Paparella: Volume IV: Plastic and Reconstructive Surgery and Interrelated Disciplines

Section 2: Disciplines Closely Associated With Otolaryngology

Chapter 22: Clinical Neurology for the Otolaryngologist

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The nervous system spreads diffusely throughout the body, but there is a very large focus about the head and neck. Thus, it behooves the head and neck surgeon to have a special overview of this complex system. This chapter attempts to do just that. It is organized into two broad categories: (1) pathologic processes within the nervous system and (2) symptoms frequently encountered in neurologic disease. It is not intended to be all-encompassing but is meant to lead the reader in the proper direction to diagnose and treat neurologic disease.

When faced with a difficult diagnostic dilemma, experienced clinicians structure their thinking along pathologic lines. The clinicopathologic profile often allows a diagnosis to be made in even the most difficult of circumstances. Neurologically, disease can be divided into acquired traumatic, acquired nontraumatic, and congenital disease. Congenital and traumatic diseases usually are not hard to separate and diagnose, but to differentiate acquired nontraumatic neurologic diseases, certain generalizations are necessary.

There are five general categories of nontraumatic neurologic disease: (1) vascular, (2) neoplastic, (3) toxic-metabolic, (4) infectious, and (5) degenerative. Each of these has its own individual manner of presentation. Clinically the abruptness of onset, the course of the clinical picture, and the distribution of abnormalities within the nervous system often reveal to which category the process belongs.

Vascular disease is known for its abrupt onset. After the acute onset the process stabilizes, and usually the clinical course is one of improvement. Vascular disease is focal in distribution. The clinician usually has little difficulty ascertaining where in the brain the problem lies. Thus, stroke is fairly straightforward diagnostically.

Neoplastic disease is much slower in its presentation, which is insidious and progresses over time. The focal distribution is not often confused with stroke because of the progressive, slower clinical picture.

Toxic-metabolic disease is quite variable depending on the toxin. Generally the process is subacute in onset, leaning toward the slower side. The process continues unless the toxin is removed. The usual presentation in the nervous system is diffuse. There are a number of exceptions to this rule, but the generalization holds: toxic-metabolic disease localizes poorly in the nervous system.

Infectious disease also distributes in a nonfocal manner, and also presents subacutely and progresses unless treated. Although fever, increased white blood cell counts, and stiff neck may aid in distinguishing infectious disease from toxic-metabolic disease, these are not consistent. Consequently these processes are often confused. Spinal fluid analysis settles the confusion; thus, if infection is suspected, a spinal tap is mandatory. If the fluid is without cells, a toxic process must be considered; if cells are present, infection can be diagnosed.

Degenerative disease presents slowly and is diffuse in its distribution within the nervous system. Alzheimer's disease is the classic example of this type of process. Demyelinating diseases (eg, multiple sclerosis) may present acutely, have multifocal distribution, and fluctuate greatly; they thus are an exception to the general rules.

Cerebrovascular Disease

Cerebrovascular disease is the third most common cause of death in the USA. As such it occupies a prominent position in the activities of physicians of all specialties.

Anatomically the brain is divided into two general circulations interconnected through the well-known circle of Willis. The anterior circulation is formed by the bifurcation of the internal carotid artery into the middle cerebral and the anterior cerebral arteries. At or around this point a third vessel, the posterior communicating artery, brings the anterior circulation together with the posterior circulation. The anterior circulations from each side are connected via the anterior cerebral arteries through an anterior communicating artery. The posterior circulation is composed of two vertebral arteries that course through the neck and unite at the pontomedullary junction to form the basilar artery. These arteries send a number of short penetrating branches to the brain stem, short circumferential branches around the brain stem (to provide circulation to the middle of the stem), and longer circumferential branches to the top of the brain stem. These longer branches include the posteroinferior cerebellar artery, the superior cerebellar artery, and (at the tip of the basilar artery) the posterior cerebral artery. The posterior cerebral artery is then connected with the anterior circulation via the posterior communicating artery, thus completing the circle of Willis.

This interwoven circulation provides for a significant amount of collateral circulation to the brain. There is also collateral from the external carotid and its branches and also from dural blood vessels. The venous system is composed of a complex series of veins and venous sinuses (dural) that provide low-pressure, slow flow from the brain back to the general circulation.

Cerebral circulation is regulated by a number of factors. Of importance is the autoregulatory system inherent in the circulation of the brain. It is this process that allows for a constant blood flow to the brain despite changes in general perfusion pressure. Thus, even though changes may occur in the circulation, the brain attempts stability. General blood pressure obviously plays a role in perfusion pressure, as do blood viscosity, biochemical factors, and oxygen saturation. With disease of the cerebral circulation the autoregulatory process is often lost, forcing the brain to rely far more on hydrostatic blood pressure.

Cerebrovascular disease may be divided into hemorrhagic and ischemic varieties. The latter may be subdivided into thrombotic and embolic. Decreased blood flow to a part of the brain (ischaemia) causing symptoms that last 24 hours or less with full resolution is defined as a transient ischemic attack (TIA). If resolution is full but the symptoms have lasted for longer than 24 hours, the process is described as a reversible ischemic neurologic deficit (RIND). If only part of the symptoms resolve, the process is called a partially reversible neurologic deficit (PRIND). An ischemic process continuing in front of the observer's eyes is a stroke in evolution. When all is settled and there remains deficit, a completed stroke (cerebrovascular accident, CVA) has occurred.

Initial workup and treatment of ischemic cerebrovascular disease is dictated by the region of the brain involved. Because the carotids are accessible surgically, an attempt is made to determine whether they are involved in the pathologic process. Thus, if a transient ischemic attack has occurred, it has to be decided whether the process is from the anterior or posterior circulation.

Symptoms of anterior circulation ischemia include amaurosis fugax (transient blindness in one eye), aphasia, hemiparesis with or without field cut, apraxia, agnosia, and hemisensory loss. Symptoms of posterior circulation ischemia include vertigo, ataxia, dysarthria, dysphagia, diplopia, crossed sensory loss (ipsilateral face, contralateral body), and cranial nerve palsies.

If there are no contraindications, heparin is usually used during the acute aftermath of a transient ischemic attack. Before anticoagulation is considered, an unenhanced computed tomographic (CT) scan of the head should be performed to search for the possibility of hemorrhage. Examination for bruits over the carotids and subclavian and vertebral areas is important. Blood pressure is allowed to be higher than normal to allow increased perfusion pressure of the ischemic area. If the attack involved the anterior circulation, the carotid vessels should be studied looking for an embolic source. Study may be with angiography or, with somewhat less accuracy, by carotid ultrasonography. If an appropriate lesion is noted, surgery may be contemplated to remove the source of the transient ischemic attack. If no source is discovered or if thrombotic (atherosclerotic) disease that is inappropriate for surgery is found, long-term management with aspirin is recommended.

If the attack involved the posterior circulation, visualization of the vessels is less important and more difficult. Surgery (except in experimental situations) is not contemplated. Longer-term anticoagulation with warfarin is controversial but often utilized. Aspirin again is recommended. A large number of brain stem syndromes have been recognized. Their eponyms are of historical interest and are noted in Table 1.

Table 1. Vascular Syndromes

Insufficiency of internal carotid artery
: Transient unilateral muscular weakness and sensory disturbances on same side; intermittent or permanent disturbance of vision in ipsilateral eye.

Posteroinferior cerebellar artery thrombosis (Wallenberg's syndrome)
: Vertigo, ipsilateral facial hypesthesia, dysphagia, ipsilateral Horner's syndrome, ipsilateral incoordination, contralateral hypesthesia body.

Basilar artery insufficiency
: Recurrent vertigo, transient visual dimness, diplopia, dysphagia, dysarthria; transient hemiparesis that shifts from side to side, falling or dropping.

Anteroinferior cerebellar artery thrombosis
: Ipsilateral ataxia, loss of facial sensation, Horner's syndrome, facial palsy, contralateral incomplete loss of pain and temperature.

Superior cerebellar artery thrombosis
: Ipsilateral ataxia, Horner's syndrome, contralateral hemihypesthesia.

Thrombosis of vertebral artery (Avellis' syndrome)
: Ipsilateral paresis of palate, pharynx; contralateral loss of pain and temperature.

Céstan-Chenais syndrome
: Paralysis of palate, ipsilateral ataxia, Horner's syndrome, contralateral hemiplegia.

Babinski-Nageotte syndrome
: Ipsilateral paresis of palate, loss of taste, loss of pain and temperature, ataxia, Horner's syndrome, contralateral hemiplegia.

Basilar artery thrombosis
: Bilateral cranial nerve and long tract findings.

Thrombosis of medial pontine branches
: Ipsilateral facial paralysis, lateral rectus paresis, contralateral hemiplegia, decreased sensation.

Thrombosis of lateral pontine branches
: Ipsilateral ataxia; ipsilateral paresis of nerves V, VII, VIII; contralateral loss of pain and temperature.

Jackson's syndrome
: Ipsilateral palate weakness, weakness of sternocleidomastoid and trapezius.

Tapia's syndrome
: Ipsilateral palate weakness, atrophy of tongue.

Vernet's syndrome
: Ipsilateral paresis of nerves IX, X, XI.

Villaret's syndrome
: Ipsilateral paresis of nerves IX, X, XI, XII; Horner's syndrome.

An echocardiogram may be necessary to rule out the possibility of emboli from the heart.

Hemorrhagic cerebrovascular disease may occur from the rupture of a small blood vessel with or without antecedent hypertension, rupture of a berry aneurysm, other developmental

defects, rupture by infection, blood dyscrasias, and unknown causes.

Management of cerebral hemorrhage is determined by the cause. Primary subarachnoid hemorrhage either from rupture of an arterio-venous malformation or, more commonly, from a berry aneurysm is evaluated with an eye toward a surgical resolution. Thus, after the patient is stabilized (blood pressure, oxygen, and so forth), angiography is performed to determine the location of the aneurysm or other vascular anomaly. CT scanning may be of aid in determining the site of the lesion. Spasm of the involved vessels may postpone or otherwise interfere with a good surgical result. Nonetheless, the process is toward surgery.

On the other hand, if the hemorrhage is secondary to hypertension without aneurysm, surgery should be postponed, unless the intracranial pressure is high enough to be contributing to a herniation of brain through the foramen magnum. Small (and sometimes large) hemorrhages often resolve with surprisingly little deficit.

Occasionally, inflammatory diseases cause cerebrovascular disease. Systemic lupus erythematosus may lead to thrombosis and infarction. Polyarteritis nodosa is less common than lupus but may also cause cerebrovascular disease. Temporal arteritis may lead to headache and blindness. Diagnosis of these processes is made by looking for the characteristics of the general disease involving other organs. Blood tests are very helpful in distinguishing these from primary cerebrovascular disease.

After treatment of the acute stroke, rehabilitation becomes of significant importance. Thus, after stabilization an aggressive rehabilitation program may allow for significant regaining of lost function.

Tumors

Ten per cent of tumors within the body occur in the central nervous system (CNS) and its interrelated structures. These include tumors from the primary cells of the nervous system, gliomas; tumors from the meninges, meningiomas; tumors of the pituitary; tumors of the covering of the extended nerves, neurilemmomas; tumors of the blood vessels; and metastatic tumors.

Heredity rarely plays a role in the formation of CNS tumors except in von Recklinghausen's disease and familial phakomatosis (tuberous sclerosis).

In the CNS all tumors become important. The distinction between benign and malignant is relative: some grow slowly and others fast; some are easier to remove surgically; some are radiation sensitive, some are more sensitive to chemotherapy. Clearly, growth in the CNS takes up space; the CNS is enclosed by unmovable bone, and thus space is at a premium. Growth of a tumor creates an increase in intracranial pressure, which results in papilledema and eventually herniation of the brain.

Symptoms of a brain tumor include headaches, vomiting, seizures, decreased level of consciousness, and change in mentation. The neurologic signs are dictated by the location of the tumor. The signs are usually insidious at first but with progression they become apparent.

Glial tumors are graded according to cell type and potential for fast, unimpeded growth. Grade I is more formed and slow compared with grade IV (glioblastoma), which is undifferentiated and rapid.

Astrocytomas are the most common CNS tumor. Cerebellar astrocytomas are common in children and often may be easily removed as a cystic lesion. Brain stem astrocytomas are more common in adolescents.

Ependymomas are derived from ependymal cells along the ventricles and are most commonly found in children and adolescents.

Oligodendrogliomas make up less than 10 per cent of gliomas. they frequently calcify and are slow growing.

Medulloblastoma is the second most common tumor in the posterior fossa in children (cerebellar astrocytoma is number one). It is rarely seen in adults.

Pinealomas cause extraocular muscle impairment of upward gaze and ptosis (Parinaud's sign).

Hemangioblastomas may arise in the cerebellum or medulla. As the name implies, they are blood vessel tumors and can be associated with cysts of the kidney (Lindau's disease) or angiomatosis of the retina (Von Hippel's disease).

Tumors are seen in the middle ear, producing a decrease in auditory acuteness, the glomus jugulare tumor. These appear to arise from embryonic cells and can grow through the petrous temporal bone into the pontine region.

Meningiomas are the second most common tumor of the CNS; they can grow flat (en plaque) or more commonly spherical and encapsulated.

Neurofibromas are the most common tumor of cranial nerve VIII. They also occur in relation to cranial nerves V, IX, and X.

Nasopharyngeal carcinomas may also grow back into the brain stem area and cause problems with cranial nerve VIII.

Computed tomography (CT) and magnetic resonance imaging (MRI) have made the diagnosis of CNS tumors much easier. If the suspected region is heavily encased in bone as at the cerebellopontine angle, MRI is preferable. Treatment depends on the tumor type and location.

Infections

Infections of the CNS may be bacterial, viral, fungal, of unknown cause, or a combination of these. Under normal circumstances infection in the CNS is unusual because of the blood-brain barrier. This physiologic barrier prevents the passage of many substances from the CNS. However, infection occasionally does reach the brain and spinal cord via the bloodstream, through direct trauma to the CNS, via nerve roots, or as a result of poor technique during treatment, eg, surgery or spinal tap. CNS infection may take the form of leptomeningitis, abscess, sinus thrombosis, or a combination.

A subdural empyema is not often seen but develops from infection of the middle ear or its surroundings. It presents with increased intracranial pressure, decreased consciousness, and eventual coma. Focal neurologic abnormalities are not common with this condition. Treatment consists of antibiotics and surgery.

Epidural abscess associated with the petrous temporal bone may occur and produces paralysis of cranial nerves V and VI (Gradenigo's syndrome). Treatment again consists of surgery and antibiotics.

Brain abscess is a more common problem, but because of the availability of excellent antibiotics is not as frequent as it once was. Abscess formation may occur via the middle ear or the paranasal sinuses, from penetrating wounds, from septic emboli, or from septicemia. The most common organisms include streptococci, staphylococci, pneumococci, and meningococci. In immunocompromised individuals other organisms may be predominant. Clinically, abscesses may present as space-occupying lesions. Fever, chills, and generalized illness are often associated with the mass lesion. CT has made the diagnosis much easier. Spinal fluid analysis is almost always contraindicated because of the high pressures associated with the abscess, which increase the risk of herniation. Treatment is with appropriate antibiotics and surgery.

Thrombosis of the dural sinuses is a complication of infections of the middle ear, sinuses, or facial structures. The superior longitudinal sinus, transverse sinus, sigmoid sinus, and cavernous sinus can all thrombose. Symptoms include increased intracranial pressure, headache, seizures, fever, and a decreased level of consciousness. Angiography with delayed films showing the sinuses is used to make the diagnosis. Treatment is with antibiotics and generalized supportive measures.

Meningitis (leptomeningitis) occurs secondary to infection of the cerebrospinal fluid (CSF) with inflammation of the arachnoidal coverings. The organism is often dependent on the age of the individual: in neonates, *Escherichia coli*, streptococci, *Staphylococcus aureus*, and *Diplococcus pneumoniae;* in infants and children, *Haemophilus influenzae*, *Neisseria meningitidis*, and *D. pneumoniae;* in adults, *D. pneumoniae*, *N. meningitidis*, streptococcus, *S. aureus*, and *H. influenzae.* In immunosuppressed individuals different organisms predominate and are somewhat regional. Treatment is with antibiotics. There are new antibiotics every year, and each increases the spectrum of activity.

Tetanus is caused by the tetanus bacillus and its exotoxin. The toxin affects the anterior horn cells and causes severe muscle spasms, resulting in trismus (tightening of the jaw musculature). Opisthotonus of a painful nature may also occur. Treatment consists of debridement of the wound, administration of tetanus immune globulin, and supportive measures.

Fungi may invade the CNS sometimes insidiously and sometimes acutely. Depending on the region of the USA, different fungi predominate. Some are prevalent in immunosuppressed individuals. Treatment is with amphotericin B along with whatever is specifically available for the primary condition.

Encephalitis is caused by viral invasion of the CNS. There are many different viruses that may invade and some are helped by insect vectors. Among the viruses and virus-induced disorders are the equine encephalitides St. Louis and Japanese B; yellow fever; tick-borne, rabies, poliomyelitis, coxsackie, herpes, echo, mumps, and measles viruses; lymphocytic choriomeningitis; influenza; infectious mononucleosis; and cytomegalovirus. Many are self-limited, but it is especially important to diagnose herpes simplex encephalitis because if left untreated it often is fatal. This severe encephalitis often involves the temporal lobes, causing headache, lethargy, irritability, confusion, decreased memory, myoclonic jerks, paresis, and seizures. Early treatment with acyclovir is important and may be life saving. Cytomegalovirus has been seen more frequently in immunocompromised individuals.

Degenerative Diseases

Multiple sclerosis (MS) is a demyelinating disease of the CNS characterized by its extreme variability. Although significant disability is possible, most people with this progressive condition live fairly normal lives. Scattered areas of demyelination produce a variety of neurologic signs and symptoms, commonly including numbness, dizziness, blurred vision, diplopia, bladder disturbances, weakness, and clumsiness.

Trigeminal neuralgia in a young person may be an indicator of demyelination. Bilateral internuclear ophthalmoplegia is thought to represent brain stem involvement by multiple sclerosis. Hearing may be transiently affected but severe hearing loss over a prolonged period is unusual.

Pathologically scattered areas of inflammation with the presence of immunocompetent cells indicate an autoimmune process. Epidemiologic studies point to a predominance in Northern Europeans. The female-to-male ratio is 1.8:1 and the disease is more prominent in temperate regions north and south of the Equator. The distribution favors involvement of a virus, but none has been isolated.

Diagnosis is made in individuals between the ages of 15 and 40 (the commonage group for initial onset of symptoms), who have a fluctuating neurologic process with scattered, multiple abnormalities within the CNS. Evoked response testing, CSF analysis in a search for oligoclonal IgG banding, and MRI may be helpful in pinning down the diagnosis.

Treatment is aimed at decreasing the length of the attacks with cortisone (used as an anti-inflammatory agent), and symptoms are managed with a variety of rehabilitative and medical measures. Immunosuppressive agents may be used if progression of the disease is evident.

There are a few unusual demyelinating diseases of children that may produce brain stem signs and symptoms. The etiology of some appears to be metabolic; others are not clearly defined. These include Schilder's disease, metachromatic leukodystrophy, and Pelizaeus-Merzbacher disease.

The brain stem and upper cervical cord may fall prey to a degenerative process producing literally a hole in the region, a syrinx. In the brain stem the process is called syringobulbia; in the spinal cord, syringomyelia. This may occur secondary to a neoplasm, to congenital problems, to trauma, to infection, or to an unknown cause. The process may be stable or progressive. Symptoms include weakness and atrophy in the extremities with dissociated sensory loss (loss of pain and temperature but preservation of proprioception). The cerebellar tonsils may be low and herniated through the foramen magnum (Arnold-Chiari syndrome).

MRI and myelography aid in the diagnosis. Treatment may involve a surgical procedure.

Amyotrophic lateral sclerosis (ALS) is a progressive disease involving the pyramidal tracts and anterior horn cells. This sporadic killing disease usually progresses rapidly, causing death in 3 to 5 years. A few cases apparently stabilize for long periods. The cause is unknown. Toxins occasionally mimic the process and thus should be evaluated. This disease ascends or descends the spinal cord and brain stem, producing weakness and difficulty with swallowing and respiration. There is no known cure and treatment is with supportive measures.

There are many muscular dystrophies. Common among most is a hereditary influence. Most occur in children, although a few begin in adulthood. The most common is Duchenne's, a sex-linked dominant disease of males. Progressive weakness and wasting of muscles leads to respiratory depression, swallowing problems, and eventual death.

The dystrophies of young adults (limb-girdle, facio-scapulohumeral) are less severe and produce weakness that is often more adaptable.

Diagnosis is made by family history, neurologic signs and symptoms, electromyography, muscle enzymes, and finally muscle biopsy. Treatment is symptomatic.

Dementia may be caused by many factors, including any pathologic process involving the brain. The cause of Alzheimer's (senile and presenile) dementia remains unknown but the pathology is characteristic. Amyloid deposition with degeneration of nerves and the presence of neurofibrillary tangles confirm the diagnosis. Clinically, recent memory begins to falter initially, followed by a general decline of memory that can be significant.

There may be an hereditary background, but sporadic cases are common. Insight, judgment, and reasoning ability are involved. The process is insidious and progressive. Toxins (internal and external), depression, and cerebrovascular disease must be especially checked.

Confusion increases when environment is changed, and thus stability of living situations may prolong the time before nursing home placement is necessary. A number of treatments have been tried, but none has been successful and management is directed toward the behavior with the addition of neuroleptic medication.

Huntington's disease is an autosomal dominant process that may begin in childhood but usually develops after the age of 30. Chorea (rapid, jerky movements) and athetosis (slow, writhing movements) are characteristic. The disease progresses with dementia and usually results in death in 10 to 15 years.

Parkinson's disease is of unknown origin. The parkinsonian process appears to result from a decline in dopamine levels in the substantia nigra, resulting in a general decline of this neurotransmitter in the brain (the striatal-nigral pathway).

Clinically this results in slow movements (bradykinesia); a resting, slow, gross tremor; and cogwheel rigidity. Often a masked face, poor associated arm movements, shuffling gait, seborrhea, dementia, and stooped posture result. The cause of Parkinson's disease is unknown, but clearly atherosclerosis, striatal-nigral degeneration, drug-induced problems (eg, from phenothiazine, the butyrophenones, alpha-methyldopa), post-encephalitic disease, and toxins (manganese, carbon monoxide) may result in a similar clinical picture.

The advent of L-dopa and its derivatives (eg, Sinemet), along with anticholinergic agents and dopamine agonists, has aided the treatment of this problem. In time, problems result with the medicine's metabolism and with sensitivity of the neurons (the "on-off" phenomenon). This produces dyskinesias (abnormal involuntary movements).

Dystonia musculorum deformans (torsion dystonia) is an autosomal dominant disease that occurs within the first two decades of life. The turning of the head into a tonic contraction of the neck generalizes into major spasms affecting the whole body and at times respiratory difficulty.

Spasmodic torticollis is a regional dystonia limited to the head and neck. It may appear less threatening than dystonia musculorum deformans, but it is a severely disabling process psychologically as well as physically. Medication used in combination, including relaxants, antidepressants, and anticholinergics, may be of help.

Dizziness

Dizziness is one of the more common problems seen by both otolaryngologist and neurologist. It is a complaint, a symptom, and not a disease. It results in uncertainty of position or motion in space. There are many classifications of dizziness, but clinically it can be broken

down into four types:

Type I. Vertigo, presenting with a definite rotational component.

Type II. Syncope-like, presenting with a feeling of loss of consciousness.

Type III. Disequilibrium, presenting with multisensory difficulties that produce a lack of balance and coordination.

Type IV. A light-headed sensation, diffuse and non-descript, including all dizzy sensations not otherwise included in types I, II, and III.

Type I dizziness is true vertigo. This implies that a vestibular abnormality is present. This may be a central abnormality, emanating from the vestibular connections in the CNS (vestibular nuclei, tracts in the brain stem, cerebellum, or cerebellar outflow tracts), or may be peripheral (semicircular canals, vestibular nerve). Sometimes the characteristics of the vertigo make it possible to distinguish peripheral from central vertigo. Peripheral vertigo is often explosive, severe, and paroxysmal. Central vertigo is more commonly insidious, continuous, and less intense. Vertical nystagmus and other cranial nerve abnormalities are more often seen in central vertigo. In either case there is a spontaneous and false sensation of movement, a feeling of spinning.

Type II dizziness most often accompanies cardiovascular abnormalities. This may be an arrhythmia with a decrease in cerebral blood flow, or orthostatic hypotension of any other cause. Transient ischemia of a general nature to the posterior circulation may give this picture.

Type III dizziness occurs in persons who lack the appropriate sensory input to maintain normal equilibrium. This can happen in frontal lobe disease. It presents as an apraxia of gait (Bruns' apraxia). Patients walk as if their feet are magnetized to the floor. Strength is usually normal and patients can lift their feet if requested, but cannot maintain the pattern for very long. Cerebellar disease can present with type III problems. Ataxia or incoordination is the result of disease of the cerebellum. Many etiologic entities can damage the frontal lobes or cerebellum; among the more common are multiple sclerosis, cerebrovascular disease, hereditary ataxias, neoplasm, toxins, and infections (discussed later in this chapter). The discrepancy of sensation distorts movement.

Type IV dizziness includes everything else that does not fit neatly into the other three categories. This may include hyperventilation and psychological or functional problems.

To stave off dizziness, patients need their senses to be fully operational. These senses include vision for which the vestibular nuclei have primary visual afferents that bypass the occipital lobe) and vestibular sensation. Acceleration is measured by vestibular sensory inputs. Proprioception from the limbs provides a relationship of the body to the head and allows for smooth movements. Touch plays a role via pressure sensations from the feet and even from

clothing. Hearing adds a major measure of sensation allowing for proper placement of the head and body in space. Obviously, if any of these senses are lost, compensation can take place, but acute loss results in significant dizziness.

The neurologic workup of dizzy patients begins with an appropriate history. The age of the individual sets the stage of the differential diagnosis. Older patients are more likely to have vascular disease, Parkinson's disease, frontal lobe dysfunction, Ménière's disease, or chronic labyrinthine imbalance. Younger patients develop infections, tumors of the brain stem, benign positional vertigo, acute and recurrent vestibulopathy, and multiple sclerosis. Drugs can affect any age group.

It is especially important to determine whether a change of position or motion has any effect. This distinguishes positional vertigo from positioning vertigo. Positional vertigo occurs after a movement has taken place; positioning vertigo occurs as the movement is being carried out. The latter is nonspecific. The former is seen in the entity benign positional vertigo. In this process there is a latent period of seconds followed by nystagmus (rotational or horizontal) and a sensation of disaster. The key is the latency before the onset of nystagmus. The nystagmus adapts in 30 seconds to a minute. It adapts, habituates, is direction fixed, and does not feel good. However, it is benign and not indicative of significant structural disease.

The course of the process also helps in finding a cause. An abrupt course signifies vestibular or cardiovascular disease. Continuous disease usually leads to a psychiatric diagnosis. Gradual onset and course is nonspecific.

An attempt should be made to reproduce the dizziness. Orthostatic blood pressure measurements are necessary. A Valsalva maneuver may reproduce syncope and its attendant dizziness. Gentle carotid sinus stimulation may be of help. Hyperventilation can be induced by deep breathing, and if the symptoms match a diagnosis may be easily made.

Positional testing (Barony and Hallpike maneuvers), turning while walking, neck twisting, electronystagmography, audiometry, brain stem auditory evoked response testing, MRI, complete blood count, Holter monitoring, cervical spine x-rays, and the Minnesota Multiphasic Personality Inventory (MMPI) may increase diagnostic accuracy.

Treatment of vertigo ideally is dependent on the cause, which thus it is essential to seek. However, the exact etiology often is not obvious, in which case general measures should be taken. Initial treatment is often with hydration plus meclizine or diphenerimine. If these fail, scopolamine is often introduced. Diazepam derivatives directly inhibit the labyrinth and may settle a troublesome vertigo. Intravenous procaine is a dated but nonetheless occasionally helpful agent. Vertigo may be an extremely disabling symptom and needs to be taken very seriously. The therapeutically oriented neurologist is continuously searching for ways to manage the dizzy patient.

Pain

Pain in and about the head can be divided into three general categories: (1) headache, (2) neuralgia, and (3) atypical facial pain. The pain-sensitive structures intracranially include the skin, bone, dura, and blood vessels. Any headache of significant intracranial pathology almost inevitably involves the blood vessels. Problems with the brain itself do not lead to pain without traction on the above structures.

Headache can be divided into two broad categories: throbbing and nonthrobbing. Proper management of headache is based on proper diagnosis. It is far preferable to treat the cause of a problem than to treat only the symptom. Thus, pain pills should be used sparingly in headache management. To make a proper diagnosis, five questions must be answered:

1. What is the type of headache: throbbing or non-throbbing?

2. What is its location?

3. What is its frequency?

4. What is its duration?

5. What are the associated symptoms?

There are four headaches that present in a nonthrobbing manner. First is the ocular headache, located around the eyes, and often found when the eyes are being used for fine work. It tends to clear overnight or when the eyes are not being taxed. It is associated with decreased vision. Despite the belief that this is a common headache, it is actually quite uncommon. Often the first physician sought by patients for headache diagnosis is the ophthalmologist. Patients nurture the false belief that the eyes are a common cause of headache. When the eyes are indeed the cause, proper treatment involves correction of the visual problem.

Second, sinus aches are a relatively rare cause of headache. They present as a nonthrobbing ache located over the sinuses. They are worse in the morning, and as the sinuses drain the ache improves. They are frequent and last until drainage or treatment relieves the pressures. They are often accompanied by fever, stuffiness, and an ill feeling. Treatment consists of antibiotics, antihistamines, drying agents, and possibly surgery. Chronic pain medications should not be used. A head and neck surgeon may see this condition more frequently, but it is not a common cause of headache.

The third cause of nonthrobbing headaches is far more common and is the result of disease of the cervical spine and its associated structures. Cervical disease is caused by whiplash (to-and-fro) injuries in the young, and often by degenerative joint disease (osteoarthritis) in the older population. The ache is located in the back of the head, spreading up from the neck. It is present all of the time and is relieved only intermittently. It may be associated with numbness

or weakness of an arm (nerve root compression). Treatment of this type of headache includes physical therapy (heat, massage, ultrasound, and gentle traction). Skeletal muscle relaxants, anti-inflammatory medications, and mild analgesics are useful. Imaging (x-ray, CT, and MRI) may have a limited diagnostic value.

Muscle contraction (tension) headaches are by far the most common type of nonthrobbing headache. Muscle contraction aches may also throb as the muscles of the forehead, back of the head, and temples clamp down around blood vessels and nerves and cause pain. These are usually associated with tension, depression, frustration, or anger, but they can occur in the absence of these. Thus, they are properly termed muscle contraction aches. They are frequent aches that may never retreat. The simple, routine "tension" headaches do not get as far as the physician's office; they are managed at home and present no management problem. To treat difficult muscle contraction headaches properly, the physician must understand that the symptoms result from the conversion of anxiety into a physical problem. The patient lacks insight into the anxiety and usually does not wish to obtain that insight. Thus, the immediate suggestion of counseling is received with a lack of enthusiasm. Mild analgesics given in combination with increasing dosages of tricyclic antidepressants may be of value. Patients often have accompanying vegetative signs of depression, including sleep and appetite disturbances. As patients begin to feel better, they are often much more amenable to counseling.

Throbbing headaches are far more common than the nonthrobbing variety. As stated, the muscle contraction headache may present in a throbbing manner. Throbbing implicates the blood vessels. The most common vascular headache is the migraine, of which there are many types. Only four are discussed here.

The classic migraine is a hemicranial headache that is usually inherited. It presents with a warning (aura) of flashing lights, strange feelings in the stomach, and increasing anxiety. The ache is often explosive and accompanied by nausea, vomiting, hyperacusis, and photophobia; this may last 2 to 24 hours. Rest may help to alleviate the symptoms. Pathophysiologically, there is a vasoconstriction followed by vasodilatation. The stretching of the walls of the vessels and the pain fibers within creates the pain. Serotonin, bradykinin, prostaglandins, and other chemicals are released. Migraines may occur seldom (less than once a year) or frequently (every 2 to 3 days). If the aura is nonfocal and definite, vasoconstricting medications (ergot alkaloids) may be taken to prevent the painful phase. Care must be taken not to overdose with these, as medication-induced vasoconstriction will be occurring elsewhere in the heart, kidneys, and extremities. If the aches are frequent, migraine prophylaxis is necessary. If taken on a regular basis, beta blockers, calcium channel blockers, and tricyclic antidepressants all help to prevent migraines. Pain medications rarely offer comfort and should be used sparingly. During the acute phase of the headache, adrenocorticotropic hormone (ACTH) or cortisone may break the cycle and is preferable to narcotics.

The distribution of common migraine differs from that of the classic variety and is often the whole head. There may not be an aura, and thus the value of vasoconstricting agents may be limited; otherwise, management is similar.

Complicated migraine is similar to classic migraine, but during the aura phase the vasoconstriction is so intense that focal cerebral ischemia results. This transient ischemia presents with focal neurological symptoms such as paresis, aphasia, and hemianopia. Under these circumstances the vasoconstricting agents used in classical migraine may lead to a cerebrovascular accident, and are thus contraindicated. Management with the other types of prophylactic agents is necessary.

Ophthalmic migraine presents with pain behind the eyes that is often burning, stabbing, and severe. There usually is blurred vision. Nausea is also frequent. Management is similar to that for other types of migraine.

Cluster headaches are often placed in the migraine category but represent a distinctive vascular headache. As the name implies, they occur in clusters, often involving part of the head. There often is unilateral eye pain and Horner's syndrome. Alcohol (a vasodilator) makes all vascular headaches more severe, but this is especially true for cluster aches. Clusters tend to occur more often in males. Treatment as for migraine may be effective, but longer-term steroids are often necessary. Lithium carbonate may be of value in this type of headache.

Brain neoplasms produce headache via traction on vessels. It may initially be nonthrobbing but throbbing soon becomes evident. The headache may be intermittent at first but becomes more regular over time. The location of the ache may be somewhat removed from the tumor because of the intracranial traction, but it is often deep in the region of the tumor. Antiedema agents may be useful in decreasing the intensity of the headache.

Postspinal headache results from the leakage of CSF through the hole produced by the spinal needle. This miserable, throbbing ache over the whole head occurs in the upright position and disappears in the lying position. Management involves the patient remaining recumbent. Fluid intake should be increased. Occasionally a patching of the dural tear with homologous blood, allowed to clot in the epidural space (an epidural blood patch), is effective.

There are three headaches that present with severe pain and cannot be distinguished on first appearance. Migraine, encephalitis-meningitis, and subarachnoid hemorrhage all present in a similar manner. Fever may be a clue to infection but is not an absolute indication. Stiff neck may indicate arachnoid irritation but also is not absolute. Only spinal fluid analysis allows differentiation: encephalitis-meningitis involves white blood cells; subarachnoid hemorrhage involves red blood cells; migraine involves no cells. After this test, the primary problem can be treated.

Toxic-metabolic, withdrawal headache (eg, as during a hangover or the withdrawal from caffeine) is produced by vasodilatation. The diagnosis is usually self-evident, the condition is normally self-limited, and management is symptomatic.

Proper headache management is based on proper diagnosis. When the mode of presentation is known, a satisfactory result is often achieved.

Neuralgia

Neuralgia is the result of an irritative disturbance to a peripheral nerve. Cranial neuralgias are secondary to cranial nerve irritation. Irritation of cranial nerve I (olfactory) may produce parosmia, an unusual smell; irritation of II causes scintillating scotoma or teichopsia (viewing of wavy lights); irritation of III produces hippus (instability of the pupil); irritation of V causes tic douloureux or trismus; irritation of VII produces hemifacial spasm; irritation of VIII results in tinnitus or vertigo; irritation of IX causes glossopharyngeal neuralgia; and irritation of X produces hiccoughs.

Painful neuralgias include ciliary neuralgia, tic douloureux, and glossopharyngeal neuralgia. The trigeminal nerve (V) has three branches: ophthalmic, maxillary, and mandibular. Irritation of the ganglion or the nerve may produce a severe, lightning-like pain along the distribution of the nerve. It is typical for a trigger point to be present. The pain is severe and unrelieved by analgesics. Relief may be accomplished with appropriate anticonvulsant treatment. Carbamazepine (Tegretol) is quite effective, whereas phenytoin (Dilantin) is less effective. Surgery (cryotherapy or alcohol injection) is also useful if the medication regimen fails.

Glossopharyngeal neuralgia is located in the posterior pharyngeal region. One-half of the face may also be involved. There is no trigger point. Cocainization of the pharynx may temporarily relieve the problem. Treatment with carbamazepine is appropriate.

Ciliary neuralgia involves one-half of the face (the forehead and lower face) and is accompanied by a reddened eye and teichopsia. This is extremely rare.

Raeder's paratrigeminal syndrome involves pain along nerves V1 and V2. There is ptosis and meiosis, with no loss from sweating. The cause is unknown but the condition appears to be secondary to damage to the oculosympathetic fibers in the sheath of the internal carotid artery.

Sphenopalatine (Sluder's) neuralgia presents as a paroxysmal problem. Sneezing or congestion usually precedes the attack. Unilateral pain then follows in the region of the nose or the eye, extending back to the dura and associated with photophobia, lacrimation, and salivation. Occasionally a metallic taste is noted. The attack may last 10 to 30 minutes, three to four times a day. Treatment involves anesthetizing the sphenopalatine ganglion.

Occipital neuralgia presents with pain, possibly paroxysmal, of a sharp, twisting nature in the region of the posterior head (the occipital nerve). Examination results are negative. Treatment is with carbamazepine and steroids.

Temporomandibular joint disease may present with pain on opening and closing the mouth. There may be pain on pressure over the joint, with crepitance over the joint. The jaw may deviate with movement. Common causes include trauma, arthritis, and malocclusion. The pain may spread and be quite atypical. Workup and treatment are aimed at the joint and are discussed elsewhere in this book.

Atypical facial pain by definition is facial pain that is not classifiable into a routine clinical picture. If the cause of the facial pain is not evident, there is a strong likelihood statistically that depression is playing a major role. Thus, if after exhaustive workup no abnormality is found, consideration should be given to treatment of depression with medication and counseling.

Diplopia

Double vision, diplopia, is one of the more common neurologic symptoms caused by disease in the head. The control of extraocular movement is relatively complex but usually involves the brain stem. The nucleus of cranial nerve III lies in the midbrain, that of IV sits just posterior to it, and that of VI posterior to that in the pons. Together these make up the nerves to the extraocular muscles. Cranial nerve VI innervates the abducens, which pulls (abducts) the eyes outward; IV innervates the superior oblique, which pulls the eye downward, and rotates it outward with torsion. All the rest of the eye muscles are directed by cranial nerve III. The coordination of eye movement through these nerves is accomplished by a thinly myelinated fiber tract that connects nerves III, IV, and VI with input from the ear (the vestibular apparatus) and the cervical (neck) muscles. This tract is called the medial longitudinal fasciculus (MLF). Lesions along its distance cause abnormalities of coordination of eye movement, leading to nystagmus that is different in each eye, dissociated nystagmus. This is called an internuclear ophthalmoplegia (INO). In its classic form a lesion of the right MLF causes a paresis of that eye's movement to right lateral gaze, while the left eye develops gross nystagmus on gaze to the right. The opposite is true for a left MLF abnormality. Bilateral INOs are seen most commonly in multiple sclerosis, while any structural disease of the brain stem can cause a unilateral MLF (INO) syndrome.

Cranial nerve III leaves the brain stem and traverses the cavernous sinus to the eye, carrying the parasympathetic fiber tracts right alongside. As it travels adjacent to the tentorium, it is vulnerable to pressure from space-occupying lesions in the head. This results in dilatation of the pupil, resulting from the unopposed sympathetic stimulation. The lesion is most often ipsilateral to the pupillary dilatation. A unilateral cranial III nerve palsy without other signs of brain stem dysfunction presents as an eye that can move laterally and not much in any other direction. There is a ptosis of the eyelid resulting from paresis to the levator palpebrae superioris (also a cranial nerve III muscle). The pupil is large and unreactive. Diabetes mellitus is the most common cause of this isolated process, although structural disease adjacent to the nerve or its nucleus must be ruled out. Structural disease includes an aneurysm of the posterior communicating artery, tumor of the brain stem, multiple sclerosis, and atherosclerotic vascular disease of the brain stem. Migraine may also result in ophthalmoplegia via vascular spasm. Myasthenia gravis, an autoimmune disease of the synapse, may also mimic localized cranial nerve III disease.

Disease of the orbit may entrap any or all of the extraocular nerve muscles, giving the appearance of a neuropathic process. Hyperthyroid disease may result in a hypertrophic exophthalmopathy with invasion of the eye muscles, and the resultant entrapment can appear as

extraocular neuropathy. This can also occur in the absence of known endocrine disease and results from inflammation about the orbit, causing the syndrome described as a pseudotumor of the orbit (Tolosa-Hunt syndrome). This painful process may be difficult to diagnose and can be confused with a thrombosis of the cavernous sinus, which also causes entrapment of the nerves and painful ophthalmoplegia and exophthalmos.

In the absence of special equipment, cranial nerve IV paresis is hard to diagnose. The superior oblique muscle acts to depress and externally rotate the eye. Trauma to the orbit of a relatively minor degree may cause a malfunction of this nerve; the head then turns to avoid the double vision. Rest usually heals the condition.

Cranial nerve VI (the abducens) travels a very long distance to get to the lateral rectus, and consequently is a prime nerve to be irritated by any increase in intracranial pressure. The space-occupying lesion may be far away from the nerve involved, and thus may be a false localizing sign. Diabetes mellitus, thyroid disease, multiple sclerosis, and atherosclerotic cerebrovascular disease may all affect this nerve. Diagnosis is relatively easy when the eye fails to abduct. Sometimes, more sophisticated cover testing with a red glass or Maddox rod is necessary.

The workup of an extraocular palsy begins with thorough history taking. Often, related complaints can make the diagnosis of multiple sclerosis, migraine, diabetes, thyroid disease, or even tumor relatively easy.

CT or MRI in combination with a good thorough neurologic examination offer a noninvasive way to diagnose disease of the orbit, brain, or brain stem. If results from these are negative, angiography may be necessary to rule out aneurysm. Edrophonium (Tensilon) testing can be performed to exclude myasthenia gravis.

Treatment obviously must be aimed at the primary problem. Occasionally, prisms may be of value if the diplopia is constant and uncorrectable in any other manner.

Facial Nerve

The anatomy of the facial nerve is relatively complex and discussed in detail elsewhere in this text. Suffice it to say that, from a neurologic point of view, it is a very long nerve. It exits from the pons and traverses a bony canal before it reaches its goal of innervation of the muscles of facial expression.

Complicating the clinical neuroanatomic expression of the facial nerve is the fact that, like many other cranial nerves, its nucleus receives bilateral input from the cerebrum. Because of this bilateral input, lesions of the cerebrum and its connections through the internal capsule spare the forehead. This sparing occurs because the input from the ipsilateral cortical tract remains untouched. The lower face is not blessed with this "double" innervation. Thus, upper motor neuron lesions in the brain often result in a loss of facial movement of the lower face

contralateral to the lesion. Any disease process that interrupts these tracts may contribute to this weakness. These include, but are not limited to, trauma, tumor, degenerative disease, and occasionally infectious disease.

Diseases of the brain stem or peripheral nerve (VII) lead to loss of movement in both the forehead and lower face ipsilateral to the process.

Because treatment may alter the course of some facial nerve (or nucleus) palsies, it is incumbent upon the clinician to attempt to determine the cause. Bell's palsy is one of the processes involving the facial nerve that is not clearly understood. It is assumed that something (possibly a virus) has caused inflammation along the course of the nerve, inducing swelling in the bony canal, which leads to compression and facial palsy. The patient presents with loss of movement of the upper and lower face. The eyelid is involved and often will not close under its own power. In most Bell's palsies, time usually solves the problem. The eye must be protected and kept from scarring, aided by appropriate lubricants and coverings. The judicious use of cortisone may decrease the length of disability and increase slightly the quality of recovery.

If the nerve is "stunned" in the facial canal, apraxia results. Because the nerve is not destroyed faster, more complete healing is common. Steroids decrease the inflammation and allow recovery to proceed more rapidly. A suggested regimen is dexamethasone, 4 mg po qid for 3 days, followed by 4 mg po tid for 3 days, then 4 mg bid for 3 days, then 4 mg po qd for 3 days. If more than a stunning of the nerve has developed and destruction has been predominant, the healing process is slower: it takes time for the nerve to regrow. If signs of improvement are at all obvious, the clinician must be patient and express confidence in repair. Very rarely the new growth of nerve is aberrant and can result in anomalous innervation, so that the eye closes when the jaw is opened (jaw winking).

If there is no apparent regrowth of the nerve after 6 to 8 weeks consideration must be given to surgery on the nerve and its canal. Most neurologists are conservative in considering surgery for this condition, since their experience and studies indicate an overwhelming healing rate.

It should not be assumed that every facial palsy is idiopathic Bell's. The herpes zoster virus (Ramsay Hunt syndrome) may be invasive and produce a similar picture with vesicles in the ear canal. Treatment is again with steroids. Acyclovir may be helpful in the healing process.

Lyme disease caused by a tick bite and infusion of a spirochete may also be a culprit; thus, a history of a well-defined red rash, systemic illness, malaise, and lymphadenopathy should be sought. Treatment of this is with tetracycline or penicillin.

Demyelinating disease (multiple sclerosis) may present with a facial palsy.

Facial palsy may be evident in brain stem stroke and basilar meningitis (tuberculosis, sarcoid), but these patients usually present critically ill and the diagnosis is not often confused

with a "routine" Bell's.

The Möbius syndrome is a congenital process involving the brain stem that results in paralysis of the facial muscles and difficulties with eye movement.

Guillain-Barré syndrome, a disease with a viral, autoimmune connection, may present with facial weakness. It is usually bilateral and may progress to (or from) weakness elsewhere in the body. At presentation it may be confused with Bell's, but when limb weakness and dysphagia develop differentiation is not difficult.

A number of eponyms have been associated with the facial nerve and its various syndromes (Table 2).

Table 2. Conditions of the Facial Nerve

Disorders of Facial Paralysis

Bell's palsy: peripheral facial paralysis of unknown etiology, thought to be viral.
Millard-Gubler syndrome: lateral rectus paralysis accompanied by ipsilateral facial paralysis.
Foville's syndrome: ipsilateral paralysis of conjugate gaze with ipsilateral facial paralysis.
Ramsay-Hunt syndrome: peripheral facial paralysis secondary to herpes zoster infection.
Melkersson's syndrome: recurring facial palsy, facial edema, congenitally fissured tongue.
Möbius syndrome: facial weakness with extraocular muscle weakness, particularly of abducens.
Brissaud's syndrome: hemifacial spasms with opposite side pyramidal involvement.

Facial Nerve Reflexes

Orbicularis oculi reflex: produced by percussion at outer aspect of supraorbital ridge over glabella, followed by reflex contraction of eye.
Myerson's sign (glabellar reflex): persistent orbicularis oris reflex.
Palpebral reflexes: reflex contraction of orbicularis oculi muscle to various stimuli.
Oculogyric-auricular reflex: retraction of auricle on extreme lateral gaze.
Palpebraloculogyric reflex: eyeballs turn upward with closure of eyes.
Orbicularis oris reflex: contraction of ipsilateral lip on percussion of upper lip.
Snout reflex: abnormal orbicularis oris reflex.
Palmomental reflex: ipsilateral contraction of chin after stimulation of palm.

Dysphagia

Dysphagia (swallowing difficulty) is a very common symptom referable to the head and neck. It is present in a number of disease entities that involve the brain stem (bulbar) or the supratentorial innervation of the brain stem (pseudobulbar). Care must be taken to differentiate

neurologic disease from local esophageal or pharyngeal disease. This may involve cine studies as well as esophageal motility and pressure studies.

Vascular disease may cause swallowing problems by involvement of the posterior circulation. Dysarthria almost invariably accompanies the dysphagia. Dizziness and sometimes vertigo may precede the condition. Ataxia and diplopia are also common.

If the vascular disease involves the cerebrum bilaterally with small strokes (lacunae), the control of the brain stem is altered and apraxia of swallowing results. Usually there is accompanying dementia. Emotional lability is prominent. This is called a multi-infarct dementia or pseudobulbar palsy.

Bulbar palsy may be caused by vascular disease of the posterior circulation, but also is inevitable with amyotrophic lateral sclerosis. This moderately to rapidly progressive degenerative disease usually attacks adults aged 40 years and over with a progressive weakness secondary to anterior horn and corticospinal tract degeneration. When the brain stem becomes involved, swallowing, speech, and breathing are affected. This deadly disease leaves the intellect untouched. Death usually occurs within 3 years, although occasional remission has been reported.

Guillain-Barré syndrome may cause severe bulbar palsy and accompanying swallowing and speaking difficulties. This potentially reversible disease appears to have a viral origin and may progress to respiratory failure. However, unlike amyotrophic lateral sclerosis, the usual course of this polyradiculopathy is to reverse, allowing complete or almost complete recovery.

Multiple sclerosis in its more severe form may cause dysphagia of both a bulbar and a pseudobulbar variety. This CNS demyelinating disease initially affects people between 20 and 40 years of age and characteristically fluctuates throughout the course.

Myasthenia gravis is a rare cause of dysphagia. It results from an autoimmune reaction against the post-synaptic receptor cells, which causes impaired neuromuscular transmission. Treatment is aimed at the autoimmune process with steroids, plasma exchange, or (more commonly) antiacetylcholinesterase medication (eg, Mestinon).

Brain stem tumors, gliomas, meningiomas, and ependymomas may cause bulbar palsy and dysphagia. Treatment with radiation and/or surgery is individualized, depending on tumor type and location and the age of the patient.

Poliomyelitis is now rarely seen as a bulbar palsy but at one time was quite common. This viral disease attacks the anterior horn cells of the spinal cord, causing paralysis.

Other less common causes of dysphagia include Parkinson's disease, myopathy, myositis, peripheral nerve palsies, botulism, and diphtheria.

Routine management involves dietary manipulation. Mechanically ground food appears

to be swallowed easier by patients with bulbar and pseudobulbar palsy. Milk and milk by-products tend to stick and thus must be used with care. Clear liquids may be aspirated, but individualized treatment is essential. Management of the primary problem is the most practical therapy. At times feeding tubes are the only way to avoid aspiration.

Dysarthria

Dysarthria and dysphagia often accompany each other and the same causes, by the same mechanisms, are involved. Phonation is the sound produced by the vocal cords vibrating; thus, dysphonia occurs when a cord is paralyzed. Hoarseness results when the cords are not working properly. Causes include laryngitis, Parkinson's disease, cerebellar disease, cerebral disorders, and paralysis of the recurrent laryngeal nerve (X) secondary to inflammation or tumor.

Aphonia implies total bilateral vocal cord compromise because of paralysis or laryngeal disease. Hysteria is a common cause of an "aphonic-like" problem.

Articulation is the sound produced by the lips, teeth, tongue, and palate. This depends on neuromuscular control. Cranial nerve VII (facial) paralysis produces problems with labials and dentals (P, B, F, V); nerve IX paralysis results in palatal weakness and nasal speech; nerve XII paralysis produces a weak tongue and problems with linguals (Th, S, Z, Sh, L, R).

Cerebellar disease may change speech to a labored, irregular, explosive variety: staccato speech. Speech may also be slow and jerky, each syllable appearing as a separate word: scanning speech. A good speech pathologist may be of value in making the patient more understandable.

Tinnitus

Tinnitus may be extremely bothersome. The ringing may be in one ear, in both ears, or not localized. Despite its significant disabling characteristics, tinnitus is rarely associated with primary neurologic disease. It occurs in Ménière's syndrome, after trauma to the ear, and occasionally in acoustic neuromas and other neoplasias (glioma, nasopharyngeal carcinoma, meningioma). Neurologically it is usually a frustrating, nonlocalizing, untreatable symptom. Workup should take into account these associated disorders and then be directed toward a primary otolaryngologic etiology.

Seizures

Seizures are a symptom of an electrical dysfunction within the brain, not in a diagnostic category of their own. They may occur because of a primary problem within the brain as seen in brain tumors, scars, stroke, degenerative brain disease, and infection of the brain. They may also be noted with toxic-metabolic problems of the body when the brain is affected by changes in body chemistry. This commonly occurs with glucose, oxygen, and other common metabolic dysfunctions.

The key to the management of seizures is the proper diagnosis of the cause. No matter what the problem, the etiologic process must be treated. If the process is toxic-metabolic, that dysfunction must be rectified before management of the seizure itself can be effective. If the problem is a primary brain process, that process must be treated (eg, stroke, tumor). Appropriate anticonvulsant medication is then given and the proper dose and route of administration become important. Seizures that start deep within the brain and spread over the whole brain simultaneously are called generalized seizures. Medications for control of these include valproic acid, carbamazepine, phenytoin, and phenobarbital. Seizures that start in a more peripheral location within the brain and then spread from there are called partial or focal seizures and are managed with phenytoin, carbamazepine, and primidone. Seizures that continue and are prolonged are termed status epilepticus. These are managed with low doses of diazepam followed by loading (large) doses of phenytoin or phenobarbital. Proper oxygenation during the seizure is important to ensure a good outcome.