Chapter 182: Central Vestibular System Disorders

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Many diverse pathologic conditions affect the central nervous system (CNS) or its vascular supply and cause dizziness, vertigo, or imbalance. To differentiate these disorders from those affecting the peripheral labyrinthine system, the otolaryngologist-head and neck surgeon must know the associated signs and symptoms of the CNS processes and the natural history of their appearance. Definitive diagnostic evaluation and treatment usually involve cooperation with neurologists or neurosurgeons. A classification of CNS disorders capable of eliciting vestibular symptoms includes intracranial complications of suppurative otitis, vascular disorders, neoplasms, disorders of the craniovertebral junction, multiple sclerosis, familial ataxia syndromes, and focal seizure disorders.

Intracranial Complications of Otitic Infections

Abscesses in the extradural or subdural spaces and those in the temporal lobe and cerebellum can cause vestibular complaints, as can a suppurative process in the apex of the petrous bone. Chapter 158 considers these conditions and their clinical presentations in detail; Chapter 154 discusses malignant external otitis.

Vascular Disorders

Abnormalities of blood flow to the vestibular system are very common causes of vestibular symptoms and are often difficult to distinguish from well-known end organ disorders. Clinical presentations vary for many reasons. Functional deficits may be restricted to one or both end organs or their parts, may involve portions of vestibular nuclei and their afferent and efferent pathways and connections, or may affect both peripheral and central regions. Diverse pathologic processes can result in either a permanent or temporary loss of function in these areas. For instance, permanent losses typically result from thrombotic or embolic arterial occlusion or from hemorrhagic infarction. Transient effects can accompany arterial stenosis, vascular spasm, inadequate perfusion pressure, or reversed arterial flow with shunting, as seen in the subclavian steal syndrome. Although many symptoms complexes seem possible, certain common clinical entities are discernible.

Migraine

Migraine is a vascular syndrome characterized by periodic headaches (Wolff, 1963). It is estimated to affect nearly 25% of women, 15% of men (Waters and O'Connor, 1975), and 5% of children (Bille, 1962). Approximately 20% of adults with migranie indicate that their headaches began before the age of 10 years (Selby and Lance, 1960). In the prepubertal period, boys with migraine outnumber girls by nearly 2 to 1, but at puberty migraine decreases in boys and increases in girls so that a 2.5 to 1 female preponderance is established by adulthood. Although migraine is said to begin after age 40 in only 10% of patients, the disorder is so prevalent that a substantial number of adults become affected later in life.

The terminology and classification of migraine and other headache disorders has been a controversial issue for decades. The most recent effort, that of the Headache Classification Committee of the International Headache Society (IHS) (1988), shows great promise for acceptance as a classification and set of clinical definitions that will allow meaningful comparison of groups of patients among centers. The IHS headache classification headings are:

1. Migraine

2. Tension-type headache

3. Cluster headache and chronic paroxysmal hemicrania

4. Miscellaneous headaches unassociated with structural lesion

5. Headache associated with head trauma

6. Headache associated with vascular disorders

7. Headache associated with nonvascular intracranial disorder

8. Headache associated with substances or their withdrawal

9. Headache associated with noncephalic infection

10. Headache associated with metabolic disorder

11. Headache or facial pain associated with disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures

12. Cranial neuralgias, nerve trunk pain, and deafferentation pain

13. Headache not classifiable.

Subclassifications of migraine are shown in the box. The most commonly occurring form is *migraine without aura* (1.1), which replaces the term common migraine. Diagnostic criteria for migraine withou aura are also in the box.

Box: IHS classification of migraine

1.1 Migraine without aura
1.2 Migraine with aura
    1.2.1 Migraine with typical aura
    1.2.2 Migraine with prolonged aura
    1.2.3 Familial hemiplegic migraine
    1.2.4 Basilar migraine
    1.2.5 Migraine aura without headache
    1.2.6 Migraine with acute onset aura
1.3 Ophthalmoplegic migraine
1.4 Retinal migraine
1.5 Childhood periodic syndromes that may be precursors to or associated with migraine
    1.5.1 Benign paroxysmal vertigo of childhood
    1.5.2 Alternating hemiplegia of childhood
1.6 Complications of migraine
    1.6.1 Status migrainosus
    1.6.2 Migrainous infarction
1.7 Migrainous disorder not fulfilling preceding criteria

IHS diagnostic criteria for migraine without aura (1.1)

A. At least five attacks fulfilling B-D.
B. Headache attacks lasting 4 to 72 hours (untreated or unsuccessfully treated).
C. Headache has at least two of the following characteristics:
    1. Unilateral location
    2. Pulsating quality
    3. Moderate or severe intensity (inhibits or prohibits daily activities)
    4. Aggravation by waling stairs or similar routine physical activity
D. During headache at least one of the following:
    1. Nausea and/or vomiting
    2. Photophobia and phonophobia
E. At least one of the following:
    1. History and physical and neurologic examination do not suggest one of the disorders listed in groups 5-11
    2. History and/or physical and/or neurologic examinations do suggest such disorder, but it is ruled out by appropriate investigations
    3. Such disorder is present, but migraine attacks do not occur for the first time in close temporal relation to the disorder

IHS diagnostic criteria for migraine with aura (1.2)

A. At least two attacks fulfilling B

B. At least three of the following four characteristics:

1. One or more fully reversible aura symptoms indicating focal cerebral cortical and/or brainstem dusfunction

2. At least one aura symptom develops gradually over more than 4 minutes, or two or more symptoms occur in succession

3. No aura symptom lasts more than 60 minutes. If more than one aura symptom is present, accepted duration is proportionally increased

4. Headache follows aura with a free interval of less than 60 minutes. (It may also begin before or simultaneously with the aura.)

C. At least one of the following:

1. History and physical and neurologic examinations do not suggest one of the disorders listed in groups 5-11

2. History and/or physical and/or neurologic examinations do suggest such disorder, but it is reuled out by appropriate investigations

3. Such disorder is present, but migraine attacks do not occur for the first time in close temporal relation to the disorder.

The other category of migraine relevant to this discussion is *migraine with aura* (1.2) which is described by the IHS as an idiopathic recurring disorder manifesting with attacks of neurologic symptoms unequivocally localizable to cerebral cortex or brainstem, usually gradually developed over 5 to 20 minutes and usually lasting less than 60 minutes. Headache, nausea, or photophobia usually follow neurologic aura symptoms directly or after a free interval of less than an hour. The headache usually lasts 4 to 72 hours, but may be completely absent. In this classification, the term *aura* is used to denote the occurrence in isolation or aggregate of the neurologic manifestations that are experienced as a result of the migraine disorder, whenever they occur, and is not restricted to their occurrence as a prodrome immediately preceding the headache. The diagnostic criteria for the clinical varieties *migraine with aura* are shown in the box on p. 3178.

To be able to ask the appropriate questions to elicit a history adequate to make the diagnosis, one must understand the various symptoms that compose the aura. In descending order of frequency they are (1) scotomata, or blind spots; (2) teichopsia, or fortification spectra, a zigzag pattern in the visual field resembling a fort; (3) flashing (photopsia) or colored lights; and (4) paresthesia.

Visual symptoms are, by far, the most common, and according to Bartelson (1984):

... usually affect both eyes simultaneously but can affect one eye alone. The patient may experience negative phenomena only and describe hemianopia or quadrantanopia, complete blindness, tunnel vision, asymmetric field deficits, monocular blindness, altitudinal defects, or one or more scotomata. More frequently, positive phenomena occur and consist of stars, sparks, unformed flashes of light (photopsia), simple and complex geometric patterns, or jagged zig-zags of lights (teichopsia or fortification spectra). These visual hallucinations are usually white but can be any color. They are typically present with eyes open or closed. The slowly evolving images have a shimmering, flickering quality. The positive and negative

visual phenomena are frequently combined, and the term *scintillating scotoma* is used. The patient may report wavy lines like heat off of pavement or as though looking through rain-covered glass. Visual distortion or misperception, such as micropsia, is uncommon and implies temporal or parietal lobe dysfunction.

The most common sequence is the slow onset of bilateral central scotomata or luminous phenomena, which move slowly in an arc to the periphery of one visual field. The leading edge is a zigzag of light followed by moving geometric patterns, which in turn leave behind expanding homonymous scotomata.

Paresthesias, or somatosensory symptoms, present as a numbness, tingling, or both, which affect the upper extremities, face, lower extremities, or occasionally the trunk. Fisher (1980) suggests "a reliable sign of migrainous paresthesias is the 'march' of numbness as it gradually spreads over the face or fingers and hand and migrates from face to limb or vice versa or crosses the face and hand on the opposite side". He also observes that 15% of patients report the appearance of paresthesias at all sites simultaneously, and that in some patients the paresthesias are very localized; for example, to a single digit, lip, or cheek. These paresthesias are not the result of hyperventilation.

Neurootologic symptoms can also occur as part of the spectrum of the migraine aura, and, in fact, are frequent and sometimes quite prominent or severe. Kayan and Hood (1984) prospectively interviewed and examined 200 consecutive patients with migraine and 116 patients with tension headache and compared the frequency of neurootologic symptoms in the two groups (Table 182-1). They found that some vestibulocochlear symptoms occurred in 59% of the migraine group but only in 30.2% of the tension headache group, significant at $P < 0.001$. While cochlear symptoms were less frequent than vestibular symptoms, cochlear symptoms did not occur at all in patients with tension headaches. The distribution of the individual symptoms is shown in Table 18-2. Neurootologic symptoms are discussed individually.

Vestibular symptoms

Nonspecific dizziness. This symptom occurs frequently in patients with either tension or migraine headaches, and therefore, does not help differentiate between the two groups.

Vertigo. Vertigo is extremely common in migraine and occurred in 26.5% of Kayan and Hood's (1984) group of migraine patients compared to 7.8% of those with tension headache ($P < 0.001$). Also significant ($P < 0.05$) was the difference in severity of the vertigo, with 10 migraine patients (5% of the entire population) experiencing it severely enough to seek medical attention for relief, compared to only one patient (0.86%) in the tension headache group. In a smaller series of patients, Kuritzky et al (1981) studied the occurrence of vertigo not associated with headaches in groups of patients who experienced migraine, tension, or cluster headaches, noting a significant difference ($P < 0.05$) between the migraine group and the other two groups. The difference was attributed to the subset of patients with classic migraine, 42% of whom experienced vertigo disassociated with their headaches.

An awareness of the temporal relationship of the vertiginous spells to the headaches is important because the commonly accepted view that symptoms of the aura immediately precede the headache as a prodrome is not the most common occurrence when vertigo is considered. In fact, in the 53 patients with vertigo and migraine interviewed by Kayan and Hood (1984), the vertigo immediately preceded the headache in only 15%, whereas it occurred during the headache in 47% of patients and during the headache-free interval in 36% of patients. In a series of 50 patients with basilar migraine, Olsson (1991) also found vertigo to occur much more often during the headache-free interval than as a prodrome for the headache.

Motion sickness. Several authors have studied the relationship between migraine and motion sickness. In a classic study of 9000 Swedish schoolchildren, Bille (1962) matched children with more pronounced migraine to a similar group without migraine. Severe motion sickness was present in 49% of the children with migraine and only 10% of the control group. Barabas et al (1983) found that 45% of 60 children with migraine experienced at least three episodes of motion sickness culminating in vomiting, compared with 5% to 7% of three similar-sized control groups of subjects with nonmigraine headaches, seizure disorders, and learning disabilities/neurologic perceptual impairments. These differences were statistically significant ($P < 0.0001$). In adults, Kuritzky et al (1981) noted a significantly greater incidence of motion sickness in patients with classic migraine compared to patients with tension and cluster headaches ($P < 0.05$). In the series by Kayan and Hood (1984) of 200 unselected patients with migraine, 50.7% reported motion sickness compared to 20.1% of the 116 unselected patients with tension headaches.

A small subset of these migraine patients experience such severe motion sickness that it significantly restricts their lives. They curtail their head motion to a minimum, plan their day's activities to minimize movement, and frequently rest during the day, remaining motionless for 30 to 60 minutes until symptoms abate. If they cannot avoid continuous vestibular stimulation, a full-blown migraine attack can be triggered. Caloric responses in these individuals tend to be symmetric and very brisk, often culminating in emesis. If treatment provides a dramatic response, the patients often are surprised at how much they have restricted their lifestyle. Neither the physician nor the patient usually suspects that the migraine has any relationship to the other symptoms. They discuss only the vestibular symptoms and ignore or discount any effects of the underlying problem. When prophylactic migraine therapy is instituted, dramatic improvement often (but not always) results.

Auditory symptoms

Twenty percent of the patients in the study by Kayan and Hoow (1984) complained of hearing loss, tinnitus, or pitch distortion; and one fourth of the 20% had multiple auditory symptoms. In descending order of frequency, these symptoms occurred during the headache, during the headache-free interval, and immediately before the headache, which exactly mirrored the temporal relationship of vertigo to the headache in the same series of patients.

Hearing loss. Many authors have reported isolated patients who have hearing loss that seemed to be related to migraine; this finding was reported in 6.5% in the series by Kayan and Hood (1984). Olsson's careful prospective study of 50 patients with basilar migraine has provided new information (Olsson, 1991). In this select population, 52% of individuals noticed a change in hearing as part of the aura immediately preceding the migraine headache; a fluctuating low-frequency sensorineural hearing loss involving at least the two lowest tested frequencies was documented in over 50% of patients.

Tinnitus. Tinnitus was experienced by 15% of the patients with migraine, none with tension headaches (Kayan and Hood, 1984), and in over 60% of those with basilar migraine (Olsson, 1991).

Distortion. Although distortion was noted by 4% of patients in the series by Kayan and Hood (1984), its actual prevalence may be higher. More attention may be given to more bothersome symptoms and auditory distortion may be ignored as unimportant.

Phonophobia. An aversion to loud noise, as much as any other symptom, distinguishes patients with migraine from those with other types of headache. Only 12% of patients with tension headaches experienced phonophobia, but 81% of those with migraine reported this symptom (Kayan and Hood, 1984) ($P < 0.001$). Olsson (1991) detected phonophobia in basilar migraine patients at a rate of 70% during the headache and 76% during headache-free intervals. Furthermore, he documented an abnormal loudness discomfort level in 78% of these patients, whereas only 14% had an abnormal speech reception threshold.

Migraine associated with neurootologic symptoms

Basilar migraine. Bickerstaff (1961) described a form of migraine similar to what was then called classic migraine in that it consisted of an aura followed by a severe headache. The principal distinguishing feature was that the symptoms of the aura could be ascribed to ischemia in the distribution of the basilar artery. The majority of patients were adolescent girls in whom the migraine attacks often occurred premenstrually. Others verified his observations and emphasized special aspects of the clinical picture such as stupor (Lee and Lance, 1977), loss of consciousness (Lees and Watkines, 1963), neurotologic symptoms (Harker and Rassekh, 1987; Love, 1987), and its occurrence in children (Golden and French, 1975). The IHS Headache Classification Committee selected basilar migraine as the name for this category of patients, replacing basilar artery migraine and posterior fossa migraine. The description of migraine with aura applies to basilar migraine, as do the diagnostic criteria in the box on p. 3178. To be classified as basilar migraine, the episodes must contain two or more of the symptoms listed in the box on the bottom of p. 3179.

Box: IHS symptoms of basilar migraine

Visual symptoms*	Double vision
Dysarthria	Ataxia
Vertigo	Bilateral paresthesia
Tinnitus	Bilateral pareses
Decreased hearing	Decreased level of consiousness

* Involving temporal and nasal fields of both eyes

The pathophysiology of these symptoms reflects dysfunction and presumably ischemia of the brainstem, cerebellum, cranial nerve nuclei, and occipital lobe cortex, the latter receiving its blood supply from the posterior cerebral arteries, which are the terminal outflow of the basilar artery. When neurootologic symptoms of migraine are severe enough that medical attention is sought (as they were in 5% of unselected migraine patients (Kayan and Hood, 1984)), the patients will consult or be referred to otolaryngologisgts. The single-most helpful anamnestic finding is the association, on more than one occasion, of neurootologic symptoms with their symptoms that reflect neurologic dysfunction of the CNS excluding the auditory and vestibular systems. This association can only be identified if all the symptoms in the box are explored in the history of all patients with neurootologic symptoms.

Migraine aura without headache. This category of migraine is the most confusing to many physicians and patients. Often neither group even considers migraine as a possible cause for vertigo because they are conditioned to believe that severe headaches must be present to make this diagnosis.

Whitty (1967) reported a series of patients who had symptoms typically experienced during a migraine aura, but who had no headaches associated with their symptoms. Some reported the same aura symptoms associated with migraine headaches earlier in life, or developed typical migraine headaches with their aura after their initial evaluation. Other patients experienced the aura symptoms alone on some occasions and together with a typical migraine headache on other occasions. Other investigators have described similar patients and used the terms *migraine equivalent* (Brown, 1977; Harker and Rassekh, 1988; Watson and Steele, 1974) or *migraine accompaniments* (Fischer, 1980, 1986). The key to the diagnosis is the elicitation of a history of repeated episodes in which neurootologic symptoms coexist with symptoms typical of a migraine aura. A past history of migraine, motion sickness, premenstrual clustering of attacks, associated headaches with some of the attacks, or a family history of migraine is frequently present in these patients.

Etiology

The time-honored concept that migraine headache results from dilatation of extracranial and dural arteries, causing stretching of sensitive pain fibers in the walls of the arteries, and that the symptoms of the aura reflect ischemia secondary to intracerebral vasoconstriction has considerable supportive research. However, not all the complex phenomena associated with migraine's protean manifestations are so easily explained. There is also considerable support for a neural-based theory (Leao, 1944; Pearce, 1987) and, more recently, a combination of the vascular and neural theories (Welch, 1987).

Abundant evidence also implicates the metabolism of serotonin and other vasoactive amines in pathogenesis. Plasma serotonin levels fall during an attack of migraine, and headaches can be precipitated by reserpine, which releases serotoning from body stores. Intravenous infusion of serotoning can relieve a migraine headache. Stabilization of serotonin neurotransmission by depressing the activity of serotonergic neurons may be the common mechanism of action of several of the effective drugs for migraine. These drugs alter serotonin's effective bioavailability by several known mechanisms, including receptor agonism, prolongation of the biologic half-life, inhibition of release, and activation of cyclic adenosine monophosphate (cAMP). Serotonin receptor sites are present both in intracranial

blood vessels and within nerve terminals throughout the central nervous system (Peroutka, 1990; Raskin, 1990), so the exact site and mechanism of its action in migraine events remain uncertain.

Management

Management of migraine headaches can be divided into two categories: symptomatic and prophylactic (Ziegler, 1983). Certain drugs are useful in ameliorating the symptoms of the acute attack, whereas others are effective in reducing the frequency and severity of attacks or eliminating their occurrence entirely. Symptomatic treatment includes analgesics, antiemetics, antivertiginous drugs, sedatives, and vasoconstrictors to relieve headache, nausea, vomiting, and vertigo, which are the most distressing symptoms. Decreased gastric motility occurs during migraine attacks and can decrease absorption of oral drugs as well as contribute to the nausea and vomiting. Metoclopramide (Reglan) promotes normal gastric motility and may improbe absorption of oral drugs and thus facilitate treatment. For many patients aspirin and rest are adequate to relieve the headache, and many of the combination preparations for symptomatic treatment of migraine contain sedatives that are effective, partially because they enhance sleep. Fiorinal (a combination containing aspirin, caffeine, and butalbital) and Midrin (which contains the sympathomimetic amine isometheptene, the mild sedative dichloralphenazone, and acetaminophen) are examples.

The most effective drug for relief of headache is ergotamine, an alpha-adrenergic blocking agent with direct action on smooth muscle fibers of peripheral and cranial vessels. Ergotamine acts as a vasoconstrictor if the vascular resistance is low but induces vasodilatation if there is increased resistance (Hellig and Berge, 1969). It also affects serotonin turnover in the brain (Sofia and Vassan, 1975) and interacts with a large number of neurotransmitted receptors. The major side effect is severe nausea and vomiting, which may preclude the use of the drug if it can not be improved by metoclopramide. Pregnancy is an absolute contraindication to the use of ergotamine because of its uterine effects. Hypertension, hepatitis, and renal disease are also contraindications for the use of ergotamine preparations.

Ergotamine relieves the acute migraine headache in nearly 90% of patients. It should be given as soon as possible after the onset of symptoms, either the aura in classic or basilar migraine or the headache in common migraine. Ergotamine may be prescribed for oral use (Gynergen and others), sublingually (Ergostat and others), or rectally with caffeine (Cafergot suppositories). Caffeine potentiates ergotamine by increasing its absorption, but it also prevents sleep, which can by itself felieve a migraine attack. The total daily dosage of oral or rectal ergotamine should not exceed 4 to 6 mg/day or 10 mg/week. Vascular occlusion and gangrene have been recorded with higher doses, and the risk of severe "rebound" headache is significant. Ergotamine preparations are generally restricted to persons over 10 years of age but are occasionally used in younger children who have a very well-recognized migraine aura.

When migraine headache is severe and ergotamines cannot be used, the next most effective symptomatic medication is Midrin. It neither aggravates nor induces nausea, and its sedative effects help relieve the tension component of the headache. Usually one to two capsules at the beginning of an attack are followed by one capsule every 1 to 2 hours if necessary. No more than five capsules should be given in 24 hours.

Another drug for aborting the acute migraine attack which is not yet available ni the USA, is the serotonin 5HT1D receptor-agonist sumatriptan (Imitrex). European trials indicate that in 90% of cases the drug is effective at aborting migraine symptoms within 10 to 30 minutes after intravenous administration (Perrin et al, 1989). Used subcutaneously, the drug exhibited the same efficacy within 60 minutes (Baar et al, 1989), and oral dispersible tablets provided relief for 70% to 85% of patients within 2 hours (Perrin et al, 1989; Doenicke et al, 1989). As yet, no serious adverse effects have been noted, making it a much more promising drug than ergotamine for aborting attacks. Because of the chronicity and recurrence of the headaches and the potential addiction liability, narcotics should almost never be used in the treatment of migraine headache.

Prophylactic treatment is often necessary when migraine attacks are frequent or the severity can not be ameliorated by symptomatic medicines.

In most instances, propranolol (Inderal) is the drug of choice to prevent migraine episodes. Multiple clinical studies have demonstrated that approximately 50% to 70% of patients will derive some benefit from prophylactic propranolol therapy (Peroutka, 1990). It is contraindicated by the presence of asthma, congestive heart failure, peripheral vascular disease, diabetes, and hypothyroidism. Patients receiving propranolol may be subject to hypertensive crisis if topical anesthetics containing epinephrine are injected in significant concentrations (Fostor and Aston, 1983). The principal side effects are fatigue and lethary; but weakness, dizziness, insomnia, gastrointestinal symptoms, and weight gain have occasionally been reported. Side effects can be minimized by slowly increasing the dose from a low starting level. Adults usually respond to 80 to 120 mg/day; the most common fist sign of effectiveness is a decrease of the severity of individual attacks or an improved response to symptomatic medication during an acute attack, rather than an actual decrease in the frequency of episodes. A trial of propranolol should be continued for at least 2 to 3 months at the highest level of tolerance before it is considered a failure. Discontinuation should be gradual over several days.

A variety of beta-adrenergic agents also have been used for migraine prophylaxis. Atenolol (Tenormin), metoprolol (Lopressor), nadolol (Corgard), and timolol (Blocadren) appear to be at least as effective as propranolol. However, a number of other beta-adrenergic antagonists (for example, acebutolol, oxprenolol, alprenolol) do not appear to be effective in migraine prophylaxis, and it has been suggested that effective prophylaxis require pure beta-adrenergic antagonism without intrinsic sympathomimetic activity.

Amytriptyline has also been demonstrated to decrease the frequency and severity of migraine attacks. It is especially useful when the episodes are triggered by tension or are closely associated with tension headache. A single nocturnal dose of 50 to 100 mg is usually effective and avoids the major side effects of sedation and anticholinergic activity.

Cyproheptadine (Periactin) can also prevent migraine, probably because of its serotonin-antagonist properties. It also has antiplatelet-aggregation properties that may play a role in its effectiveness. Its use is limited in adults because of the major side effects: sedation and weight gain. These occur in up to 40% of patients but are adequately tolerated by most children. Because of atropine-like effects, the drug is not used in children with asthma or hypertension.

Calcium channel blockers (nifedipine, verapamil) can also reduce the frequency of migraine attacks but are not as yet approaved for use in migraine in the USA. Their effectiveness appears to be slightly less than that of beta-blocking agents (Albers et al, 1989; McArthur, 1983). Side effects such as constipation, orthostatic hypotension, and erythematous swelling of the feet are fairly common. The drugs are contraindicated in pregnancy (Peroutka, 1990).

In those women whose migraines are periodic and premenstrual, naproxen, a nonsteroidal anti-inflammatory agent, or Bellergal, a combination of ergotamine tartrate, phenobarbital, and the parasympathomimetic inhibitor bellafoline, can be taken for the few days each month when the migraine is expected, to prevent its appearance. When Bellergal is chosen, no additional ergotamine-containing preparations should be used because of the danger of ergotamine over-dosage.

In young children, anticonvulsants - both phenyton (Dilantin) and phenobarbital - are effective in preventing migraine. Adverse effects of phenyton include hypersensitivity rash, hirsutism, gingival hyperplasia, nausea and vomiting, and ataxia. Behavioral disturbances and hyperactivity can occur with phenobarbital use.

Vestibular disorders related to migraine

Benign paroxysmal vertigo

Basser (1964) described a clinical entity in children under the age of 4 that he called *benign paroxysmal vertigo.* The physician is usually consulted because the child exhibits spells of peculiar behavior. A completely normal child suddenly becomes frightened; cries out; clings to the parent or staggers as though drunk; and exhibits pallor, diaphoresis, and frequently vomiting. Symptoms are accentuated by head motion, and sometimes nystagmus and torticollis are observed. Many children can describe vertigo or a spinning sensation. The spells last only several seconds to 5 minutes, and afterwards the child is immediately normal again and resumes playing as though nothing happened.

Between spells the child remains entirely normal and has no audiometric abnormalities, unless concomitant middle ear disease is present. Unilateral or bilateral vestibular paresis on caloric stimulation is common. Neurologic examination and the electroencephalogram are normal.

The spells usually begin before age 4 and occur up to several times a month. After 2 to 3 years, they decrease in number and gradually disappear entirely. Most children have no further spells after the age of 7 or 8 years.

The cause of benign paroxysmal vertigo is unknown, although most authors suspect a vascular disturbance affecting the posterior cerebral circulation. Follow-up studies of patients with typical benign paroxysmal vertigo during childhood indicate that up to 50% subsequently develop classic migraine (Lanzi et al, 1986; Watson and Steele, 1974). When the clinical picture is clear, neither treatment nor extensive evaluation is necessary.

Paroxysmal torticollis

Snyder (1969) described 12 patients who developed recurrent attacks of head tilt between 2 and 8 months of age. The spells lasted from a few hours to 3 days and were occasionally accompanied by pallor, vomiting, and agitation at the beginning of the spell. Subsequent observers (Lipson and Robertson, 1978; Sanner and Bergstrom, 1979) have indicated that the disease can have a familial occurrence, and that symptoms can occur periodically and include lateral curvature of the trunk and occasionally extension of one leg.

Nystagmus was not reported in these patients, and caloric testing was normal in a very few patients. In most children the spells stop spontaneously by age 5, but Dunn and Snyder (1976) noted that 4 of 33 patients with benign paroxysmal vertigo of childhood previously exhibited paroxysmal torticollis of infancy. No treatment is necessary.

Vertebrobasilar insufficiency

Vertebrobasilar insufficiency is a common cause of vertigo, particularly in the elderly (Fisher, 1967; Williams and Wilson, 1962). Vertigo with vertebrobasilar insufficiency is abrupt in onset, usually lasts several minutes, and frequently is associated with nausea and vomiting. Associated symptoms resulting from ischemia in the remaining area supplied by the posterior circulation include visual illusions and hallucinations, drop attacks and weakness, visceral sensations, visual field defects, diplopia, and headaches. These symptoms occur in episodes either with the vertigo or alone. Vertigo may be an isolated initial symptom of vertebrobasilar insufficiency or may occur in isolation intermixed with more typical episodes (Grad and Baloh, 1989), but repeated episodes of vertigo without other symptoms should suggest another diagnosis (Fisher, 1967).

Vertebrobasilar insufficiency usually is caused by atherosclerosis of the subclavian, vertebral, and basilar arteries. Fig. 182-1 shows areas with a predilection for atherosclerotic plaques. Occasionally, episodes of vertebrobasilar insufficency are precipitated by postural hypotension, Stokes-Adams attacks, or mechanical compression from cervical spondylosis (Naritomi et al, 1979). Rarely, occlusion or stenosis of the subclavian or innominate artery just proximal to the origin of the vertebral artery results in the so-called subclavian steal syndrome. In this syndrome vertebrobasilar insufficiency results from siphoning of blood down the vertebral artery from the basilar systems to supply the upper extremities. Vertigo and other symptoms of vertebrobasilar insufficiency are precipitated by exercise of the upper extremities.

Angiography may be helpful in localizing the site of lesion with vertebrobasilar insufficiency, but often a poor correlation exists between angiographic and clinical findings. The physician should not undertake angiography unless the diagnosis requires it, or it is expected to lead to definitive treatment, such as removal or bypass of a focal atherosclerotic lesion.

In most cases treatment of vertebrobasilar insufficiency consists of controlling risk factors (diabetes mellitus, hypertension, hyperlipidemia) and using antiplatelet drugs (aspirin, 330 mg/day) (Caplan, 1986; Kistler et al, 1984). Anticoagulation is reserved for patients with frequent incapacitating episodes or in patients with symptoms and signs suggesting a stroke

in evolution, particularly basilar artery thrombosis. In these instances heparin is administered, with an intravenous bolus of 5000 units followed by a continuous infusion of 1000 units/hr. The dose is titrated to keep the partial thromboplastin time at approximately 2.5 times control. After 3 to 4 days, warfarin is administered with an oral dose of 15 mg. The daily dose is then adjusted (5 to 15 mg) until the prothrombine time is between 1.5 and 2 times the control value. Heparin is then discontinued. Although surgical reconstruction and revascularization procedures have been performed successfully in the vertebrobasilar system, their specific indications have not yet been defined.

Brainstem infarction

Lateral medullary syndrome

The zone of infarction producing the lateral medullary syndrome (Wallenberg's syndrome) consists of a wedge of the dorsolateral medulla just posterior to the olive (Fig. 182-2). The syndrome usually results from occlusion of the ipsilateral vertebral artery and rarely from occlusion of the posterior inferior cerebellar artery (Fisher et al, 1961). Characteristic symptoms include vertigo, ipsilateral facial pain, diplopia, dysphasia, and dysphonia.

On neurologic examination the following abnormalities may be found:

1. Ipsilateral Horner's syndrome resulting from involvement of the preganglionic sympathetic fibers originating in the hypothalamus.

2. Ipsilateral loss of pain and temperature sensation on the face caused by involvement of the nucleus and descending tract of the trigeminal nerve (CN V).

3. Ipsilateral paralysis of the palate, pharynx, and larynx resulting from involvement of the nucleus ambiguus and the exiting fibers of the glossopharyngeal and vagus nerves (CN IX and X).

4. Ipsilateral lateral rectus muscle and facial weakness caused by involvement of abducens and facial nerves (CN VI and VII).

5. Ispilateral dysmetria, dysrhythmia, and dysdiadochokinesia resulting from involvement of the cerebellum.

6. Spontaneous nystagmus caused by involvement of the vestibular nucleus.

7. Contralateral loss of pain and temperature sensation on the body resulting from damage to the crossed spinothalamic fibers.

Hearing loss does not occur because the lesion is caudal to the cochlear nerve entry zone and cochlear nuclei.

Some patients with Wallenberg's syndrome develop a prominent motor disturbance that causes their body and extremities to deviate toward the lesion site as if being pulled by a strong external force (Bjerner and Silfverskiold, 1968). This so-called lateral pulsion also affects the oculomotor system, producing excessively large saccades directed toward the side of the lesion, whereas saccades away from the side of the lesion are abnormally small. Most patients with Wallenberg's syndrome have major residual neurologic deficits years after the acute infarction.

Lateral pontomedullary syndrome

Ischemia in the distribution of the anteroinferior cerebellar artery results in infarction of the dorsolateral pontomedullary region and the inferolateral cerebellum (Adams, 1943). The middle cerebellar peduncle is typically the core of the affected territory (Amarenco and Hauw, 1990) (Fig. 182-3). Because the labyrinthine artery arises from the anteroinferior cerebellar artery in approximately 80% of individuals, infarction of the membranous labyrinth is a common accompaniment. Severe vertigo, nausea, and vomiting are the initial and most prominent symptoms. Other associated symptoms include unilateral hearing loss, tinnitus, facial paralysis, and cerebellar asynergy.

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Neurologic signs include the following:

1. Ipsilateral hearing loss and tinnitus caused by infarction of the labyrinth and cochlear nerve.

2. Ipsilateral facial weakness resulting from involvement of the facial nucleus and facial nerve.

3. Ipsilateral cerebellar dysfunction caused by involvement of the anteroinferior cerebellum.

4. Ipsilateral loss of pain and temperature sensation on the face resulting from involvement of the nucleus and descending tract of the trigeminal nerve.

5. Contralateral loss of pain and temperature sensation on the body caused by involvement of the crossed spinothalamic fibers.

6. Spontaneous vestibular nystagmus resulting from involvement of the labyrinth and vestibular nerve.

The clinical course consists of an acute onset followed by gradual improvement over a variable period. Vertigo may persist for several weeks to months because of damage to central compensation mechanisms.

Cerebellar infarction

Occlusion of the vertebral artery, the postero-inferior cerebellar artery, the anterior inferocerebellar artery, or the superior cerebellar artery may result in infarction confined to the cerebellum without accompanying brainstem involvement (Duncan et al, 1975; Sypert and Alvord, 1975). The initial symptoms are severe vertigo, vomiting, and ataxia; because typical brainstem signs do not occur, the physician may mistakenly diagnose an acute peripheral labyrinthine disorder. The key differential point is the presence of prominent cerebellar signs such as ataxia of gait and the extremities and gaze paretic nystagmus. The diagnostic procedure of choice is magnetic resonance imaging (MRI) (Bogousslavsky et al, 1986). After a latent interval of 24 to 96 hours, some patients develop progressive brainstem dysfunction caused by compression of a swollen cerebellum. A relentless progression to quadriplegia, coma, and death follows unless the compression is relieved surgically.

Cerebellar hemorrhage

Spontaneous intraparenchymal hemorrhage into the cerebellum causes multiple neurologic symptoms and signs that often progress to coma and death (Freeman et al, 1973; Ott et al, 1974). In most patients the cause of the hemorrhage is hypertensive vascular disease. The initial symptoms are often vertigo, nausea, vomiting, headache, and inability to stand or walk. This early syndrome is distinguished from more benign labyrinthine disorders by findings on neurologic examination of nuchal rigidity and prominent cerebellar signs. Obstructive hydrocephalus and brainstem compression typically follow within hours to days of the acute hemorrhage. Approximately 50% of patients lose consciousness within 24 hours of the initial symptoms, and 75% become comatose within 1 week of onset. The condition often is fatal unless surgical decompression is performed. The earlier the syndrome is recognized, the more likely that surgery will be successful. Once patients become comatose, almost none survive (Brennen and Bergland, 1977). Computed tomography (CT) scanning is indicated in all patients who display the clinical picture already described (Fig. 182-4). Midline cerebellar hemorrhage is particularly difficult to diagnose because it produces bilateral signs and generally runs a more fulminant course than lateral hemorrhage. If the CT is negative an MRI may still show evidence of cerebellar infarction (Bogousslavsky et al, 1986).

Neoplasms

Space-occupying lesions can induce vestibular symptoms by compressing or destroying neural tissue within the temporal bone, in the cerebellopontine angle, or intraaxially within the brainstem and cerebellum. Symptoms also can be produced or augmented by vascular compression. Chapter 193 discusses neoplasms and tumors affecting the temporal bone; Chapter 188 discusses acoustic neuromas and other cerebellopontine angle tumors.

Brainstem neoplasms

Gliomas of the brainstem usually grow slowly and infiltrate the brainstem nuclei and fiber tracts, producing multiple symptoms and signs. The typical history is relentless progressive involvement of one brainstem center after another, often ending with destruction of the vital cartiorespiratory centers of the medulla. These tumors are 5 to 10 times more common in children than in adults. Vestibular and cochlear symptoms and signs are common and occur in approximately 50% of patients, but a brainstem origin usually is obvious because of the multiple associated findings. Tumors originating in the pons or midbrain are likely initially to cause long tract signs, cranial nerve deficits, and ataxia. Although less common, tumors that originate in the medulla are likely to cause recurrent vertigo and vomiting. MRI is most helpful for identifying brainstem gliomas (Bradac et al, 1985) (Fig. 182-5). Radiation therapy is the treatment of choice for these lesions (Kim et al, 1980). Prolonged survival (more than 5 years) is not uncommon with more benign astrocytomas.

Tumors arising in the fourth ventricle and compressing the vestibular nuclei in its floor also commonly produce vestibular symptoms. Medulloblastomas, occurring primarily in children and adolescents, are rapidly growing, highly cellular tumors that arise in the posterior midline or vermis of the cerebellum and invade the fourth ventricle and adjacent cerebellar hemispheres (Fig. 182-6, A). Headaches and vomiting occur early from obstructive hydrocephalus and associated increased intracranial pressure (Fig. 182-6, B). An attack of headache, vertigo, vomiting, and visual loss may result from a change in head position, producing transient cerebrospinal fluid (CSF) obstruction (Bruns' syndrome. Positional vertigo and nystagmus may be the presenting symptom and sign (Grand, 1971). Other fourth ventricular tumors that produce similar clinical pictures include ependymomas, papillomas, teratomas, epidermoid cysts, and, in endemic areas, cysticercosis. The diagnosis of a fourth ventricular mass is made readily with MRI and CT scanning, but frequently the exact nature of the tumor cannot be determined before surgical exploration and biopsy. The tumor is removed completely whenever possible. Medulloblastomas are particularly sensitive to radiation therapy (Landberg et al, 1980).

Cerebellar neoplasm

Gliomas of the cerebellum may be relatively silent until they become large enough to obstruct CSF circulation or compress the brainstem (Bucy and Thieman, 1971). The most common symptoms are headache, vomiting, and gait imbalance. Approximately 90% of patients have papilledema from increased intracranial pressure. Positional vertigo is occasionally the initial symptom of a cerebellar glioma (Gregorius et al, 1976). Paroxysmal positional nystagmus, when present, is atypical because it can be induced in several different positions and is nonfatigable. Other tumors that produce identical symptoms and signs include teratomas, hemangiomas, and hemangioblastomas. Each of these tumors has a characteristic appearance on MRI and CT scanning, although occasionally the exact tumor type cannot be determined before surgical biopsy.

Disorders of the Craniovertebral Junction

Disorders of the craniovertebral junction are severe problems that are poorly understood and relatively uncommon. The patients are frequently referred to surgeons because of brainstem and lower cranial nerve signs and symptoms such as tinnitus, vertigo, hearing loss, pharyngeal dysfunction, hoarseness, or airway obstruction; occasionally they are seen for problems unrelated to the craniovertebral junction. Physical examination is sometimes difficult, and appropriate radiographic evaluation to establish the diagnosis can be difficult and complex. An understanding of these problems helps to facilitate appropriate evaluation and management and to prevent inappropriate evaluation and management such as middle ear explorations, endolymphatic sac operations, vocal cord injections, cricopharyngeal myotomies, or arytenoidectomies.

The basic physiologic problem common to these disorders is physical compression of the central nervous system at the upper spinal cord and medulla (cervicomedullary compression) (Menezes et al, 1980; Michie and Clark, 1968). The rostrocaudal area of compression is variable, and the impingement can be ventral, dorsal, or (rarely) both. A second, less common mechanism is vascular insufficiency of the anterior spinal or vertebral arteries from angulation, stretching, or extrinsic obstruction (List, 1941; Schneider and Schemm, 1961). The way these two types of events occur are variable and often multiple.

Classification

Basilar impression

Basilar impression is an upward indentation or invagination of the normally convex skull base, which appears as though the ridged cervical spine were being pushed cephalically into a softer, more plastic base that yields under the weight of the head (Bertrand, 1982). The normal anatomy of the foramen magnum, fist cervical vertebra (atlas), second cervical vertebra (axis), and its odontoid process (dens) can become altered by disease, trauma, or congenital deformities. When a softening of the skull base results, the skull literally descends onto the spinal column and odontoid. As the odontoid projects intracranially, it compresses the ventral aspect of the medulla.

Associated erosive changes of the occipital condyles and atlas accentuate the protrusion of the odontoid, narrow the diameter and circumference of the foramen magnum and spinal canal, and provide mechanisms for dorsal cervicomedullary compression. The odontoid not only projects above the foramen magnum but also occupies a significant portion of its circumference.

Disorders known to cause basilar impression include Paget's disease, rheumatoid arthritis, osteomalacia, osteogenesis imperfecta, cretinism, and rickets (Menezes et al, 1980). The diagnosis is confirmed when lateral radiographs of the skull demonstrate that the tip of the odontoid either extends above Chamberlain's line (a line drawn from the posterior edge of the hard palate to the posterior lip of the foramen magnum) (Chamberlain, 1939) or projects posterior to Wackenheim's clivus-canal line (Fig. 182-7). The term *platybasia* has been used synonymously with *basilar impression* by some authors. Technically it is not a measure of basilar impression, and although the two often coexist, platybasia by itself causes

no symptoms.

Assimilation of the atlas

Assimilation of the atlas (also called occipitalization of the atlas) is a bony union between the first cervical vertebra and the skull. The amount of union varies, but motion between the two structures does not occur. As a result the odontoid often impinges on the effective anteroposterior diameter of the foramen. Frequently there is associated fusion of the axis to the third cervical vertebra.

The Klippel-Feil syndrome is the most common cause of cervical vertebral fusion. The extent of fusion is quite variable, as are the manifestations related to abnormalities distant from the neck. The most common type is characterized by cervical vertebral fusion and associated atlantooccipital fusion. Although vestibular and auditory symptoms of Klippel-Feil syndrome can result from an associated Mondini deformity (McLay and Maran, 1969), cervicomedullary junction compression must also be considered and its associated symptomatology sought (described later). Assimilation of the atlas facilitates the development of atlantoaxial dislocation.

Atlantoaxial dislocation

During flexion and extension of the neck, congenital fusion of the occiput to the atlas increases the strain on the structures that normally restrict the motion of the atlas on the axis, especially if there is fusion of other cervical vertebrae as well. The transverse ligaments that normally secure the odontoid against the anterior aspect of the arch of the atlas may weaken because of this repeated strain, and the resultant laxity allows the odontoid to move posteriorly into the lumen of the foramen magnum with neck flexion. Flexion or extension of the neck may then produce symptoms, depending on whether the predominant neural compression is anterior from the odontoid or posterior from the posterior arch of C1. When the atlas or congenital cervical fusion has been assimilated, the transverse odontoid ligament is sometimes hypoplastic, which makes laxity and atlantoaxial dislocation even more likely.

Atlantoaxial instability is also associated with a number of congenital and acquired disease processes. It occurs in 18% to 30% of individuals with Down syndrome (Martel and Tishler, 1966; Nordt and Stauffer, 1981) and is frequently seen with spondyloepiphyseal dysplasia, Hurler's syndrome, and Morquio syndrome and in achondroplastic dwarfs. Of patients with rheumatoid arthritis, 25% have atlantoaxial instability secondary to destruction of the normal stabilizing mechanisms by inflammatory rheumatoid tissue in the synovial membrane (Bland, 1974). Similarly, ligamentous laxity can result from inflammatory conditions affecting retropharyngeal soft tissues or cervical bony structures, such as tuberculous (or other bacterial) osteitis, retropharyngeal abscess, or lymphadenitis (Greenberg, 1968; Sullivan, 1949).

Chiari malformation

Chiari malformations are a developmental defect of the cervicomedullary junction not infrequently associated with vestibular symptoms. With this soft tissue malformation, the caudal brainstem and cerebellum are elongated and protrude down into the cervical canal. It appears in two forms: Chiari type I and type II malformations. Type II is more common and becomes manifest in the first few months of life. It is associated with hydrocephalus and other CNS malformations, usually meningomyelocele.

The Chiari type I malformation is more difficult to diagnose and more important to the surgeon. The onset of symptoms and signs is delayed until young adulthood. The afflicted patients often have subtle neurologic symptoms and signs and are usually unassociated with other developmental defects. The malformation can usually be visualized with CT scanning of thin cuts (1.5 mm) through the foramen magnum region. Intrathecal metrizamide enhances the contrast. MRI provides the best visualization (Fig. 182-8). Suboccipital decompression of the foramen magnum region can stop the progression and occasionally lead to improvement of neurologic symptoms and signs (Spooner and Baloh, 1981).

Symptoms and signs

Patients with congenital abnormalities of the craniovertebral junction often exhibit associated morphologic abnormalities of the neck, such as a low hairline, short neck, abnormal head position, limitation of motion, and painful torticollis. Accentuation of symptoms by coughing, straining, or change in neck position is common.

A recent review of 50 patients with Chiari malformation (41 type I) who did not have an associated myelomeningocele revealed that ataxia and nystagmus were both reported in 24% of patients, whereas vertigo was a complain in only 8% and tinnitus in only 2%. The most common symptoms were weakness of the arms or legs (60%), pain (54%), and sensory loss (34%) (Dyste and Menezes, 1989).

Clinical manifestations of cervicomedullary compression from other causes are usually relentless and severe, progressing over several weeks to many months. Because the compression can occur over a wide rostrocaudal distribution and be either ventral or dorsal, manifestations are understandably variable. The addition of intermittent vascular insufficiency from temporary kinking adds transient symptoms, as do changes during flexion or extension and during sudden increases in cerebrospinal fluid pressure.

The most common symptom is occipital pain with radiation toward the vertex, and it is almost universally present. Other symptoms can be classified as motor, sensory, cranial nerve, and intrinsic brainstem.

Motor symptoms can be limited to mild transient weakness, sometimes characteristic of the central cord syndrome, with greater weakness in the upper than the lower extremities. The weakness may progress to permanent disability and eventually to complete spastic quadriparesis. When there are associated musculoskeletal problems - for example, from rheumatoid arthritis - motor assessment can be difficult, and long tract rigns such as Babinski's sign are helpful.

Most patients report sensory changes, which may be limited to transient limb paresthesias or fixed deficits in pin-prick detection (especially over the posterior scalp) or pain and light-touch sensation.

Cranial nerve signs are especially important to the surgeon. In these patients the abnormality often results from brainstem nuclear compression, but the manifestations are as if the nerves themselves were involved. Ptosis, diplopia, and facial diplegia may be seen; but these are relatively uncommon. Episodic vertigo, tinnitus, and hearing loss occur in some combination in between 25% and 75% of the patients (Elies and Plester, 1980; Menezes et al, 1980, 1985).

Elies and Plester (1980) studied 180 consecutive patients with tinnitus, hearing loss, or vertigo; 32 (17.8%) exhibited craniovertebral junction abnormalities, 26 patients had a unilateral fluctuant but slowly progressive sensorineural hearing loss, 17 had tinnitus that was predominantly unilateral, and 22 experienced vertigo that was symptomatically different from that caused by Ménière's disease. In these patients the vertigo was related to head movement, body flexion, physical effort, or changes in body position. It was oscillatory or vertical and only rarely rotational (as experienced by Ménière's disease patients), although nausea accompanied the vertigo.

Audiometric tests showed normal hearing in only 6 of the 32 patients; the remainder had variable degrees of sensorineural impairment. Five of these 26 had unilateral anacusis, and one had bilateral anacusis.

Lower cranial nerve involvement (CN IX through CN XII) can result in difficulty eating because of pharyngeal plexus and tongue dysfunction, as well as hoarseness or airway embarrassment. Intrinsic brainstem involvement is indicated by the presence of internuclear ophthalmoplegia, downbeat vertical nystagmus, sleep apnea, respiratory arrest, syncopal episodes, and sensorinum changes.

Preoperative assessment

Careful radiologic assessment has allowed both accurate diagnosis and appropriate choice of surgical treatment. The critical assessment features include (1) whether the abnormality is reducible and (2) the direction of encroachment on the cervicomedullary junction. Pluridirectional tomography is performed in the frontal and lateral projections, the lateral with the patient's head in both the neutral and the extended positions (with the attending neurosurgeon supervising the procedure).

Metrizimide myelotomography is performed at the end of the period of traction, which in severe cases may be 2 weeks. This is usually followed by CT scanning, and in some instances by MRI.

Management

A series of operations has been developed to correct the craniovertebral abnormalities, eliminate the cervicomedullary compression, and prevent its recurrence (Fig. 182-9). They are designed to reduce the odontoid from its cranial position, to remove any bony, ligamentous, or inflammatory soft tissue compression of the cervicomedullary junction; and to fix the skull to the cervical vertebral column in the reduced position when necessary. In expert hands, results are impressive.

In a series of 45 patients surgically treated for cervico-medullary compression secondary to rheumatoid arthritis and cranial settling (Menezes et al, 1985), there were no operative deaths and no infections. All patients improved to a functional class two grades above the preoperative level, and some improvement in cranial nerve function occurred in all patients who had preoperative deficits. All tracheotomies were successfully removed.

When patients who are at risk for craniovertebral junction abnormalities are scheduled for elective or emergent surgery because of diseases in other organ systems, the surgeon should be alert to the possibility of operative CNS complications, even though the patient may have no CNS symptoms before the operation. In high-risk patients, such as those with Down syndrome and rheumatoid arthritis, lateral cervical spine radiographs are appropriate preoperative examinations.

In patients seen for cranial nerve deficits or those with cochlear or vestibular symptoms, a careful history and physical examination should prompt clinical suspicion to pursue the necessary diagnostic studies.

Special emphasis should be given to patients with rheumatoid arthritis. Because up to 25% have significant craniovertebral abnormalities, one cannot simply assume that the complaint of tinnitus reflects salicylate intoxication, that an immobile vocal cord reflects cricoarytenoid ankylosis, or aspiration from pharyngeal dysfunction is best managed by a cricopharyngeal myotomy. The otolaryngologist may be the key person to recognize the gravity of the situation and arrange for appropriate evaluation and treatment.

Multiple Sclerosis

Multiple sclerosis is a demyelinating CNS disorder of unknown cause with onset usually in the third and fourth decades of life (McAlpine et al, 1972). The key to the diagnosis is the finding of disseminated signs of CNS dysfunction manifested in an alternating remitting and exacerbating course. Although many symptoms of multiple sclerosis occur, certain ones deserve emphasis because of their consistent appearance. Blurring or loss of vision caused by demyelination of the optic nerve (retrobulbar neuritis) is the initial symptom of multiple sclerosis in approximately 20% of patients. Diplopia, weakness, numbness, and ataxia also occur early in the disease process. Vertigo is the initial symptom in about 5% of patients and is reported sometime during the disease in as many as 50%. Hearing loss occurs in about 10% of patients (Noffinger et al, 1972). No apparent relationship exists between the severity or duration of multiple sclerosis and the hearing loss; auditory impairment may be part of the initial episode or may occur more than 10 years after the onset (Daugherty et al, 1983). The hearing loss can be acute (hours to a few days), subacute (over months), or

insidious in onset. Partial or complete remission after the onset of hearing loss is common.

The findings on examination are as diverse as the symptoms. In most long-standing cases there are signs of involvement of the pyramidal tracts (hyperreflexia, extensor plantar responses), cerebellum (intention tremor, ataxia, slurred speech), and visual pathways (decreased visual acuity and pallor of the optic disk). The finding of dissociated nystagmus on lateral gaze or acquired pendular fixation nystagmus is particularly helpful in diagnosing multiple sclerosis and relatively unusual with other disease processes (Aschoff et al, 1974). All varieties of positional nystagmus, including fatigable paroxysmal positional nystagmus, can be seen with multiple sclerosis, and caloric examination is abnormal in approximately 25% of patients (Dam et al, 1975). Pure-tone hearing level, when abnormal, have no characteristic pattern with multiple sclerosis. Special audiometric studies (speech discrimination, tone decay, acoustic reflex) indicate a neural site for the lesion. Brainstem auditory-evoked responses can detect subclinical lesions of multiple sclerosis (Stockard et al, 1977) and usually are abnormal when hearing loss is present (Daugherty et al, 1983). There are not specific laboratory tests for multiple sclerosis, but abnormalities in CSF examination can be identified in approximately 90% of patients sometime during the course of the disease. These findings include an elevated gamma-globulin level, increased gamma-globulin synthesis, oligoclonal banding of gamma-globulin, and elevated myelin basic protein (Waxman, 1983). Unfortunately none of these findings are specific for multiple sclerosis. MRI will identify white matter lesions in from 70% to 95% of patients with multiple sclerosis (Fig. 182-10), although similar lesions are sometimes seen in patients without the clinical criteria for the diagnosis of multiple sclerosis (Debaene et al, 1986; Ormerod et al, 1987).

There is no definitive treatment for multiple sclerosis. Collaborative studies (Rose et al, 1970) suggest that steroids and adrenocorticotropic hormone (ACTH) hasten the remission of symptoms and signs after an acute exacerbation in some patients, but no evidence demonstrates that these drugs alter the natural history of multiple sclerosis. The potential benefit of other immunosuppressant drugs, such as cyclophosphamide and azathioprine, currently are being investigated, particularly regarding the risk-benefit ratio.

Familial Ataxia Syndromes

Auditory and vestibular symptoms occur occasionally in the hereditary ataxia syndromes, although in many instances the site of pathologic involvement has not been investigated adequately. Of the well-defined syndromes, Friedreich's ataxia, olivopontocerebellar degeneration, Refsum disease, Roussy-Lévy's disease, and episodic vertigo and ataxia are associated with loss of vestibular and auditory function. In addition, several isolated families with atypical ataxis syndromes associated with hearing loss and absent vestibular function have been reported. The Richards-Rundle syndrome consists of progressive spinocerebellar ataxia, hypogonadism, deafness, and mental retardation (Sylvester, 1972). Other reports describe a familial condition with vestibular impairment, a progressive spastic ataxia, and retinal degeneration (Bergstedt et al, 1962), and, a sex-linked disorder with onset in infancy with deafness and progressive ataxia (Schmidley et al, 1987). A rare familial disorder including cerebellar ataxia, deafness, and hyperuricemia with renal impairment also has been described (Rosenberg et al, 1970).

Recurrent or progressive cerebellar ataxia is the primary neurologic deficit in several well-defined hereditary disorders. These disorders include ataxia-telangiectasia, Bassen-Kornzweig syndrome, or hypo-beta-lipoproteinemia; hexosaminidase deficiency spinocerebellar degeneration variant; the late-onset form of Menkes' disease; the delayed-onset variant of maple syrup urine disease; and the syndrome of familial nephropathy, retinitis pigmentosa, and ataxia. Progressive cerebellar ataxia with cortical hearing loss on occasion can be the presenting manifestation of sex-linked adrenoleukodystrophy, preceding the appearance of dementia. The increase in serum very-long-chain fatty acids provides diagnostic confirmation of this disorder.

Clinically, cerebellar or spinal cord findings overshadow the loss of vestibular function, and ataxia rather than vertigo is usually the principal complaint. In some the symptoms are constant; in others there is episodic accentuation of symptoms. Only after performing caloric or rotatory testing can the physician recognize the impaired vestibular function. Vertigo rarely is experienced because the progressive loss of vestibular function is bilaterally symmetric. Many types of pathologic nystagmus are encountered, including gaze-paretic nystagmus, spontaneous vestibular nystagmus, positional nystagmus, and rebound nystagmus (Baloh and Honrubia, 1990). The diagnosis of the familial ataxic syndromes is primarily clinical, based on a characteristic profile for each syndrome (Gillman et al, 1981).

Friedreich's ataxia

Friedreich's ataxia is an autosomal recessive disorder, which is considered to be the prototype of the hereditary spinocerebellar ataxias. It has a mean age of onset between 8 and 13 years of age, although the insidious onset of gait incoordination often precludes establishment of the precise time of onset. Instability of gait is by far the most common initial manifestation of Friedreich's ataxia, but on rare occasion the disorder presents with progressive hearing loss or with manifestations of the associated cardiomyopathy (Harding, 1981; Salih et al, 1990). As the illness progresses, ataxia becomes more severe, involving both axial and appendicular structures. Scoliosis is a customary component of the illness, as is the development of pes cavus deformities of the feet. Dysarthria and, less often, nystagmus become evident as the process continues. In addition, neurologic examination reveals muscle wasting, diffuse reflex loss despite extensor plantar responses, and posterior column signs in the form of loss of vibration and position sensations, especially in the feet. Optic atrophy with visual loss can occur but is infrequent until the later stages of the illness. Dementia, likewise, is a feature of the later stages, but is not invariably present. Friedreich's ataxia is a slowly progressive disease with most affected patients losing their ability to ambulate independently 10 to 20 years after onset. Duration of the illness is variably with many surviving into middle age.

Cardiomyopathy is an integral component of Friedreich's ataxia and can become the most life-threatening aspect of the disease. Asymmetric septal hypertrophy leading to left ventricular outlet obstruction and cardiac arrhythmias complicate the clinical course in some patients. Fibrotic and degenerative myocardial changes are associated with coronary artery stenosis and recurrent myocardial ischemia in others. Congestive heart failure resulting from hypertrophic cardiomyopathy on rare occasion precedes the customary neurologic manifestation. The other metabolic abnormality customarily associated with Friedreich's ataxia is carbohydrate intolerance or overt diabetes mellitus. The type of diabetes varies among

patients, with many being insulin dependent. Among older children and adolescents who develop a progressive ataxic disorder not explained by metabolic or image studies, evaluation of cardiac status and carbohydrate tolerance are important diagnostic measures.

A variety of metabolic aberrations have been postulated or suggested in Friedreich's ataxia, including deficiency of pyruvate dehydrogenase and abnormalities in lipoamine dehydrogenase. No definite conclusions have as yet been made. Neuropathologic findings include degeneration of the posterior columns, spinocerebellar pathways, and pyramidal tracts. Degeneration of peripheral sensory pathways and large neurons in the dorsal root ganglia also occur, as do cerebellar changes, although they are less striking than those affecting the spinal cord.

Roussy-Lévy's syndrome

Roussy-Lévy's syndrome was once classified with Friedreich's ataxia but is now recognized to be a variant of hereditary motor-sensory neuropathy type I or Charcot-Marie-Tooth disease. Most cases are autosomal dominant. The disorder is characterized by lower extremity weakness and variable sensory loss in addition to tremor and ataxia in some cases. It is primarily a peripheral nerve disease with neural pathology including a hypertrophic neuropathy with characteristic onion-bulb formation. Pes cavus is common, as is loss of deep tendon reflexes and progressive muscle atrophy. Ataxia in this disorder can be on the basis of cerebellar dysfunction or secondary to proprioceptive abnormalities associated with the peripheral nerve pathology. Vertigo or other types of vestibular dysfunction are not usually seen with this condition.

Olivopontocerebellar atrophy

Olivopontocerebellar atrophy is a group of familial progressive degenerative disorders that can be inherited in autosomal recessive or dominant fashion or can occur sporadically. Large families with progressive hereditary ataxis disorders reported near the end of nineteenth century by Marie, Sanger Brown, and Menzel are now considered to be somewhat heterogeneous but included among the various types of olivopontocerebellar atrophy. The Schut-Haymaker type, described more recently, is now classified among the autosomal dominant types of olivopontocerebellar atrophy. Although the primary pathology is within the cerebellum, pons, and inferior olivary nuclei, neuronal degenerative changes usually occur in various parts of the brain and spinal cord. The disorder can begin in childhood or anytime during the adult years. The onset is usually with unsteadiness of gait and dysarthria. With progression, some affected patients develop extrapyramidal signs, including bradykinesia, tremor, and a slow hesitant gait, reminiscent of Parkinson's disease. Bulbar dysfunction is common and dementia sometimes appears during the later stages. Retinal-macular degeneration is a common feature of the disorder and is associated with progressive visual decline and extinguished electroretinograms. This manifestation is diagnostically important, as the combination of progressive retinal degeneration in association with an acquired and progressive cerebellar ataxic syndrome is suggestive of olivopontocerebellar atrophy. Image studies, including CT (Fig. 182-11) are diagnostically useful. The latter are especially helpful in revealing cerebellar and brainstem atrophy in the sagittal dimension. Some cases of olivopontocerebellar atrophy have been associated with partial deficiency of glutamate dehydrogenase by study of leukocytes or cultured fibroblasts (Plaitakis et al, 1980).

Refsum's syndrome

Refsum's syndrome is a familial disorder that occurs mainly in persons of Scandinavian origin, and is believed to be inherited in autosomal recessive fashion. The onset of symptoms usually occurs in oldren children or young adults. When first described, it was labeled *heredopathia atactica polyneuritiformis.* Clinical disabilities that characterize the disorder include progressive visual decline secondary to retinitis pigmentosa, weakness due to a chronic polyneuropathy, cerebellar ataxia that sometimes has an intermittent course, sensorineural hearing loss, and ichthyosis. CSF generally contains an increased protein content without a cellular response. Cardiomyopathy with abnormal electrocardiographic findings is present in some patients. Refsum's disease is caused by an enzymatic deficiency that leads to serum and tissue storage of a branched-chain fatty acid (3,5,7,11-tetramethyl hexadecanoic acid) called phytanic acid. Serum analysis of the content of phytanic acid provides a diagnostic test for the disorder (Rosenberg and Pettegrew, 1983). Because phytanic acid is predominantly exogenous (dietery) in origin, the illness can be treated with dietary restriction (Refsum, 1981). Meats and fish oils provide the highest dietary content of this fatty acid. Proper diet management reduces the serum phytanic acid content to normal or near normal. In addition to these conditions, an infantile form of phytanic acid oxidase deficiency has been identified (Weleber et al, 1984). It is characterized by retinal dystrophy, deafness, mental retardation, dysmorphic facial features, and phytanic acid storage. The serum phytanic acid content is usually considerably lower in the infantile form than in the adult form of the disease. Peroxisomal dysfunction in infantile phytanic acid storage disease accounts for the elevation of the very-long-chain fatty acids found in this condition.

Episodic vertigo and ataxia

Episodic vertigo and ataxia is an important but small group of poorly understood familial ataxias manifested by recurrent vertigo and ataxia often associated with other signs and symptoms such as diplopia, vertical nystagmus, and dysarthria (Donat and Auger, 1979; Farmer and Mustian, 1963; Griggs et al, 1978; Whigte, 1969; Zasorin et al, 1983). The episodes begin between early childhood and early adulthood, last minutes to 2 or 3 days, and are often periodic. The inheritance pattern is autosomal dominant. Their major significance to otolaryngologists is that they represent another treatable cause of vertigo. Symptoms are eliminated by administration of acetazolamide (Diamox) (Donat and Auger, 1979; Griggs et al, 1978; Zasorin et al, 1983). It is not known whether the therapeutic effect results from the mild metabolic acidosis produced by the drug or from direct action.

Late onset corticocerebellar atrophy

Late onset corticocerebellar atrophy is characterized by the gradual onset of ataxia beginning in late life (Baloh et al, 1986). This disorder can be sporadic or inherited as an autosomal dominant trait. Both varieties are characterized by a highly localized atrophy of the cerebellum, particularly the archicerebellum and the paleocerebellum. There is extensive loss of Purkinje cells in the vermis (especially the anterior vermis) and the flocculonodular lobe, with relatively little neuronal loss in other areas of the brain. Cerebellar eye signs are common, including several types of pathologic nystagmus (especially spontaneous downbeat or upbeat). MRI typically identifies atrophy remarkably localized to the midline cerebellum (Fig. 182-12).

Focal Seizure Disorders

Vertigo can be part of an aura of a focal seizure. Presumably a focal discharge activates the cortical projections of the vestibular system. Smith (1960) studied 120 patients with focal seizures who experienced vestibular symptoms as part of their aura. He attempted to define the cortical focus of origin on the basis of associated symptoms. The most common vestibular symptom was a sense of spinning, occurring in 55% of patients, followed by a sense of linear movement in 30%. Common associated symptoms and their frequency included visceral and autonomic symptoms in 62% of patients, visual symptoms in 45%, auditory symptoms in 28%, and somatosensory symptoms in 22%. Of the visceral and autonomic complaints, an abnormal epigastric sensation was most common, followed by nausea, mastication, and salivation. Visual illusions and hallucinations occurred frequently, suggesting a close functional relationship between cortical visual and vestibular projections. Auditory symptoms included tinnitus, auditory hallucinations, and auditory illusions. Mapping the suspected cortical foci on the basis of these associated symptoms suggested that lesions of the frontal, parietal, and temporal cortex can result in vestibular symptoms as part of the aura phenomenon. It should be emphasized, however, that episodes of vertigo as an isolated manifestation of a focal seizure disorder occur rarely, if at all.