates in the lens during periods of hyperglycemia, causing it to swell, and the lens appears responsible for the shifts in refraction because these shifts do not occur in aphakic or pseudophakic eyes. The same mechanism may account for accommodation paresis, because lens resiliency probably is decreased from the swelling. Persistent loss of accommodation can occur in patients with both controlled and uncontrolled diabetes mellitus from damage to the parasympathetic innervation to the eye. In this setting, accommodation paresis and mydriasis are due to denervation injury rather than to effects on the lens. Thus, either metabolic or neurologic mechanisms can be responsible for reduced accommodation in patients with this disease.

Lieppman described 12 professional divers who had visual complaints after decompression sickness (448). All of the divers had evidence of severe accommodation and convergence insufficiency that was thought to be caused by a "central" lesion. These findings were reproduced in two rhesus monkeys experimentally subjected to similar hyperbaric conditions.

**Accommodation Insufficiency Associated with Trauma to the Head and Neck**

Theoretically, any cerebral injury could impair the highly complex neurophysiologic system involved in the coordination of the near response. Similarly, abnormal input from the upper posterior cervical roots or contusion to the side of the cervical cord could disturb transmission in the ascending spinotegmental and spinomesencephalic pathways that influence parasympathetic outflow from the Edinger-Westphal nuclei. Symptoms of difficulty with focusing at near and at far, commonly associated with headache and pains about the eyes, are common complaints in patients who have suffered cerebral concussion or craniovertebral extension injuries (449). These vague and ill-defined complaints are most prominent during the first weeks or months after injury.

The prevalence of accommodation dysfunction in patients with head or neck trauma is not exactly defined and may be influenced in part by the average age of patients studied, as older patients likely have presbyopia prior to trauma.

In a series of 161 patients with head injury (average age 29 years), Kowal found that 16% had poor accommodation, 19% had over-accommodation (pseudomyopia), and 14% had convergence insufficiency (450). In about half of these patients, near vision complaints improved or resolved within the first year after their injury. Similarly, among 39 patients with whiplash or indirect injury to the neck, Burke et al. found decreased accommodation and convergence in nine (23%), six of whom were symptomatic (451). Five of these six patients recovered accommodation after 9 months.

Tests of accommodation depend upon an earnest, voluntary effort by a motivated patient. Thus, patients who have cortical deficits, loss of concentration, poor comprehension, excessive somnolence, or pain often perform poorly on accommodative tests. Other patients attempting to gain material or psychological compensation may intentionally perform poorly on these tests (452). The persistence of symptoms for many months or even years is most common in patients who are seeking compensation for their injury through litigation.

**Accommodation Insufficiency and Paralysis from Pharmacologic Agents**

Most topical pharmacologic agents that produce pupillary mydriasis also produce cycloplegia, including atropine, scopolamine, homatropine, eucatropine, tropicamide, cyclopentolate, and oxyphenonium. Various investigators have compared the duration and effectiveness of cycloplegia produced by these agents when used as ocular solutions (453,454). None of these agents causes persistent paralysis of accommodation after discontinuation, although there may be some confusion when loss of accommodation occurs after treatment of a severe viral uveitis (e.g., herpes zoster, varicella) with a cycloplegic agent. In such cases, the accommodation...
paralysis occurs from the effects of the virus on the ciliary ganglion and not from the cycloplegic drug.

When cycloplegic agents or related substances are incorporated in medications that are taken internally or applied to the skin as ointments or plasters, there may be sufficient absorption to produce paresis of accommodation. In such cases, the accommodation deficit is partial and recovery begins shortly after the medication is discontinued.

**Accommodation Paralysis for Distance: Sympathetic Paralysis**

Lesions of the cervical sympathetic outflow may produce a defect that prevents the patient from accommodating fully from near to far, but most reports describe an increase in accommodative amplitude on the side of the Horner syndrome (455). Cogan described an ipsilateral increase in near accommodation in five patients with Horner syndrome and noted an apparent paresis of accommodation in one patient (456).

**ACCOMODATION SPASM AND SPASM OF THE NEAR REFLEX**

**General Considerations**

Accommodation spasm is due to excessive activity of the ciliary muscle that results in an abnormally close point of focus. Clinically, there is an apparent or increased myopia that disappears following cycloplegia (pseudomyopia). Accommodation spasm typically affects both eyes, but unilateral cases have been reported (457). It can occur in isolation as pseudomyopia or in association with convergence spasm and excessive pupillary miosis in varying combinations and degrees, all of which probably represent the spectrum of clinical presentations of spasm of the near reflex (458).

Symptoms of isolated accommodation spasm are blurry vision, especially at distance, fluctuating vision, asthenopia, eyestrain, poor concentration, brow ache, and headaches. The diagnostic finding is a greater myopia on manifest refraction compared with cycloplegic refraction, the difference ranging from 1 to 10 diopters. Additionally, the patient will not accept the majority of the cycloplegic refraction, preferring instead the greater myopic correction for visual improvement.

In addition to the symptoms of accommodation spasm, patients with spasm of the near reflex who have convergence spasm also complain of a horizontal diplopia that often is variable in nature. Because of the diplopia and apparent esotropia, such patients initially may be mistaken as having a unilateral or bilateral abducent nucleus palsy or ocular myasthenia and undergo extensive neurologic and neuroimaging investigations (428,459). Spasm of the near reflex should be suspected in a patient with an apparent unilateral or bilateral limitation of abduction that is associated with severe bilateral miosis (459,460). The diagnosis is confirmed by demonstrating that the miosis resolves as soon as either eye is occluded with a hand-held occluder or patch (461). Additionally, the apparent abduction weakness present on horizontal gaze testing with both eyes open will disappear when the opposite eye is patched (monocular ductions testing) or when the oculocephalic maneuver is performed. Refraction with and without cycloplegia will establish the presence of pseudomyopia as well.

**Accommodation Spasm Unassociated with Organic Disease**

Most cases of accommodation spasm (usually as part of spasm of the near reflex) appear to be nonorganic, being triggered by an underlying emotional disturbance or occurring as part of malingering. In such cases, spasm of the near reflex typically occurs as intermittent attacks lasting several minutes (462,463). The degree of accommodation spasm and convergence spasm in such patients is variable; however, miosis is always present and impressive (Fig. 16.32). However, many young persons, when undergoing a noncycloplegic refraction, can accept increasing degrees of overcorrecting concave (minus) lenses. When these same patients undergo a cycloplegic refraction, they are found to be emmetropic or at least significantly less myopic than they appeared to be when not cyclopleged. However, unlike patients with accommodation spasm who prefer the greater myopic correction, these otherwise healthy young persons prefer their cycloplegic refraction for best-corrected visual acuity.

The management of most patients with nonorganic spasm of the near reflex begins with simple reassurance that they have no irreversible visual or neurologic disorder. In other instances, referral for psychiatric counseling is appropriate. Symptomatic relief may be necessary with a cycloplegic agent and bifocal spectacles or reading glasses. Glasses with an opaque inner third of the lens to occlude vision when the eyes are esotropic have been proposed for the convergence spasm (464). Nonorganic spasm of the near reflex also is discussed in Chapter 27.

**Accommodation Spasm Associated with Organic Disease**

Accommodative spasm has been reported, mostly as single case occurrences, in association with various organic diseases and CNS lesions. These include neurosyphilis, ocular inflammation, Rader paratrigeminal neuralgia syndrome, cyclic oculomotor palsy, congenital ocular motor apraxia, congenital horizontal gaze palsy, pineal tumor, Chiari malformation, pituitary tumor, metabolic encephalopathy, vestibulopathy, Wernicke-Korsakoff syndrome, epilepsy, cerebellar lesions, and acute stroke (465–471).

Several investigators have reported spasm of the near reflex in patients with MG (472,473). Romano and Stark described a 26-year-old man who developed isolated pseudomyopia as a presenting sign of ocular MG (428). The pseudomyopia was thought to have occurred from “substitute convergence” that the patient used to compensate for bilateral medial rectus weakness rather than from true accommodation spasm.

Isolated accommodation spasm and spasm of the near reflex appear to be increasingly recognized as a consequence of head injury (450,474–477). In one series, accommodation...