Textbook of Oral and Maxillofacial Surgery

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Chapter 26

Neurological disorders of the maxillofacial region

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The neurological apparatus of the maxillofacial region is unique in many respects. It has special anatomical characteristics such as the highest sensory innervation density in the body and a peripheral intermingling of major cranial nerve branches whose central connections do not follow the orderly segmentation typical of spinal cord innervation. Developmentally, oral tissues are the first to respond reflexly to in utero tactile stimulation. These tissues simultaneously perform the vital processes of feeding, sensory perception, respiratory activity, and external communication by means of facial expression and speech. In spite of their importance to the organism, maxillofacial nerves course tortuously through bony crevices and canals and run dangerously close to cutaneous and mucosal surfaces where they are vulnerable to various injuries. The terminal branches must coexist with local tissues that have an extremely high pathological incidence: the paranasal sinuses, the teeth, and the periodontium. Therefore, it is not surprising that the maxillofacial neurological apparatus is a prime target for characteristic pathology that is frequently different from neuropathology in other body regions.

There are maxillofacial neurologica disorders of somatic sensation and visceral and motor activity. Many are responses to acute lesions in adjacent tissues, such as partial seventh nerve paralysis that results from advancing parotid gland carcinoma. Others are part of systemic disease states such as diabetic trigeminal neuropathy. The pathological condition may be primary to the nerve tissues, as in trigeminal neuralgia caused by multiple sclerosis. Finally, when there is chronic facial pain, the symptoms themselves may take on the dimensions of a disease process, since neurological disorders in the facial region carry an especially high emotional impact for many individuals.

The diagnosis of maxillofacial neurological problems depends on an orderly process of (1) interpreting the symptoms and signs of altered neurophysiology, (2) determining the anatomical localization of the disease process, (3) understanding the basic pathological processes that exist, and (4) when possible, identifying the etiological or precipitating factors. Treatment of these problems is a challenge to practitioners from many disciplines, and effective action may depend on a team approach. However, the dentally educated clinician is in a position to make a strong contribution in this field because of his extensive knowledge of maxillofacial symptomatology, anatomy, physiology, and pathology.

Psychophysiology

Terminology. The clinical effects of alterened nerve physiology are identified by certain useful terms. *Paralysis* means loss of or impairment of motor function in a body part,

and *paresis* is incomplete paralysis. Although these terms are usually reserved to describe neuromuscular deficits, they may also be applied to malfunction in autonomic nerves. *Anesthesia* refers to the loss of any and all sensation and should be distinguished from the loss of specific sensations, such as *ageusia,* the loss of taste, and *analgesia,* the loss of sensitivity to painful stimuli. *Hyperesthesia* means excessive sensitivity, and *hypoesthesia,* also called *hypesthesia,* refers to diminished sensitivity, usually to touch. *Hyperalgesia* is an excessive sensitivity to painful stimuli, and *hypoalgesia* implies lowered pain sensitivity.

In cases of altered sensitivity, the concept of thresholds is introduced, and two clinically useful pain thresholds have been described: pain detection and pain tolerance thresholds. The *pain detection threshold,* which is the lowest level at which a given stimulus is considered painful, is known to be remarkably similar for most humans and is influenced little by minor environmental factors. The detection threshold is altered in rarely occurring cases of congenital insensitivity to pain and in neuropathological conditions such as hyperalgesia resulting from incomplete nerve regeneration. The *pain tolerance threshold* is the level of maxillary tolerated stimulus and is highly variable between individuals as well as for a given person tested at different times. Tolerance is greatly influenced by cultural, psychological, and environmental factors, and it is this pain threshold that is often changed therapeutically by pharmacological and hypnotic techniques.

Sensory dissociation refers to a loss of certain senses with the simultaneous maintenance of other senses. For example, sensory dissociation is seen in incomplete local anesthetic block, in which fine tactile and pinprick sensitivity is lost, although proprioception and deep pain awareness persist. The terms paresthesia and dysesthesia are both used to describe abnormalities of sesory quality and both occur spontaneously. A *paresthesia* represents any altered sensation and may be described as itching, tingling, numbness, crawling, or feelings of tissue fullness or swelling. A *dysesthesia* is a painful paresthesia and may be reported as burning, boring, or stabbing and occasionally may be a sensation of "phantom" pain such as the awareness of a previously extracted tooth or a burning tongue after glossectomy.

Components and mechanisms of pain. Pain may be defined as an unpleasant experience that involves three main components: (1) perception, (2) affect or emotion, and (3) reaction. The first proposed component, somatic sensory perception, raises the question of whether pain is really a specific sensation like vision, touch, and taste, which have specific forms of energies in the environment, specialized receptors that may be excited by chemical mediators, and transmission along precise pathways to isolated brain centers. Clinical support for this specificity theory of pain comes from the observation that pathology and surgery at many levels of the nervous system may induce a sensory dissociation in which pain is eliminated and other primary sensations retained. Indeed, a class of nerve fibers of relatively small diameter, 1 to 5 microns, have been identified as *nociceptors,* or fibers that respond specifically to noxious stimuli, meaning stimuli that have the potential to produce tissue damage. It is generally agreed that the nociceptors are unmyelinated or thinly myelinated fibers of the *A delta* and *C* type that are activated by specific biogenic amines such as serotonin and prostaglandin AG. Current theories stress that transmission of "pain impulses" to higher integration centers is greatly influenced by the balances and interactions between the A delta and C fiber nociceptors and the larger A alpha and beta fibers. The net effect of large fiber activity on the transmission of nociceptive information beyond the primary brain stem trigeminal synaptic region is an inhibitory one. Conversely, the smaller incoming fiber components, if unchecked by the inhibitory larger fibers, would tend to encourage the transmission of pain impulses to higher centers. This physiological phenomenon is known as

"afferent inhibition" and is a basic tenet of the gate control theory of pain as described by Melzack and Wall.

A key point of afferent inhibition for the maxillofacial region and the "gate" through which pain information must pass is thought to be located in the caudal portions of the brain stem trigeminal synaptic regions, specifically in the subnucleus caudalis of the descending trigeminal tract. From this theory it follows that any process that disturbs the inhibitory balances of incoming small and large fiber populations that arrive at the subnucleus caudalis would have the effect of changing the amount and nature of pain information transmitted to higher centers. Later it will be shown that many conditions of maxillofacial neuropathology may upset the balances of incoming fiber populations. More recent extensions of the gate control pain theory have stressed that afferent inhibition at the brain stem gate also may be influenced by the activity of fibers that converge on this area from other brain regions such as the reticular formation and the cortex (see *Descending control system)*. The descending control neurons converge on the trigeminal "gating" neurons of subnucleus caudalis from a number of higher centers in the brain stem, particularly from the reticular gray substance surrounding the aqueduct of the midbrain. The midbrain periaqueductal gray (PAG) region is important because it is a well-studied part of the *opiate receptor* system, the sites of action of the opiate analgesics. The PAG region is also of interest because it is activated not only by exogenous opiates but also by a recently discovered class of endogenous polypeptides, the endorphins, or endogenous morphinelike substances. The endorphis appear to originate as products of the hypothalamus, are secreted by the anterior pituitary in response to stress and excitement, and are distributed throughout the body as well as to the opiate receptors. One of the functions of the endorphin system, therefore, appears to be an intrinsic changing of pain threshold levels at the gating level brought about by an activating of the opiate receptor-descending neurons.

There is another vey important link to the opiate receptor-descending control system. A bidirectional pathway links the midbrain PAG region with the limbic paleocortex, the seat of human emotion, arousal, and memory functions. These connections provide a partial anatomical explanation for the well-known influence that emotion, personality, learned behavior, psychopathology, and culture have on pain threshold responses.

When pain impulses pass through the "threshold" gate in the trigeminal system, they travel rapidly and directly along discrete ventral secondary ascending tracts in the brain stem to synapse in the lateral thalamus and finally distribute diffusely to many areas of the neocortex (see *Perception component)*. Neurosurgical sectioning of the trigeminothalamic tracts eliminates the perceptive and informative phase of the pain experience. But, as is unfortunately demonstrated in many of these cases, the perceptive phase of pain does not constitute all the pain experience.

The second major component of pain, affect or emotion, is also inseparable from the pain experience. Perception of a noxious stimulus without feeling emotion is not recognized as pain by the individual. It is suspected that the affective phase of pain is carried in entirely different anatomical pathways and reaches brain integration regions that are far different from the perception component. Specifically, fiber projections from the nucleus of the descending tract of the trigeminal nerve enter the reticular formation at many levels of the caudal brain stem core where they activate diffuse and multisynaptic fiber groups. The reticular formation in these regions also contains the vital centers for maintenance of consciousness and for control of basic cardiac and pulmonary function. These anatomical associations may explain why painful stimuli evoke profound reflex changes, both in conscious attention levels and in

heart and lung function (see *Vital center effects).* Anterior to these vital centers in the midbrain gray matter and in the medial thalamus are reticular formation nuclei that probably receive pain-related fiber projections and whose stimulation brings about an extreme aversion response. These reticular centers project forward to many regions of the paleocortex in the ventral core of the brain, known collectively as the limbic lobes. The limbix cortex has long been considered an important integration center for human emotion. It is also known to exert a powerful influence over the hypothalamus to initiate endorphin release as previously described (see *Affect component).* Therefore, by way of complex projections of the brain stem reticular formation to the limbic cortex, the hypothalamus, and the pituitary, pain information gains access to the seat of human emotion and to the central nervous system and major endocrine glands. In this way, an emotional response becomes a direct part o fthe pain experience and a "visceral" reaction also spreads throughout the body. It may be these associations that link chronic pain with widespread systemic and visceral diseases of a "psychosomatic" nature, such as gastric hyperacidity and vascular headache.

The third component of pain, reaction, therefore includes an automatic visceral phase that can often be detected clinically by observing such signs as pulse and blood pressure elevations, pupillary dilation, sweating, and changes in saliva consistency. There is also a skeletal muscle reaction phase of the pain response that is more obvious (see *Reaction component).* Painful stimuli evoke reflex facial grimaces, turning of the head, and protective clenching of masticatory muscles. Skeletal muscle reactivity to chronic pain may also set up secondary pain foci, such as in the temporomandibular joint myofascial pain dysfunction syndrome.

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In summary, the experience of pain is the most common problem in maxillofacial neuropathology and should be viewed as more than a predictable specific sensation or a warning sign of disease. Rather, pain may become a disease syndrome in itself. Pathological factors that disturb peripheral and central inhibitory-excitatory balance must be detected, and the relative strengths of perception, emotion, and reaction components must be considered for each individual. Having accomplished this, one can best tailor control of the pain problem to offset each of the affected pain components.

<div align="center">**Anatomical Features**</div>

Gross sensory supply. Somatic sensation from the maxillofacial region is mainly carried by branches of the maxillary and mandibular divisions of the trigeminal nerve, which extensively branch before entering the skull. Because of this peripheral separation of branches, acute lesions (such as infection, compression, and neoplasia) frequently cause highly specific symptoms that aid in localizing the disease. However, neurological symptoms felt in peripheral tissues may also result from lesions as far central as the trigeminal ganglion and its sensory root because the peripheral fields of innervation are known to project precisely onto specific regions of the central trigeminal system. This concept is called *somatotopic organization* and may explain the mimic effect of peripheral disease and the highly localized "trigger points" that are part of many trigeminal neuralgia states. For example, certain forms of acute and highly localized trigeminal neuralgia may be caused by the irritation and destruction of specific fiber groups in the trigeminal ganglion overlying a carotid artery aneurysm. In spite of the occasional specificity of neurological symptoms, it is more typical for subacute or chronic maxillofacial lesions to produce poorly localized symptoms. They are often changeable or inappropriate for the stimulus and may even be *referred,* a situation in

which symptoms are felt in distant tissues and are unrelated to the true pathological site. These characteristics may be partially explained on the basis of anatomical features in the brain stem trigeminal complex. All sensory input from the maxillofacialregion has its primary synaptic termination in a laterally placed columnar region, the descending tract nuclei of the trigeminal system, which extends from the rostral pons to the third or fourth cervical spinal cord levels. It is typified by many fibers that synapse on one another and bring about considerable overlap and convergence of fibers on common secondary neurons. These complex interplays and convergences may help explain the diffuseness and also the referred nature of maxillofacial symptoms. For example, an irritative pathological condition from an alveolar osteitis in a third molar socket may cause pain anterior to the ear because the same irritated inferior alveolar brain stem synaptic regions are also shared by converging fibers of the auriculotemporal nerve. Similarly, pain accompanying angina pectoris may be referred to the supraclavicular and mandibular angle regions because incoming fibers of the cervical plexus are known to converge in the caudal portions of the descending trigeminal nucleus.

Microscopic sensory supply. The maxillofacial somatosensory tissues also have unique microanatomical features that help explain pathology in the region. On the basis of fiber size, trigeminal nerve branches contain the largest proportion of myelinated axons and the smallest proportion of unmyelinated axons in the entire somatic sensory system. This leads to the prediction that primary diseases affecting the myelin sheaths will show some predisposition for the trigeminal system. This does appear to be the case in multiple sclerosis, a demyelinating disease in which trigeminal neuralgia occurs in up to 5% of cases. A preponderance of myelinated fibers in peripheral nerves is also significant in those systemic disease states in which vascular pathology is a significant feature because the Schwann cells that are responsible for laying down and maintaining the myelin sheaths are known to be especially vulnerable to ischemia. This is the case in diabetes mellitus, in which trigeminal nerves may display *polyneuropathy,* defined as a symptomatic degeneration of many nerves.

The trigeminal ganglion cells themselves are known to be significantly larger on the average than the spinal ganglion cells. This may explain the high incidence of viral disorders in the trigeminal ganglion, such as trigeminal herpes zoster in which the inclusion bodies preferentially inhabit the larger cell bodies of the ganglion. A selective effect on a specific trigeminal cell population may be significant in certain congenital disorders such as the Riley-Day syndrome and in congenital insensitivity to pain. It has been suggested that these conditions result from an interference with the maturation of a particular segment of the sensory ganglion. In the case of the trigeminal ganglion, it seems that ganglion cells may develop from at least two separate sources, the neural crest and the epidermal placode. A teratological influence may act preferentially on one of these cell sources.

Autonomic supply. The *sympathetic* nerve supply to the maxillofacial region originates in the cervical spinal cord and, following synapse in the superior cervical chain ganglion, distributes to glands and smooth muscles by coursing in network fashion along the arteries of the head and neck. Lesions of the sympathetic fibers at any point distal to the cervical ganglion may produce signs and symptoms in the maxillofacial region. For example, an interruption of sympathetic functions in the orbit may result in Horner's syndrome, which is characterized by lid ptosis, pupillary constriction, and local anhidrosis. Horner's syndrome may result from lesions as diverse as a carotid sinus tumor, cellulitis in the infratemporal fossa, and retro-orbital edema after facial trauma.

The perivascular location of sympathetic nerve nets also makes then vulnerable reflex stimulation by inadvertent stimulation during local anesthetic administration. Mechanical

contact of the nerves with the anesthetic needle or intra-arteriolar deposit of solution with vasoconstrictor may trigger a severe spasm distal to the point of contact with the perivascular nerve fibers. This produces rapid pain and tissue blanching followed occasionally by an edema response. This phenomenon is perhaps most common with infraorbital nerve blocks in which stimulation of autonomic and also sensory fibers at the infraorbital foramen triggers a firing of autonomic branches by axon reflex.

Parasympathetic neurons to the maxillofacial region arise in brain stem cell columns designated as portions of the third, seventh, and ninth cranial nerves. Nerve processes course outward to synapse in the ciliary, sphenopalatine, otic, submandibular, and other smaller ganglia before distributing to smooth muscle, salivary glands, and lacrimal glands. These fibers are small in diameter and reach their final destinations by coursing with the larger nonautonomic branches of cranial nerves, primarily the trigeminal. For this reason, lesions of somatic nerves also affect parasympathetic function as in submandibular and sublingual gland dysfunction, which may result from the severance of the lingual nerve in the retromolar region. Because of the complexity of autonomic fiber distribution, there are occasiona errors in nerve regeneration patterns, which may result in bizarre reflex syndromes. For example, the *gustatory sweating syndrome* may follow interruption of the auriculotemporal nerve and its associated autonomic fibers caused by trauma in the mandibular fossa, parotid gland injury, or condylar fracture. In this syndrome a sweating and uncomfortable flushing of the face occurs over the distribution of the auriculotermporal nerve in response to a taste stimulus. This is probably a result of an inappropriate regrowth of the ninth nerve parasympathetic fibers along vacant sympathetic pathways that terminate in sweat glands rather than salivary acini.

Motor supply. Conscious control of skeletal muscle in the maxillofacialregion, which includes extraocular, masticatory, facial, lingual, and palatopharyngeal groups, originates in the cerebral cortex as "upper motor neurons". These nerves descend in both crossed and uncrossed tracts to many levels of the brain stem, where they terminate on secondary motor nuclear groups of "lower motor neurons". It is these latter neurons that send out peripheral extracranial processes to skeletal muscle and constitute the cranial nerves. The cranial nerves diverge from the brain at points widely separated from one another and range from the midbrain to the cervical spinal cord. It is for this reason that patients presenting with signs of multiple cranial motor nerve deficit probably are not suffering a lesion of the lower motor neuron. Rather, the diagnostician should suspect a lesion of upper motor neurons in the more rostral brain stem or cerebral tissues where the upper motor neuron tracts are more grouped. In addition, since all the lower motor cranial nerves except the fourth nerve are completely uncrossed in their courses to skeletal muscles, any lesion of the lower motor nerves produces a deficit in all the muscles supplied by that nerve. For example, lesions of the sevent nerve on one side will cause deficits in all ipsilateral facial muscles equally, whereas a lesion of the upper motor neurons that course to the motor nuclei of the seventh nerve causes a deficit primarily in the lower facial muscles, sparing upper facial muscle activity. This results from the patterns of crossing and uncrossing by the upper motor nerves that result in double innervation to some portions of the facial motor nucleus.

Most cranial motor nerves are anatomically well shielded from damage. For example, the trigeminal nerves to the masticatory muscles branch from the deeply located mandibular nerves and are rarely affected by peripheral pathological conditions. Therefore, if there are signs of masticatory neuromuscular deficit, the diagnostician should suspect a centrally located pathological condition, probably within the cranial cavity. Unfortunately the seventh nerve is not well protected and is especially vulnerable to lateral facial trauma. For example, seventh

nerve paresis may result from compression over the mastoid process and at the mandibular angle during general anesthesia using the face mask. The seventh nerve is also known to be especially prone to the effects of ischemia, perhaps because of its long course within the non-expanding facial bony canal. This may explain the occasional transitory facial palsy resulting from mandibular block anesthesia, in which intravascular injection has produced an ischemia in the posterior auricular and stylomastoid artery distributions. The facial nerve is also responsive to hypocalcemia as demonstrated by the use of the Chvostek test for systemic hypocalcemia, in which tapping over the nerve trunk elicits a tetany of facial muscles.

Histopathology

Although symptomatic maxillofacial neurological disease may be caused by a wide variety of lesions acting at many levels of the nervous system, certain basic histopathological processes help explain both the disease course and the associated clinical picture.

Reversible lesions. In the peripheral nerves, many common irritative lesions caused by mild trauma, chemical irritation, and necrotic infection may induce transient paresthesias as well as mild muscle paresis. A typical incident is the compression of inferior alveolar nerve branches during tooth extraction or the excessive blunt retraction of seventh nerve branches. The essential histopathological process in these cases involves tearing, hemorrhage, microinfarcts, and cellular infiltrates of the epineural and perineural sheaths without damage to endoneural tissue. Because these lesions involve no significant damage to the nerve axons or myelin sheaths themselves, they are reversible if the irritating factors are promptly removed.

Degeneration. There are two forms of degeneration seen in peripheral nerves that produce specific clinical symptomatology and that may result in irreversible neurological changes: segmental demyelination and wallerian degeneration. *Segmental demyelination* is a selective dissolution of the myelin sheath segments and is characterized by a slowing of conduction velocity as nerve impulses travel slowly along denuded axons. The clinical picture is frequently one of *polyneuropathy,* which is characterized by simultaneous involvement of many nerve branches, and the distribution of symptoms tends to be "patchy" and to cross natural neuroanatomical boundaries. The symptoms are typically those of paresthesia or dysesthesia in which certain of the sensibilities, such as deep pain, are retained, but others, such as fine tactile and pinprick sensitivity, are absent or delayed. Segmental demyelination is most often associated with vascular and connective tissue disorders that produce small infarcts in the ischemia-susceptible, myelinated nerve segments.

Wallerian degeneration is a disease process demonstrating disintegration of both the peripheral nerve fibers and myelin sheaths that spread distally from the point of first degeneration. Breakdown products are rapidly phagocytozesz, leaving prominent Schwann cell columns that once contained the nerve elements. Although these responses are best known as an invariable response to traumatic nerve section, they may be caused by any destructive lesion that attacks the peripheral nerve, including ischemia, inflammation, or tumor. In addition, wallerian degeneration of peripheral nerve processes may occur whenever the neuron cell bodies are diseased and unable to maintain their peripheral nerve cytoplasms. For example, necrosis of trigeminal ganglion cells resulting from invasion by herpes zoster virus is also accompanied by a wallerian degeneration of peripheral trigeminal nerves. A cerebrovascular accident in the pons adjacent to the motor nucleus of the seventh nerve may also result in degeneration of the peripheral nerve branches in the face. In some cases a peculiar form of wallerian degeneration may occur in which degeneration begins in the most

peripheral nerve tissues and progresses centrally from that point. This condition, known as "dying back" neuropathy, has been associated in the trigeminal system especially with metabolic intoxications caused by heavy metal poisoning, with isoniazid and penicillin therapy, and with conditions of malnutrition. In these cases anesthesia and paresthesia appear peripherally, and, as the disease progresses, more centrally located nerve branches become symptomatic.

These degenerative changes all take place within the first 48 hours after the primary lesion, and clinically the tissue distal to the point of degeneration become rapidly unreactive. In denervated zones the monosynaptic reflexes, such as the jaw-jerk reflex, disappear, sweating ceases, salivation decreases, and a zone of anesthesia develops immediately. Initially, the acute zone of anesthesia consists of a central *autonomous* zone that is absolutely free of sensation and a narrow surrounding *intermediate* zone of hypoesthesia, which results from the overlap of fibers of adjacent intact nerves. Within the first few days after denervation, the diameter of the autonomous zone becomes smaller because of the "sprouting" of new sensory terminations from adjacent normal nerves into the autonomous zone. A similar phenomenon may occur in denervated flaccid muscular regions as neuromuscular end plates sprout from adjacent intact nerves. This has the effect of partially reducing the size of the completely flaccid zone.

Neurotrophic effects. If tissues remain denervated for longer periods of time, certain clinical changes take place, called *neurotrophic effects.* In skeletal muscles the early spontaneous muscle spasms are followed by a flaccid paralysis with progressive atrophy and lack of muscle definition and tone. Denervated skin and mucosa may typically become cold, dry, and inelastic, with a greater susceptibility to injury and poor healing capacity. Keratinization is irregular, and skin surfaces may become caked and scaly, with a shiny cyanotic appearance. Joint structures may deteriorate, especially if subjected to intermittent stress. Although many of the neurotrophi tissue effects may be results of the interruption of efferent sympathetic fibers concerned with vasoconstriction, a missing neurohumoral nutritional factor has also been postulated to account for neurotrophic effects. Because of the potential for destructive neurotrophic effects, all efforts should be made to protect the denervated tissues from damage and to stimulate artificially the remaining intact structures until proper reinnervation can take place. Classic physical and occupational therapy techniques should be applied in these cases, such as lubrication and protection of surface tissues from trauma, manual stimulation of glandular tissues, warming and temperature control for assuring effective circulation, and electrical stimulation of intact motor units.

Normal regeneration. Peripheral nerve regeneration may begin within 24 hours if the cause of the original degeneration has been eliminated. The central nerve stump of the interrupted nerve sends out a swollen tangle of newly sprouted fibers, called a *growth cone,* that advances through the scar area of original degeneration seeking contact with the residual and now vacant Schwann cell tubes of the degenerative peripheral nerve. If the growth cone fibers reach the distal passages, they enter in random fashion, grow distally at a rate of approximately 1.5 cm daily, and finally make contact with terminal receptors and neuromuscular end plates. The thin fibers will then gradually thicken to approach their original diameters, and the investing Schwann cell will elaborate new myelin sheaths. Clinically the advance of the regenerating growth cone can be detected by observing *Tinel's sign,* in which a tapping on the growth cone or proximal stump will elicit paresthesias. As functional contacts are made, the autonomous zone of anesthesia gradually shrinks in size, first with the return of proprioception and a response to deep pressure pain stimulus that may be poorly localized and is itching, burning, or "bursting" in character; however, responses to

fine tactile and pinprick stimulation are lacking. Although these neurological imbalances usually disappear as the regenerated nerves continue to mature, a persistence of this sensory pattern may occur and is called *hyperpathia.*

Abnormal regeneration. Unfortunately many other factors may detract from the return of proper function in regenerated peripheral nerves. For example, the successful briding of the gap between the intact central stump and the distal Schwann cell passageways may be hampered by scar and foreign body barriers. When this happens the growth cone may continue to proliferate at the scarred junction in an aimless tumor of small fibers that constitute a *traumatic neuroma.* In other cases poorly myelinated tubular regions in the regenerated nerve, called *neuromas in continuity,* may result and resemble in many respects the discrete lesions of segmental demyelination. Because the nerve tissues of peripheral neuromas rarely mature and myelinate properly, their stimulation may result in painful bursts of intermittent pain and bizarre paresthesias. This phenomenon may be explained on the bsis of the setting up of *artificial synpases,* in which impulses in one demyelinated fiber may excite neighboring demyelinated fibers, resulting in an abnromal chain reaction to the original stimulus. The concept of the artificial synapse occurring in pathological peripheral nerve zones, as suggested by White and Sweet, may be a common explanation for many trigeminal neuralgias, such as those accompanying multiple sclerosis lesions and the paroxysmal shocking pain of tic douloureux. A similar explanation has been used to explain the deep burning pain of posttraumatic *causalgia,* which may be caused by the excitation of demyelinated sensory nerve segments by adjacent unmyelinated sympathetic fibers. In addition to neuroma formation, there are other potential accidents of regeneration. It is known that the relocation of growth cone fibers in the distal Schwann cell passages is largely a nonspecific selection process, and the identical matching of new regenerating fibers with their former tissue receptors may not occur. Fibers may come to innervate the wrong tissues. When this occurs in reinnervated skeletal muscle and glands, motor control is inappropriate. Little is known about similar accidents in sensory tissues. It is also known that regenerated fibers rarely attain their original diameters, and the distances between nodes of Ranvier are shorter in regenerated nerves. These two factors may lead to reduced nerve conduction velocities as well as disproportionately high numbers of smaller fibers. According to the gate control theories of pain and sensory modulation outlined previously, such imbalance in the afferent fiber diameters could lead to sensory abnormality such as hyperpathia.

In addition to imbalances that may be induced by histolpathological conditions in the peripheral nerve fibers, significant imbalances may also be caused by selective effects on the nerve cell bodies themselves. For example, there is considerable evidence that trigeminal ganglion cell bodies may be selectively lost as a result of many life and disease processes. Neuron necrosis occurs more readily in immature cells, and possibly in larger cells, from such diverse causes as trauma, metabolic disease, and viral infection. In those circumstances in which the neuron cell bodies are unable to survive, there is not only a wallerian degeneration of peripheral nerve but also a disintegration of the central nerve processes. This has the effect of severing functional synaptic connection with the secondary transmission, reflex, and integration centers in the central nervous system. In sensory systems the loss of peripheral fibers and synaptic contacts that normally arrive at the primary synaptic regions is called *deafferentation,* and it is now known that trigeminal deafferentation may induce both morphological and physiological changes int he nuclei of the descending trigeminal tract. Deafferented brain stem regions take on bizarre and stereotyped electrical characteristics that have been called *epileptogenic foci* because they resemble the EEG patterns that typically inititate seizure activity. It has also been postulated that these epileptogenic firing patterns may represent the physiological change responsible for paroxysmal and atypical neuralgia

states. Deafferentation effects may eventually explain the cause of such poorly understood conditions as trigeminal neuralgia, postherpetic neuralgia, and phantom limb pain.

Neuralgias

Neuralgia may be defined as paroxysmal, intense intermittent pain that is usually confined to specific nerve branches of the head and neck. It is the modern view that paroxysmal, bursting maxillofacial pains may have a common histopathology, which is a breakdown in the insulating mechanisms between axons without destroying them. This primary condition may occur in peripheral nerve branches, in the sensory ganglion tissues, or in the posterior roots. There is good evidence that these peripheral lesions may cause pain by creating afferent imbalances and setting up abnormal pools of secondary central neurons in the trigeminal descending tract nuclei, possibly the epileptogenic foci type.

Idiopathic trigeminal neuralgia. The most dramatic and well-known neuralgia is *tic douloureux,* which displays the classic and diagnostic features of paroxysmal pain that is (1) extreme, "stabbing", or "shocking", lasting seconds to minutes, (2) rapidly provokable by gentle stimulation over surface "trigger zones", (3) confined to the distributions of trigeminal nerve branches, (4) unilateral and does not cross the midline for any given paroxysm, and (5) without objective sensory or motor loss in the affected region. In some cases atypical neuralgia features may be superimposed on this clinical picture, including an unprovoked burning or aching that persists between the paroxysms, pain that may radiate into the neck and posterior scalp, and the occurrence of mild hypesthesias. Idiopathic trigeminal neuralgia occurs most frequently in the sixth decade of life and more frequently in women (over 58%), has a predilection for the right side (over 60%), and may be cyclic or seasonal with more than 50% of patients experiencing early remissions of greater than 6 months before return of active pain. Pain occurs most often in the maxillary peroral tissues, which are grossly and microscopically normal, and the pain may be triggered by gentle touching or by facial movements during talking or mastication. Although the pain of the trigger zones and pain fiber distributions often mimic pain of odontogenic or sinus origin and cause patients to seek indiscriminate dental extractions, there have been no proved correlations between dental or antral sepsis and the occurrence of tic douloureux.

The histopathological lesion of idiopathic trigeminal neuralgia has now been established as a hypermyelination, segmental demyelination, and microneuroma formation that is localized in ventral portions of the trigeminal ganglion and its adjacent posterior root fibers. It has been theorized that such a lesion would act pathophysiologically as an artificial synapse by setting up abnormal volleys throughout the tangle of demyelinated fibers. Also, according to the gate control theory of the pain mechanism, the selective degeneration of the largest myelinated fibers in these lesions could have the effect of lowering pain inhibition of smaller fibers in the brain stem trigeminal nuclei.

The cause of this disease process is unknown. Vascular factors such as transient ischemia and autoimmune hypersensitivity responses have been proposed as causes of the demyelination. Mechanical factors have also been postulated, such as the action of aneurysms of the intrapetrous portion of the internal carotid artery that may erode through the floor of the intracranial fossa to exert a pulsatile irritation on the ventral side of the trigeminal ganglion.

More recently, an anomaly of the superior cerebellar artery has been described that has been shown to lie in contact with the sensory root of the trigeminal nerve. This anomaly has

been implicated as a cause of demyelinating pathology. Surgical elevation of the vessels away from the sensory rooty has been highly successful in relieving paroxysmal pain in cases of idiopathic trigeminal neuralgia.

The diagnosis of this syndrome is based primarily on recognizing the clinical features. Diagnostic local anesthetic blocks applied at the point of triggering should eliminate all paroxysms of pain and help differentiate atypical neuralgia and other forms of trigeminal neuralgia or neuritis conditions Treatment of this condition is both medical and surgical. However, analgesics, sedatives, and the formerly advocated vitamin B_{12} injections have no significant therapeutic effect on tic douloureux pains. The best medical results have been gained with antiepileptic agents such as phenytoin (Dilantin), which decreases the pain symptoms approximately 50% of the time, and more recently carbamazepine (Tegretol), which affords significant or total pain relief in a high percentage of cases, as much as 100% in some of the series reported. The effectiveness of antiepileptic agents in the control of paroxysmal pain lends support to the concept of the epileptogenic focus as a factor in the pathophysiology of paroxysmal pain. Reports on the use of carbamazepine have reversed early concerns about its toxicity, although its effectiveness for long-term pain control is not yet known.

Surgical approaches for maxillofacial neuralgias are varied and will be discussed in greater detail in a later section. They consist of (1) temporizing procedures such as alcohol injection and peripheral neurectomy for eliminating the peripheral trigger effects, (2) decompression and complete resection of ganglion posterior root fibers, and afferent trigeminal tracts, and (3) interruption of central trigeminal ascending pathways. In addition, neurophysiological stimulation techniques based on the gate control theory of the pain mechanism appear to hold promise for the future control of tic douloureux.

Vagoglossopharyngeal neuralgia. Formerly called glossopharyngeal neuralgia, the syndrome of *vagoglossopharyngeal neuralgia* occurs less than one-eightieth as often as idiopathic trigeminal neuralgia and is an affliction of sensory, autonomic, and motor fibers of the ninth and tenth cranial nerves. The onset is usually in the fourth decade, with there being no apparent predilection for either sex. The left side is involved more frequently than the right, and both bilateral pain or combinations with idiopathic trigeminal neuralgia are rare. The pains are felt in the base of the tongue, the adjacent tonsillar pillars, and occasionally the soft palate and external auditory canal. The paroxysms may be of lower intensity than trigeminal tic pain, with intermittent refractory periods. Pain triggering is most often caused by swallowing or surface stimulation in the tongue or pharynx, and occasionally pain will follow taste stimulation by spicy or bitter foods. As part of the paroxysmal attack there is also excessive salivation, lacrimation, mild vertigo, and involuntary movements of the pharynx and larynx, which may result in severe coughing and vomiting. Syncope may occur as well as a progressive hypotension and bradycardia; in extreme cases ECG recordings have demonstrated asystoles of over 1 minute, and there are records of associated cardiac arrest.

The histopathological finding is similar to that described for idiopathic trigeminal neuralgia, and the complex clinical features can most easily be explained on the basis of an indiscriminant artificial synapse lesion of mixed ninth and tenth cranial nerve components. The pain fiber distribution to the tongue and pharynx and the taste fibers from the posterior third of the tongue are known to be components of the ninth nerve. Irritation of the greater petrosal nerve branches explains the increased function of the parotid gland, and the pharyngeal-laryngeal motor reflexes suggest involvement of the ninth and tenth nerve pharyngeal plexuses. Most significantly, the extreme cardiovascular effects of this syndrome reflect an affliction of the nerve of Hering, which is responsible for initiating carotid sinus

reflex activity.

Possible causes include posterior root demyelination from the pressure of intracranial aneurysm, and correlations have also been made with preceding paratonsillar infection. Neuralgia association has also been demonstrated with an elongated ossified stylohyoid ligament, causing *Eagle's syndrome,* in which chronic and functional pressures may be exerted on the vagoglossopharyngeal nerve complex in the region of the foramen spinosum. The firm and enlarged stylohyoid ligament in these cases can be seen radiographically, may be palpable in the lateral soft palate or anterior tonsillar pillar, and will often precipitate the paroxysmal pain if the head is turned toward the affected side. In one retrospective study an ossified stylohyoid ligament was found in 70% of cases of vagoglossopharyngeal neuralgia, and intraoral osteotomy of the styloid process has brought about a cure in 11 of 12 patients reported in the literature.

Drug therapy similar to that used to manage trigeminal neuralgia has also been effective in this condition. Surgical peripheral neurectomy procedures have not been effective for relief in these cases, and most attention has focused on intracranial rhizotomies of the ninth and tenth nerves' posterior roots as well as the tractotomy procedures identical to those used in the control of trigeminal neuralgia.

Intermedius (geniculate) neuralgia. A rare paroxysmal pain, described as a "red-hot poker" felt deep within the external auditory canal, the auricle, and occasionally the soft palate, is attributed to involvement of the sensory or intermedius portions of the seventh cranial nerve. This unilateral condition, also known as the *Ramsay-Hunt syndrome,* has a strong predilection for women, no preference for right or left side, and has its usual onset in young to middle adulthood. The pains are typically longer in duration that the paroxysms of idiopathic trigeminal neuralgia, and they are often unprovokable, although trigger zones have been identified in the pinna of the ear. Other unusual clinical features may include excessive salivation and nasal secretions, tinnitus, mild vertigo, and a bitter taste. Diagnosis may be confirmed by stimulating the tympanic plexus and chorda tympani nerves in the middle ear of the awake patient. After the specific involved nerve branches have been identified by stimulation, the pain may be relieved by neurectomy of these branches.

Periodic migrainous neuralgia. Periodic migrainous neuralgia, known also as "cluster headache" and "histamine cephalgia", also encompasses the syndromes of ciliary neuralgia, vidian neuralgia, and sphenopalatine (Sluder's) neuralgia. Its clinical features are intermediate between trigeminal neuralgia and migraine headache in that it combines painful paroxysms with prominent facial autonomic disorder. The pain is burning or aching and usually begins deep within the midface behind or beneath the eye and then "migrates" unilaterally to the forehead, temple, and then to the lower face. The attacks increase in severity for 5 to 20 minutes before subsiding and occur in groups or "clusters" over a 2- to 3-month interval, followed by pain-free remission intervals of weeks to years. The overall incidence is lower than for classic migraine and has no known hereditary basis. Although the role of psychogenic factors is disputed, there is a common association with both emotional stress and compulsive personalities. There is no known trigger point or point of provocation, although alcoholic intake and subcutaneous injections of histamine will precipitate attacks. Along with pain attacks, there is a rapid injection of the conjunctival and nasal mucosal vessels, heavy lacrimation and salivation, and often the development of a transient Horner's sign of ptosis and miosis. The temporal vessels become dilated and tender, the eyelids and oral mucosa may swell, and the face may flush a brilliant red.

It is generally agreed that this disorder is a result of an arterial dilation as in classic migraine headache, but in periodic migrainous neuralgia the point of neuropathology is somewhere along the course of the greater superficial petrosal nerve, either intracranially, in its tympanic and petrous course, in the vidian canal, or in the sphenopalatine ganglion itself. The pain and autonomic signs typical of this syndrome appear to be explained on the basis of abnormal excessive discharges in this mixed parasympathetic nerve.

Diagnosis and pain relief can be aided by local anesthetic block of the sphenopalatine ganglion through the greater palatine foramen. Although petrosal neurectomy may be indicated in certain refractory cases, good relief of neuralgia attacks is usually gained by using either ergotamine tartarate or methysergide (Sansert, Deseril), which appears to act through vasoconstriction and serotonin antagonism. Carbamazepine appears to be of no benefit.

Multiple sclerosis neuralgia. A paroxysmal maxillofacial neuralgia, in which the pain is indistinguishable from the pain of idiopathic trigeminal neuralgia, may occur as a feature of multiple sclerosis. The pains are precisely confined, have a shocking or stabbing character, and may be triggered by tactile stimulation. However, in contrast to idiopathic trigeminal neuralgia, multiple sclerosis neuralgia more often involves multiple trigeminal divisions and more often becomes bilateral. Although paroxysms may be the initial symptom of the disease process, they usually are associated with the other progressive and various disorders that characterize this disease. In some cases the paroxysmal pains have followed an initial transient facial numbness and loss of taste on one side; late in the disease process the facial pain may become constant as secondary central pain tracts become diseased.

Multiple sclerosis is a degenerative disease prevalent in norther climates, and its cause is unknown. It runs a varied episodic course over a 10- to 30-year period and involves progressive widespread sensory and motor disability. The acute onset is usually in the third decade and appears typically as a weakness of the lower limbs, disturbed vision, and objective sensory loss in about half the cases. These early signs are typically followed by long remissions. The histopathological lesion in multiple sclerosis is the sclerotic "plaque", a discrete focus of myelin loss with maintenance of axon segments and glial proliferation. The plaques may be seen in widely divergent brain tissues, but the trigeminal neuralgia paroxysms seem to be associated specifically with plaques that span the junction of the posterior trigeminal root entry to the pons.

The differential diagnosis of this disorder depends on identifying the features of a multiple nervous system disorder. There is no known cure for the basic disease process, but the facial pain can be effectively controlled by the same surgical techniques used for idiopathic trigeminal neuralgia. Carbamazepine relieves the pain of multiple sclerosis trigeminal neuralgia in approximately 80% of cases.

Posttraumatic Sensory Disorders

Trauma to the peripheral nerves of the maxillofacial region is a common consequence of daily life, whether the damage is caused by accident or iatrogenic sources. Because of unique features of nerve tissue degeneration and regeneration, a wide range of posttraumatic pathological conditions may result. The clinical problems may range from barely perceptible paresthesias caused by small neuromas-in-continuity to a profound neuralgia with phantom characteristics that reflects central nervous system pathophysiology.

Posttraumatic trigeminal neuropathy. Maxillofacial trauma is now recognized as a common factor in facial pain syndromes that in the past may have fallen in the catchall "atypical facial pain" class. The type of trauma that precedes this syndrome usually involves direct tearing or intense compression of peripheral sensory nerves; repetitious trauma is of particular importance even with moderate force. Specific examples that have been cited are fractures of the maxillofacial bones such as basilar skull, supraorbital, and zygomatic fractures involving the infraorbital nerves and displaced mandibular fractures in which the traumatized nerves are associated with disrupted bony canals. In many cases of fractures that precede neuropathy, the displaced bony segments are not firmly fixed, permitting continued mobility and repeated injury to the regenerating sensory nerves. The development of neuropathy may be associated with indirect or iatrogenic trauma such as maxillary and frontal sinus-lining ablation, chronic denture flange irritation, cured infections of the masticator spaces, alveolar osteitis, and, most common of all, damage to the inferior alveolar nerve during third molar extraction. Although an acute onset of the pain syndrome at the time of injury has been reported, there is more often an initial period of anesthesia over the traumatized nerve zone, followed by a gradual onset of symptoms 2 months to 15 years after injury. Any of the cranial or sensory nerve distributions may be involved, although the third division of the trigeminal nerve is most commonly affected. The accompanying pain is aching, burning, boring, or pulling in character, is poorly localized along anatomical lines, and spreads out from the zone of original nerve injury. It is usually not provokable or triggered but is sustained and builds gradually, although in some cases bursts of paroxysmal pain may be superimposed. The neurological examination of the pain areas is significant in that there are often reduced fine tactile and pin-prick sensitivities as well as paresis and neurotrophic effects, such as masticator muscle atrophy.

The spreading, nonprovokable nature of the pain and its delayed onset suggests a central nervous system focus of hyperexcitability, similar perhaps to the epileptogenic focus previously described. However, it has been emphasized that more peripheral sites of pathology such as the traumatic neuroma may also result in abnormal sensation. Therefore all efforts should be made to identify any sources of peripheral nerve irritation such as bone fragments displaced into neurovascular canals, occluded nerve foramina, or foreign bodies such as transosseous wires impinging on nerve bundles.

Although peripheral neurectomy procedures may benefit some cases, the sustained burning quality in this syndrome is often refractive even to retrogasserian rhizotomy. Medical treatment with carbamazepine has been effective in the more paroxysmal forms of posttraumatic pain but less successful in relieving the overall neuropathy syndrome.

Causalgia. As a syndrome, causalgia means literally "burning pain" and occurs in the appendages as a result of penetrating missile wounds of mixed peripheral nerves. Pain begins after a postinjury delay of at least 2 weeks with a deep aching or burning that spreads out from the injured zone and beyond natural nerve boundaries. It is neither paroxysmal nor triggered, although attacks are brought on by mild pain or touch stimulation of the region, by drying of tissues, or by environmental stimuli such as loud noises. The affected tissues sweat excessively, and show color and degenerative trophic changes. All the symptoms are intensified by emotional stress. There is no apparent sexual or racial predilection, and the syndrome is most often seen in vigorous middle-aged people.

Major causalgia has been described only rarely for the maxillofacial region, yet minor causalgia-like states may account for some of the rather common complaints of persistent, focal posttraumatic burning of the gingiva, palate, tongue, and lips. Perhaps most common

are complaints of persistent burning in postextraction sites, which occurs along with subtle vasomotor and trophic changes that are made more intense by emotional stress. The specific mechanism of causalgia pain may be the artificial synapsing of efferent sympathetic fibers with somatic sensory fibers within neuromas that have formed at the site of original nerve injury. The initiation of sympathetic impulses for this bizarre nerve firing pattern probably takes place in the limbic-hypothalamic axis that normally regulates general sympathetic activity. This interaction may explain the influence of emotional factors in heightening the pain. Maxillofacial causalgia pathological lesions are likely to be located at sites of sympathetic and somatic sensory nerve intermingling in nonexpansile bony canals such as the supraorbital, trochlear, and infraorbital and mandibular canals, where mixed nerve bundles are found.

The differential diagnosis of causalgia-like lesions in the maxillofacial region includes psychalgia or hysteria reactions, persistent local pathological conditions, and the more diffuse tissue changes such as surface hypersensitivities. Selective blocking of tissues with vasoconstrictor alone may aid in the diagnosis because true causalgia attacks are relieved by sympathetic blockade. Major causalgia has been treated effectively with sympathectomy, and although minor causalgias have been occasionally cured by identifying and excising neuromas, the best treatment for minor maxillofacial causalgia-like conditions has not been determined.

Phantom facial pain. Patients who have undergone excision of a body part often experience a sense of awareness of the missing part called the *phantom phenomenon.* In cases of limb amputation, in which the phantom phenomenon is best known, the phantoms are painful in approximately 30% of cases and persist in 5% to 10%. Paroxysms are of approximately 10 minutes' duration and are described as stabbing, with extreme itching or deep burning and pressure of the missing part. They may be triggered by tactile stimulation and usually are relieved by local anesthetic block of the peripheral nerve stumps. Although little is known of the incidence and pathogenesis of phantom pain in the maxillofacial region, it should be given consideration in the differential diagnosis of facial pain and not dismissed as evidence of psychological disorder. Complaints can be anticipated from patients who have undergone radical excisions as in orbital-antral exenterations, glossectomy, and mandibulectomy operations for cancer control. Complaints of toothache and of "phantom teeth" in dental extraction sites are not unusual, especially when the teeth have been chronically symptomatic before removal.

Although neuromas occurring at the regenerated nerve stump surface may contribute to this syndrome, it appears that the primary site of pain mechanism is in the brain stem. After limb amputation about half the associated neurons die, and the regenerated fibers of the stump are usually small, poorly myelinated, and slow conducting. Stimulation of these stump tissues therefore may have the effect of activating an imbalanced gate control mechanism in the brain stem and cause inappropriate sensory phenomena such as phantom pain. Palpation and diagnostic block may reveal the presence of a contributing neuroma, and reamputation of nerves at more proximal levels may be successful in these cases. In more severe cases the pain can be eliminated by anterolateral cordotomy, although the awareness of phantom tissues is seldom eliminated. Carbamazepine therapy results in varying degrees of succes, and supportive care and reassurance is often adequate in less severe cases because phantom pain seems to diminish with time.

Anesthesia dolorosa. When the afferent fibers of the cranial nerve sensory roots are divided central to the ganglion in rhizotomy procedures, there is a profound and permanent numbness of the denervated tissues. However, the majority of these patients experience some

type of paresthesia within the anesthetic zone, and, for an unfortunate few (3% to 15%), these sensations become intolerably painful. The pain, called *anesthesia dolorsa,* has a gradual onset in the weeks to months after rhizotomy, is constant, burning, pressuring, "crawling or grinding" in sensation, occurs most often in the ophthalmic regions, and is not provokable. The likelihood of anesthesia dolorosa occurring for a patient can be accurately predicted by observing effects of long-term trial denervations with peripheral neurectomy or alcohol blockade of nerves. The site of the disorder is probably in the central nervous system, possibly in the cortex, and medical management is of questionable value. The most effective treatment to date seems to be bilateral frontal lobe leukotomy and lesions of the trigeminothalamic tracts.

Infectious Disorders

Few primary infections involve the nerves of the maxillofacial region. Leprosy is the only known direct infection of peripheral fibers, but herpes infections of the cranial nerve ganglia are proved viral infections, and rare fungus infections of the trigeminal ganglion are also known. However, numerous infections involve the central nervous tissues secondarily because they cause peripheral symptoms and signs and thus are of concern, since these infections may arise in the maxillofacial tissues and spread intracranially.

Herpes zoster (postherpetic trigeminal neuralgia). *Herpes zoster,* commonly known as shingles, may occur in the sensory nerve distributions of the trigeminal, seventh, ninth, and tenth cranial nerves. Herpes zoster is associated with sensory abnormality in its acute disease phase but, more significantly, may result ina severe postherpetic facial neuralgia that is difficult to treat. The acute disease involves the trigeminal nerve in 18% of cases, which is second to the thorax in disease frequency. Outbreaks are most commonly seen in the ophthalmic division. The acute eruptions are discrete painful vesicles with erythematous bases that correspond in distribution to the particular nerve branches, which may involve skin, mucosa, and corneal surfaces. It appears mainly in older people, although a life-threatening neonatal variety is known. It is probably contagious and appears to have peak seasons. The disease may arise after a systemic toxicity or metabolic disorder but most often follows trauma to peripheral sensory nerve branches. In the maxillofacial region, for example, herpes zoster may follow inferior alveolar nerve damage from dental extraction. Herpes zoster is the only definite viral infection of the peripheral nervous system and may be caused by a reactivation of varicella viruses that are latent in the tissues with a specific infection of the larger neuron cell bodies of sensory ganglia. This selective effect in some cases has resulted in a relative loss of the larger trigeminal nerve fibers and a shifting of the postherpetic fiber spectrum toward the small elements. Based on gate control concepts of pain mechanism, this phenomenon may explain the occasional complication of postherpetic neuralgia. Clinically this condition is a constant burning or aching often associated with a deep aggravating anesthesia within the region of the original herpetic outbreak. In addition, severe paroxysmal stabbing pains may be elicited by light touch stimulations. Unfortunately postherpetic neuralgia has been extremely refractive to surgical treatment, and most efforts toward pain management with carbamazepine have been unsuccessulful.

Infectious meningitis. It is well established that foci of infection in the maxillofacial tissues may spread into the intracranial cavity. When the process is acute, it may lead to death within 10 days, and some experts feel that chronic forms of central nervous system infection from maxillofacial foci may be responsible for bizarre forms of epilepsy and also contribute to multiple sclerosis. Exacerbations of the infection, causing inflamation and scarring of the cranial nerve rootlets that pass through the meninges, may cause focal neurological disease

such as extraocular muscle paresis and may result in transient trigeminal and facial nerve (Bell's) palsy. Clinical signs of acute meningitis are progressive headache with nuchal rigidity and neck flexion and a decreasing level of consciousness with memory deficits. Later in the course, focal signs appear such as hemiparesis, loss of extraocular muscle tone, vertigo, papilledema, and facial pain. Lumbar puncture will usually reveal a leukocytosis in the cerebrospinal fluid, and angiograms and brain scan may reveal an expansile abscess mass. A direct culture of identical organisms from both the brain abscess and the maxillofacial infection will confirm the diagnosis.

The main pathways of infection are venous routes and direct extension, although perineural spread and lymphatic and arterial bacteremias may also infect the brain. The most common route of venous spread is from anterior maxillary foci along the facial angular veins to anastomoses with the inferior and superior ophthalmic veins. Because the facial veins lack valves, these infectious emboli may reach the cavernous sinus along this route. A similar spread may occur from infratemporal space infections that involve the pterygoid venous plexus and that readily communicate with the intracranial venous sinuses adjacent to the temporal lobes. Temporal lobe abscesses may also result from a direct erosion of the temporal bone by cellulitis in the infratemporal and temporal fossae. Direct pulsations of organisms into the subarachnoid space occur almost routinely in basilar skull fractures. Likely sites of seeding in these cases afre the frontal lobes superior to the cribriform plates and the temporal lobes adjacent to the middle ear cavity. The most common maxillofacial foci for venous spread and direct invasion are the maxillary teeth, pericoronitis in the mandibular third molars, the palatine tonsils, and contaminated needles introduced into the pterygomandibular space. Prevention of intracranial spread depends on establishing early dependent drainage, wound culture, and attaining high blood levels of antibiotics, preferably penicillin. However, craniotomy and direct abscess drainage may become necessary to avoid brain necrosis and high intracranial pressures.

Leprous neuropathy. Leprosy is the most common neuropathy in the world and is caused by a communicable infection by the acid-fast bacillus, *Mycobacterium leprae,* which enters at skin or mucous membrane surfaces and invades peripheral nerve branches with the capacity to ascend as high as the sensory ganglia. It has a special affinity for the cooler body parts, especially the maxillofacial region where disfiguring lesions appear, including subcutaneous nerve nodules on the eyebrows, nares, and cheeks and cauliflower ears and depigmentation of the skin. A patchy paresis of the sevent cranial nerve branches often leads to drooping of the eyelid and oral commissures, and spotty anesthetized areas over the trigeminal nerve division make the patient vulnerable to trauma.

The essential histopathological lesion is the leprous nodule, a granuloma of bacterial colonies, epithelioid and fibroblastic cells, and plasma cells. The nodule surrounds an area of demyelination and eventually casues wallerian degeneration of the nerves. Leprous nodules can be differentiated from similarly appearing tuberculosis and actinomycosis by analysis of nasal scrapings or nerve biopsy. Modern chemotherapy with sulfones prevents further nerve destruction by these organisms.

Diphtheric neuropathy. Diphtheria is an acute neuropathy caused by the exotoxin of *Corynebacterium diphtheriae,* which spreads along the peripheral perineural channels to produce a segmental demyelination. Most of the cranial nerves may be affected, producing facial palsies, extraocular muscle paresis, nerve deafness, and hypoglossal paresis. In over 75% of cases of diphtheric neuropathy there is an unexplained predilection for the motor fibers of the vagus that supply the skeletal muscles of the palate, pharynx, and larynx.

Susceptible children are infected by droplets in the palatine tonsil and nasopharyngeal tissues, producingf a gray pseudomembrane in these areas. Early symptoms include hypernasality, nasal regurgitation associated with a deviated uvula, and absence of palatal elevation in the gag reflexes. The disease may progress to include a myocarditis and may be complicated by pneumonia. Active immunization is routinely induced in the first year of life, but if the acute disease is encountered, diphtheria antitoxin should be administered. After recovery there may be a postdiphtheric cranial nerve palsy in the palate that requires speech aid.

Neurosyphilis and tabes dorsalis. Neurosyphilis occurs in 10% of persons infected by *Treponema pallidum,* and the organism may reach the nervous system in the primary stage before the development of any cutaneous lesions. In the secondary stage of the disease process, headaches are common as are ocular palsies. Occasionally a polyneuritis occurs that may involve cranial nerves. It is the tertiary form of neurosyphilis, however, that may lead to the most serious maxillofacial neurological dysfunction. The essential pathological lesion is a vascular inflammation, which results in a necrotic and granulomatous response of the meninges called *gumma.* Gummatous lesions of the meninges may secondarily affect the cranial roots and typically cause palsy of the third, fourth, and sixth nerves as well as partial or total paralysis of the seventh nerve. The trigeminal nerve may also be involved, causing spontaneous paroxysmal pain that is poorly localized and spreading. The diagnosis of tertiary syphilis depends on recognizing the widespread neurological symptoms, and obtaining accurate history and a positive VDRL test.

A particular form of chronic tertiary neurosyphilis, called *tabes dorsalis,* involves a selective degeneration and demyelination of the largest fibers of the dorsal roots and cranial nerve afferent tracts that project through the dorsal columns of the spinal cord and brain stem. The pain of tabes dorsalis is described as "lightning" with a burning or "tearing of the flesh" character that persists for seconds and then shifts to another location. Although tabetic pain occurs most commonly in the extremities, the trigeminal and glossopharyngeal nerves may be involved. Examination may reveal loss of deep pain and proprioceptive sensitivity, with occasional loss of taste and smell. The treatment of tabetic pain is symptomatic.

Maxillofacial Neuropathies of Systemic Origin

A broad range of common systemic disease states including metabolic and nutritional disorders, intoxication, and connective tissue diseases may affect neurological function in the maxillofacial region. These manifestations are typically less predictable in location, intensity, and course than primary neuropathological conditions, and they often simultaneously involve motor, sensory, and autonomic functions. Neuropathies may result from segmental demyelination, wallerian degeneration, or dying-back processes, and clinical features may be confusing because nerve degeneration and regeneration often exist side by side.

Connective tissue disease neuropathies. Trigeminal neuropathies have increasingly been linked with the major collagen disorders, including lupus erythematosus, Sjögren's syndrome, rheumatoid arthritis, scleroderma, polyarteritis nodosa, and diabetes mellitus. The grouping of these diseases is not coincidental; their association with trigeminal neuropathy is based on a rather common mode of pathogenesis. A randomized vasculitis occurs in the nutrient vessels of the peripheral nerves, causing microinfarcts and ischemia to the nerve segments. A segmental demyelination develops rapidly, with initial preservation of the nerve fibers. The haphazard positioning of pathological areas in more peripheral tissues and the selectivity for myelinated portions of the nerves help explain some clinical features. Patchy regions of paresthesia occur, often described as "creeping" or "tightening" numbness with

infrequent shooting pains. The disturbances are often multifocal and involve motor, sensory, and autonomic nerves regionally and may change position gradually as nerve regeneration occurs in some areas. The trigeminal neuropathies are often preceded by weeks or years by the appearance of Raynaud's phenomenon, a condition in which the fingers and toes respond to cold and emotional stress with pallor, cyanosis, and trophic skin changes caused by vasospasms. The combination of Raynaud's phenomenon and trigeminal neuropathy is significant because of the strong correlation with the development of refractory postextraction dry socket.

Neurological examination of the collagen trigeminal neuropathies reveals reduced vibratory, pinprick, temperature, and light touch sensibilities and an unpleasant hyperpathia that is brought on by deep pressure stimulation. Clinical tests and nerve conduction studies may reveal that deep tendon reflexes such as the blink reflex to glabellar tapping and the jaw-jerk reflex are depressed. Vasomotor deficits may be observed in some cases.

The disease state most commonly associated with neuropathies of all types is diabeter mellitus. Trigeminal diabetic neuropathies tend to be scattered sensory and motor dysesthesias, and, as in the pure collagen disorders, the neurological disorders may be only slightly improved by controlling the underlying disease processes. Systemic steroid therapy may check the progression of trigeminal neuropathies, but circumstantial evidence exists that steroids themselves may act as neurotoxic agents.

Toxic and nutritional neuropathies. In conditions of severe nutritional deprivation and in reactions to foreign substances, the nerve cell metabolism may be so disturbed that it can no longer maintain its peripheral processes, and a dying-back wallerian degeneration results. These metabolic neuropathies are characterized by numbness and paresthesias that begin in the more distal portions of nerves and branches of nerves most acutely affecting vibratory and position senses.

The nutritional neuropathies usually occur in the appendages but may involve the maxillofacial region. They are usually related to mixed deficiencies of the vitamin B complex. Deficiencies of vitamin B_1 (thiamin) cause beriberi and may result in nerve deafness, laryngeal paralysis, and a patchy trigeminal paresthesia or anesthesia. This syndrome may also be seen as part of Wernicke's encephalopathy, which is caused by thiamin deficiency in chronic alcoholism. Pellagra, a result of niacin (vitamin B_2) deficiency, is characterized by stomatitis, glossitis, erythematous dermatitis, diarrhea, and a randomized trigeminal sensory neuropathy. Pernicious anemia neuropathy is caused by incomplete absorption of vitamin B_{12} by the small intestine and may cause burning paresthesias in the orofacial region.

Heavy metal intoxications are known to specifically induce necrosis in sensory ganglia, including the trigeminal and the geniculate. The metals that have been most frequently associated with maxillofacial neuropathies have been mercury, lead, cadmiummm, bismuth, and arsenic.

Among the nonmetals that are known to intoxicate the nervous system, a few have specific preference for the trigeminal system. Trichloroethylene, formerly used extensively in obstetrical analgesia, produces a trigeminal neuropathy by a selective necrosis of ganglion cells. Triorthocresylphosphate ("Jamaica Ginger"), an occasional contaminant of bootleg liquor, produces polyneuropathy in many body regions including the face. A transient trigeminal neuropathy with peroral paresthesias has also been linked to acute ethyl alcohol intoxication.

Finally, trigeminal sensory neuropathies have been linked with a number of well-known therapeutic drugs, including penicillin, cortisone, Stilbamidine, isoniazid, and nitrofurantoin.

Headaches

More than 90% of the population have experienced maxillofacial headache, and most headaches result from combinations of both organic and psychic factors that act cyclically and potentiate one another. The most frequent kinds of major headaches are vascular in origin, resulting from excessive vasodilation of extracranial or intracranial vessels as in the migraine headaches, or direct vessel wall pathology such as temporal arteritis. Most common mild headaches are "tension headaches", caused by abnormal skeletal muscle contractions. A most common tension headache is the temporomandibular joint-related syndrome of myofascial pain dysfunction.

Coronary artery disease may cause referred pain headaches in the lower face and neck, and hypertensive disease may induce chronic, diffuse headaches. Severe cranial trauma may produce both acute and chronic posttraumatic headaches, and distinctive head pains are known to follow both lumbar puncture and spinal anesthesia. Nonlocalizing headaches may signal progressive intracranial disease such as infection or tumor. Finally, radiating maxillofacial headaches are a frequent by-product of acute neuritis in nasal, paranasal, otological, ocular, and dental tissues.

Migraine. True migraine headaches are "sickening", often debilitating pains that throb and ache unilaterally in the temporal region. Women are affected twice as often as men, and the onset is usually before 16 years of age. There is a 57% familial tendency, and the stereotyped migraine personality is anxious, rigid, perfectionistic, and may be resentful and appear fatigued. The attacks last minutes or as long as a few days, and headaches may be separated by weeks or months and precipitated by menstruation or intake of certain foods or by alcohol. The attacks are preceded by a prodroma or "aura" of vertigo, facial flushing or pallor, and a spotty blindness or flashing of lights in the visual fields. As the headache progresses, there is increasing psychic irritability, nausea, vomiting, and constipation or diarrhea.

Migraine is caused by excess vasodilation of the extracranial vessels, such as the maxillary artery, and the dural portions of the middle meningeal, both of which are innervated by the trigeminal nerve. The prodromal symptoms are probably produced by a preparatory vasoconstriction of these same vessels. The diagnosis can be substantiated by noting rapid pain relief after digital pressure over the common carotid or external carotid arteries and also by obtaining relief with ergotamine tartarate. This condition must be differentiated from trigeminal neuralgia, periodic migrainous neuralgia, and myofascial pain dysfunction. Its treatment is almost entirely medical; aspirin may help in milder cases by acting on the peripheral vasodilation mechanisms. However, the more severe forms usually respond to systemic ergotamine tartarate, a vasoconstrictor that may be combined with caffeine (Cafergot). The syndrome may be prevented in some cases by using the serotonin antagonist methysergide (Sansert). Psychological counseling is also of benefit in some cases.

Temporal arteritis. Temporal arteritis is a primary arterial disorder that appears most commonly in women between 55 and 80 years of age. It causes intense, unilateral boring headaches over the lateral maxilla, zygoma, preauricular region, temporal region, and occipital region. A referred diffuse pain is felt over the scalp. The pain is initiated by mastication in

over 50% of cases, it may be aggravated by lying down or stooping, and digital pressure applied over the external carotid will produce temporary relief of the pain. There is a tenderness over the courses of the superficial temporal, transverse facial, and supraorbital arteries, and palpation may reveal pulseless, tortuous nodules over these vessels. Because the ophthalmic and retinal arteries may also be diseased, there is a gradual onset of blindness in more than one third of the cases.

The pathological characteristic is an arteritis with giant cell and chronic inflammatory infiltrates, intimal thickening, and frequent thromboses. Treatment with cortisone and adrenocorticotrophic hormone (ACTH) results in successful control of the headaches and, if instituted early in the disease course, will prevent blindness. In extreme cases tha pain can also be relieved by selective resection of small portions of the diseased vessels.

Coronary disease headache. The referral of a deep pressing pain to regions of the head and neck is a confusing manifestation of heart disease. It is not difficutl to recognize the association between face pain and heart disease when pain begins during exertion or emotional stress and spreads from the chest up the neck to the angle of the mandible, its most common facial location. However, facial pain may also occur without any of the classic anginal signs of chest and left arm pain, and symptoms may be localized to the maxilla, cheek, mandibular body, orbit, or occiput and may persist for longer periods of time than the typical anginal attach.

As in classic angina pectoris the pain is thought to originate from stimulation of cardiac afferent fibers by myocardial ischemia. These fibers then converge onto a common synaptic region with somatic sensory fibers that arrive from a different body region. In this case the convergence of cardiac fibers is with cervical plexus and trigeminal fibers in the upper cervical spinal cord segments. As in other forms of anginal pain, these referred headaches can be controlled by the use of nitroglycerine tablets.

Myofascial pain dysfunction. Most neuralgia-like headaches of the lower face are caused by muscle spasm in the masticatory apparatus. In their acute form, these spasms produce trismus, deviation of the mandible to the affected side, inability to occlude the teeth, and a sharp pain brought on by clenching. The pain is most often felt directly over the temporomandibular joint or above the gonial angle and less frequently over the zygoma, temporal, submandibular, or occipital regions. There may be tender palpable "knots" in the masseter and temporal muscles that can be relieved by injecting local anesthetic directly into the muscles. The acute syndrome often follows trauma such as subluxation or dislocation of the mandible and radical changes in the chewing patterns, but the most important precipitating factor appears to be emotional stress. Chronic forms of this syndrome also correlate well with underlying emotional tensions that may promote injurious jaw habits and amplify the effects of occlusal disharmonies. The chronic symptoms are most gradual in onset, appear four times as often in women, and occur most often in early adulthood, during puberty, and at the menopausal ages. Joint clicking and jerky mandibular movements are common, and sharp pains are superimposed on a dull, aching, and "drawing" pain in the lower face.

The myofascial pain syndrome must be differentiated from true osteoarthritis, migrainous neuralgias, and temporal arteritis and from acute neuritides such as maxillary sinusitis and referred odontogenic pain. Primary temporomandibular osteoarthritis may produce acute clinical features identical to acute myofascial pain. Differentiation is made by radiographically observing the hypocalcification and lipping of the condylar head, resorption of the articular tubercle, and dystrophic joint calcification.

Treatment of the acute phase of myofascial pain is aimed at interrupting the muscle spasm cycle by (1) *supportive* therapy with analgesics, tranquilizers, and muscle relaxants such as diazepam and physiotherapy such as local moist head and (2) *disengagement* of the masticatory apparatus by consciously avoiding stressful clenching, by injecting the tender muscles with local anesthetics, and by using temporary occlusal splints. The need for rest and the beginning of psychological counseling should be stressed at this time. Treatment of the chronic problems will rely even more heavily on psychological counseling to identify and compensate for abnormal muscle habits. Correction of occlusal discrepancies should proceed only after the acute myospasms have been controlled. Surgical treatments, such as cortisone joint injections, arthroplasty, and condylectomy, are indicated only after the presence of osteoarthropathy or ankylosis has been confirmed.

Maxillofacial Neuritis

The often misused trerm *neuritis* means literally "inflammation of nerve" and will be used here to identify acute, reversible irritations of maxillofacial nerves. Neuritis may occur in sensory, motor, or autonomic nerves and results from peripheral pathology that infects, compresses, entraps, or erodes the adjacent nerves. Neuritis is significant because it is a signal of an acute pathological condition and because neuritis that is allowed to persist may eventually progress to degenerative and irreversible neuropathy. Sensory neuritis is almost always manifested as pain, but its character depends on the location and nature of the primary lesion. It is also characterized by a lowering of the pain thresholds, probably as a result of alterations in the central gate control mechanisms. For example, with chronic neuritis from a periapical abscess, irritation of the smaller pain-sensitive fibers seems to have the effect of sensitizing the brain stem synaptic regions to respond more readily to any kind of incoming stimulation. This slight opening of the gate may explain why tactile or light pressure stimulation of tissues around periapical abscesses will result in pain responses, even in the locally anesthetized state.

Bell's palsy. Bell's palsy is an isolated facial paralysis of sudden onset caused by a neuritis of the seventh nerve within the facial canal. It occurs often in the young adult man with a history of recent exposure to local cold, such as sleeping next to an open window, or in some cases it occurs after infections of the nasopharynx or masticator spaces. The clinical appearance is a unilateral flaccidity of all facial muscles with loss of eyebrow and forehead wrinkles, drooping of the eyebrows, flattening of the nasolabial furrow, sagging of the corner of the mouth, and collection of food in the buccal vestibule. Patients are unable to frown or raise their eyebrows, and they cannot close their eyes or purse their lips. If the neuritis has extended as far centrally as the point at which the chorda tympani nerve joins the facial nerve trunk, then taste will be impaired over the anterior two thirds of the tongue on that side. Patients may also complain of loud noise intensification because of damage to the nerve of the stapedius muscle. The histopathological sign of Bell's palsy is edema in the nonexpansile facial canal. It is not known whether the precise source of inflammation is in the nerve fibers themselves, in related connective tissues, or in the periosteum of the canal walls. After its sudden onset the paralysis begins to subside within 2 to 3 weeks, and gradual complete recovery occurs in over 85% of patients. It is necessary to differentiate between the isolated lower motor neuron lesions of Bell's palsy and the more complex upper motor lesions that may result from vascular lesions or neoplastic lesions in the pons. In these central "upper motor" lesions, upper facial muscle function is spared.

In the early stages of Bell's palsy, inflammation may be suppressed by using systemic cortisone or ACTH. Surgical decompression of the facial canal may also aid in edema control

if it is performed during the first few days. Galvanic stimulation of facial muscles may counteract the neurotrophic effects. The cornea must be protected from abrasion by applying lubricants or in some cases by suturing the eyelids. If paralysis is permanent, the facial tissues may be given artificial support by means of prosthetic devices or by subcutaneously grafted masseter or fascia lata slings. In some cases a surgical redirection of the accessory nerve into the degenerate seventh nerve tissues has been effective in restoring some facial muscle function.

Other cranial motor neuritides. A rare trigeminal neuritis has been described that is similar to and in some cases accompanies Bell's palsy. This neuritis is unilateral and results in total anesthesia or pain, usuallly over the mandibular division. It is also associated with a unilateral total loss of function in the masticatory muscles supplied by the trigeminal nerve. The cause is unknown, and gradual complete recovery has been reported for most cases.

Cranial nerves three, four, and six are often affected by traumatic lesions, either directly by damage or indirectly by orbital edema. Third nerve neuritis is confirmed by observing the features of Horner's syndrome, which include ptosis, pupillary constriction, and anhidrosis. Upward gaze is also deficient with this lesion. Trochlear neuritis appears as an inability to rotate the eyeball down and outward. Paralysis of the sixth cranial nerve results in impaired lateral gaze. In each of these cases the primary neuritis can be differentiated from local muscle entrapment by a forced duction of the individual muscles.

Paranasal sinus neuritis. Less than 5% of all headaches are related to paranasal sinusitis, and these painful neuritides are usually accompanied by symptoms of nasal discharge, epistaxis, otalgia, and fullness in the ears. The maxillary sinus is most frequently involved, and tapping over the infraorbital and zygomatic regions elicits a dull aching response. However, the pain may also be referred to the maxillary teeth. The diagnosis of sinusitis is usually confirmed by observing pus in the middle meatus and detecting radiographic air-fluid cloudiness in the sinuses.

Salivary gland and mucosal neuritis. The most common causes of salivary gland neuritis are infection of the glands and obstruction of the ducts by bacterial plugs or sialoliths. In both cases dull pain and pressure sensations are correlated with eating or "milking" the gland.

Mucosal neuritis has many local causes. For example, an intense mucositis with stinging pain may be caused by acute drug idiosyncrasy. An itching neuritis is typical of neuritis from overuse of systemic antibiotics. Galvanism, a result of the presence of adjacent, dissimilar restorative metals, may also result in a burning ulceration.

Odontogenic neuritis. Pulpal disease is the most common cause of dental pain, and in the initial hyperemia phase, pain is often severe and exaggerated in response to stimulation by heat or cold. As pulpal disease progresses, pulpitis pain becomes more spontaneous, sharp, and throbbing because of the inflammation of poorly myelinated nerves in the rigid-walled puplp chambers. As the pulps become necrotic, however, the pain is less intense and is most likely to be elicited by tapping or pressing on the affected tooth because the nerve irritation is now primarily caused by fluid pressures in apical and periapical tissues.

The pain of periodontal disease is usually less intense, dull, and gnawing in character and is without the pulsations that occur with pulpal neuritis. Extension of pericoronitis along adjacent muscle and fascial planes may cause symptoms of neuritis, although the clinical

picture in these cases may be dominated by signs of trismus and myofascial spasm. Acute periosteal and alveolar disease may set up referred neuritis such as the preauricular pain that occurs during dry socket in the mandibular third molar region.

Central Disorders

Most central nervous system pathology is vascular in origin, and strokes at many levels of the brain may cause neurological disorder in the maxillofacial region. For example, the frequent occlusion of basilar artery branches interrupts the blood supply to the emerging rootlets of the third to seventh cranial nerves as well as the long sensory and motor tracts that course in the ventral pons. This produces simultaneous neurological deficits in many cranial nerves along with hemiparesis and hemianesthesia, a pattern that helsp to differentiate this lesion from more isolated peripheral diseases. The symptoms of intracranial neoplasia vary greatly and also may mimic peripheral disease. Differentiating features, however, include constant progressive headaches, mental deterioration, and spreading seizures. Among the more localized intracranial tumors, the lesions of neurofibromatosis (von Rechlinghausen's disease) may involve sensory cranial nerves. The acoustic neuromas often produce secondary pareses of trigeminal and facial nerves by compression. Primary trigeminal tumors are extremely rare. There are ofther forms of intracranial disease that may present problems in the differentiation of maxillofacial neurological disorder. They include multiple sclerosis, tertiary neurosyphilis, syringobulbia, thalamic syndrome, meningitis, and psychalgia

Syringobulbia. Syringobulbia is a slowly progressive and degenerative disease of the medulla oblongata that causes a great range of maxillofacial neurological change. The most striking clinical feature is a dissociated sensory loss in which there is a loss of pain and thermal sensibilities with retention of touch. Other classic signs include lingual atrophy, vertigo, palatal paralysis, nystagmus, facial weakness, Horner's syndrome, and especially hoarseness. Although pain and paresthesias are uncommon, there may be spontaneous trigeminal burning and aching that resembles the pain of tabes dorsalis. Men are more often affected, usually before age 30, and after an insidious onset the disease develops rapidly in the first few weeks.

The histopathology of both syringobulbia and the related syringomyelia is a cavitation and gliosis that begins near the central canal of the brain stem or spinal cord and then enlarges ventrally to gradually interrupt the large sensory and motor fiber tracts that cross over in these ventral regions. The cavitation (syringes) most often begin in the cervicothoracic regtions (syringomyelia) and cause sensory loss and wasting of small muscles of the head. The cavitation often then spreads rostrally into the medulla (syringobulbia) to involve first the twelfth and tenth cranial nerve nuceli and eventually the other cranial nerve nuclei. The cause is unknown but there are indications that the lesions arise congenitally. The main differential diagnosis in syringobulbia is with multiple sclerosis, poliomyelitis, and tabes dorsalis. There is no satisfactory treatment, and, because of the condition's slow course, preventive physiotherapy and dental care should be actively pursued.

Thalamic syndrome. The thalamic syndrome is a rare pain condition caused by damage to the lateral thalamic nuclei on the side opposite the symptoms. The pains are aching, burning, and gnawing; they spread unilaterally and are felt deeply in wide facial and trunk regions. The threshold for pinprick is raised in this condition, but many types of stimuli, especially deep pressures, will evoke the delayed and inappropriate pain. In this respect the thalamic syndrome resembles the hyperpathia seen in posttraumatic neuropathies of the peripheral nerves. The pathogenesis is unknown, but traumatic and vascular lesions that

involve diencephalic and cortical tissues have been implicated. This condition must be differentiated from causalgia, peripheral neuromas, and intracranial tumors. Treatment with narcotics is only partially effective, and prefrontal leukotomy may be effective in some cases.

Psychalgia. The incidence of maxillofacial psychalgia, which is pain of mental and truly nonorganic origin, is unknown, but to the patient, it may be severe and real. These pains tend to be vague and nonspecific, shifting over poorly defined nerve distributions, and usually cannot be elicited by specific stimulations. Although pain is the common complaint, hysterical conversions may result in anesthesia, paresthesia, blindness, deafness, and such objective signs as flaccid facial paralysis, tics, dermatological outbreaks, vomiting, and even angioneurotic edema. Symptoms are strongly correlated with emotional stress, and other signs of character disorder or psychosis may be prominent. Almost any form of medical or surgical treatment, even placebo, will yield temporary symptom relief, and these patietns will often prescribe their own treatment and even seek multiple deforming surgeries. Because of the high incidence of character disorders related to psychalgia, psychological screening tests such as the Minnesota Multiphasic Personality Index (MMPI) may be helpful. The only effective treatment is psychotherapeutic.

The diagnosis of psychalgia should not be made lightly or as a last resort. It should be recognized that maxillofacial neurological problems, especially pain, are almost always complicated by an emotional component. The identification of even a strong psychologic component should not be the signal to abandon the search for and treatment of organic problems. Nor should those patients whose symptoms seem bizarre or even humorous ("crawling, puffing, inside-out") be quickly relegated to the psychalgia category. Facial sensation is a personal matter, and the honest patient may not "feel" his abnormality in textbook terms.

Diagnosis

In the process of making a final diagnosis of maxillofacial neurological problems, the history and examination should lead to a descritpion of four basic disease elements: (1) *symptoms,* (2) *pathology,* (3) *location,* and (4) *etiology* (see following outline).

I. History
    A. Symptom classification
        1. System
            a. Sensory (pain, numbness, paresthesia)
            b. Motor (weakness, spasm)
            c. Autonomic (nasal, ocular, cutaneous, gastric)
            d. Special sense (visual, auditory, olfactory, taste)
        2. Quality
            a. Strength (mild, moderate, severe)
            b. Onset (spontaneous, induced, triggered)
            c. Duration (momentary, minutes, days, constant, paroxysmal)
            d. Nature (dull, aching, burning, pulsing, sharp, itching)
        3. Localization
            a. Precise (V_1, V_2, V_3, VII, IX, other)
            b. Unilateral or bilateral
            c. Migrating, spreading, radiating
            d. Diffuse

4. Influences
    a. Movement or function (face, jaw, body)
    b. Position (head, jaw, body)
    c. Activities (exertion, eating, talking)
    d. Time of day, month, or season
5. Symptom course
    a. Unchanged
    b. Response to therapy (drug, surgery, other)
    c. Changes in character

B. Systemic and environmental factors
1. Metabolic disorders (anemia, diabetes mellitus, uremia)
2. Connective tissue disorders (arthritis, scleroderma, lupus erythematosus, Sjögren's syndrome)
3. Toxicities (heavy metal, organic chemical, food, drug, alcohol)
4. Nutritional disorders
5. Infectious disorders (herpes zoster, meningitis, syphilis, leprosy, diphtheria)
6. Vascular disorders (coronary artery disease, temporal arteritis, Raynaud's syndrome, hypertension)

C. Primary neurological and psychiatric disorders
1. Neuralgia (trigeminal, vagoglossopharyngeal, intermedius, period migrainous)
2. Migraine
3. Central disorders (syringobulbia, thalamic syndrome, seizure disorders)
4. Neuroses (psychalgia)
5. Psychoses
6. Multiple sclerosis

D. Neuritis factors
1. Maxillofacial trauma (facial fracture, prosthetic irritation, iatrogenic)
2. Infection (odontogenic, periodontal, facial)
3. Paranasal sinusitis
4. Otalgia
5. Saivary gland disoders (sialolith, adenitis)
6. Mucosal disorders (mucositis, herpetic ulcer)
7. Motor neuritides (Bell's palsy, ocular palsies, Horner's syndrome, myesthesia)
8. Myofascial dysfunction

II. Examination
A. General cerebral function
1. Consciousness level
2. Gross movements

B. Cranial nerve function
1. Motor functions
    a. III, IV, VI (extraocular muscle function)
    b. V (masticator muscle function)
    c. VII (facial muscle function)
    d. IX, X (palatal, pharyngeal, laryngeal function)
    e. XI (sternocleidomastoid, trapezius function)
    f. XII (tongue function)

        2. Somatosensory functions (V, VII, IX nerves)
            a. Pinprick sensitivity
            b. Pressure pain sensitivity
            c. Pulp tester sensitivity
            d. Fine and two-point tactile
            e. Vibratory sensitivity
            f. Hot, cold sensitivity
        3. Special functions
            a. I (smell)
            b. II (vision)
            c. VII, IX (taste)
            d. VIII (hearing)
        4. Autonomic functions
            a. Sympathetic (pupillary dilation, eyelid tone, sweating, vasoconstriction, salivation)
            b. Parasympathetic (pupillary constriction, serous salivation)
    C. Special reflexes or tests
        1. Corneal reflex
        2. Jaw-jerk
        3. Electromyography
        4. Nerve conduction studies
        5. Minnesota Multiphasic Personality Index
    D. Acute maxillofacial neuritis lesions
        1. Odontogenic (caries, periodontal disease)
        2. Osteal-periosteal (cysts, osteomyelitis)
        3. Myofascial
        4. Salivary glandular (ductal obstruction, adenitis)
        5. Mucosal (mucositis)
        6. Paranasal sinus
        7. Vascular
    E. Diagnostic blocks
        1. Placebo effects
        2. Vasocostrictor effects
        3. Anesthetic effects.

History

Chief complaint and present illness. Disease symptoms should first be expressed in the patient's own terms and then evaluated by the clinician to determine whether the problem seems to be motor, sensory, autonomic, or a combination and to establish the basic nature, intensity, location, onset, and course of the complaint. At this time a tentative *symptom diagnosis* may be made (such as neuralgia, headache, paralysis, dysesthesia).

Past history. The neurological history should begin with a questioning about systemic disease and environmental conditions that have known neurological effects in the maxillofacial region. Next, patients should be questioned about the past history of major neurological and psychiatric disorders. Once the systemic, environmental, primary neurological, and psychiatric disease factors have been considered, questioning should turn to the incidence and nature of direct lesions in the maxillofacial tissues themselves. On the basis of information gained in the neurological history alone, it may be possible to make a tentative *pathology diagnosis* (such as infectious, degenerative, arteritic, or demyelinating) or

an *etiology diagnosis* (such as posttraumatic, odontogenic, psychogenic, or diabetic).

Examination

Major goals of the examination for maxillofacial neurological disorders should be detection of acute lesion sources of neuritis within the tissues and location of the disease process and site of pathology. The disease is located by examining first the general and then the more specific functions. The examination should begin with an observation of cerebral functions such as level of consciousness and gross movements and continue with a study of specific cranial nerve reflex functions. In addition to the classic techniques of inspection, tissue palpation, and reflex stimulation, the diagnostician may benefir from the use of special tests such as electromyography, nerve conduction studies, EEG, and lumbar puncture. Prsonality screeing such as the MMPI can also be done at this time. Finally, a detailed search for specific and local sources of acute neuritis should be done, using all routine dental diagnostic skills and aids such as inspection, palpation, radiography, and electrodiagnosis.

Diagnostic nerve blocks

A simple and most revealing technique for locating and characterizing neurological lesions is the diagnostic block. The level of a particular lesion may be determined by depositing small amount of local anesthetic solution, first at the most superficial nerve levels and then at progressively more central levels. After each injection the patient should be carefully questioned regarding change of symptoms, especially in response to direct stimulation of trigger points or sensitive tissues. These blocks may lead directly to a localization diagnosis. For example, blocks of the sphenopalatine ganglion by injecting deeply within the descending palatine canal may be diagnostic of periodic migrainous neuralgia. Diagnostic blocks are also helpful in differentiating between the precise superficial trigger zones of classic neuralgias and the deeper, more diffuse loci of neuritides and neuropathies. Diagnostic nerve blocks may also define for the clinician the specific type of nerve fiber that is initiating the pain response. A rigid protocol is recommended in which placebo (saline) nerve block is injected in the region of pain complaint. Relief of pain with placebo for more than a few minutes suggests a possible psychogenic basis for the pain. If pain persists, however, a second injection should be made with 0.25% procaine or lidocaine. Relief of pain after this injection indicates that pain is initiated by small, nociceptor fibers or autonomic fibers and therapy should be pointed toward correcting this neuropathy. If pain is still persistent after the second, small-fiber nerve block, then a final block of both small and large fibers should be given using 1% to 2% procaine or lidocaine. Pain relief after this block indicates larger fiber mediation of pain reflexes, commonly seen in myofascial pain syndromes. If pain is unrelieved, even after total nerve block, a central nervous system lesion or a psychogenic basis for pain or both should be suspected.

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In summary, when the neurological history and examination of a patient with a maxillofacial neurological disorder has been completed, a differential diagnosis can be developed. In each case, the four key elements of the diagnosis should have been investigated. A sample, all-inclusive diagnosis would be posttraumatic neuralgia associated with infraorbital neuroma (etiology, symptom, location, and pathology).

Treatment

Because nervous tissues often do not withstand or recover completely from repeated insults by infection, inflammation, metabolic toxicity, and especially trauma, preventive treatment should begin by removing sources of acute neuritis and barriers to nerve regeneration. It is also clear that neurological problems of the maxillofacial region are varied, since they encompass motor, autonomic, and sensory systems along with the diverse components of pain. Effective treatment therefore must have a similar broad base of medicine, surgery, physiotherapy, and psychotherapy.

Medical treatments

Analgesics. The type of analgesic selected for pain control is determined by the severity and the nature of symptoms as well as the location of the neurological lesion. Mild, chronic neuritis caused by inflammation in the skin, mucosa, and joints or when vasculitis and edema are present are bect managed by mild analgesics, such as salicylates, propoxyphene, or para-aminophenols, for which the suspected site of action is in the peripheral tissue and paravascular receptors. Potent analgesics such as narcotics, narcotic antagonists, and synthetic agents such as pentazocine are indicated for more severe pain when the suffering and the affect component of pain are predominant. Because these agents seem to act in subcortical, reticular, and limbic cortex regions, they are more effective against more diffuse and central pains such as visceral and periosteal pains, nerve invasions by carcinoma, neuropathies, syringobulbia, and the thalamic syndrome. However, not even the most potent narcotics are effective in the relief of true paroxysmal pain such as trigeminal, intermedius, or vagoglossopharyngeal neuralgias. They are of marginal value in postherpetic pain, migraine, posttraumatic neuralgias, tabes dorsalis, and periodic migrainous neuralgia. Because of the potential for addiction, chronic pain syndromes should not be managed solely by narcotic analgesics.

A number of studies have shown that the timing and approach to an analgesic drug may be more important than the particular type of analgesia used. When analgesics are used for acute or postoperative pain they are most effective when used frequently, in small doses, and early in the pain cycle on an "as needed" basis. In chronic pain syndromes, however, the "as needed" approach should be replaced by a strict "time contingency" schedule of drug use. In this manner a reasonable blood level of analgesic is maintained, and patients take medication "by the clock" rather than "as they feel". In this manner cyclical pain phenomena are more likely to be controlled and problems of drug habituation or addiction are minimized.

Anticonvulsants. Based on the concept that many paroxysmal pains are produced by the epileptiform mechanism, anticonvulsant agents have proved effective in the treatment of severe neuralgia. Phenytoin (Dilantin), administered in doses of 100 mg four times daily, has controlled the pain of idiopathic trigeminal neuralgia in approximately 50% of cases. Although phenytoin has been a safe agent for chronic use, it has not been as effective in the later stages of the neuralgia. It is known to act on peripheral nerve transmission and by a depression of brain stem trigeminal nuclei function. However, within the last 5 years the anticonvulsant drug of choice for severe neuralgia states has become carbamazepine. This agent acts similarly to phenytoin as a depressant of peripheral nerve transmission but has a more potent effect on polysynaptic systems in the trigeminal descending tract in the medulla and ultimately in the thalamic nuclei related to head and neck pain transmission.

In a large number of clinical series, carbamazepine administered in doses of 200 to 800 mg daily has been effective in controlling tic douloureux in 85% to 100% of cases. This drug has also been useful in the control of intermedius, vagoglossopharyngeal, multiple sclerosis, and posttraumatic neuralgias. However, it has been less than 50% effective in postherpetic neuralgia, phantom pain, and periodic migrainous neuralgia.

Side effects including slight sedation, dizziness, nausea, and occasional rash are seen with carbamazepine. Although the incidence of complications is low, agranulocytosis, thrombocytopenia, and trigeminal paresthesias have been reported in approximately 7% of cases; these toxic effects seem to be most prominent in older, debilitated patients. In spite of promising results reported with carbamazepine, trigeminal neuralgia pain returns in approximately 10% of patients.

Systemic vasoconstrictors and antihistamines. Classic migraine and other vascular headaches have been managed by ergotamine tartaragte, which is known to counteract directly the painful dilation of craniofacial arteries. Methysergide preparations, which may act as a serotonin and histamine antagonist, have also been effective in as many as two thirds of cases of periodic migrainous neuralgia. Recent clinical studies have also shown that the beta-adrenergic blocking agent Propanolol (Inderal) is effective in relieving vascular facial pain disorders. The usual trial dosage is 40 mg given three times daily.

Corticosteroids. Agents such as ACTH and adrenal corticosteroids are indicated when the neurological disorder is a result either directly or indirectly of inflammation. They may be helpful in the early stages of seventh nerve neuritis (Bell's palsy) to prevent degeneration of the nerve trunk within the facial canal. Steroids may also be indirectly effective in preventing further degeneration when vasculitis is a factor, such as in connective tissue diseases, diabetic neuropathy, and temporal arteritis.

Tranquilizers. Minor tranquilizers such as the benzodiazepines (Valium, Librium) have no direct analgesic effects but may be helpful as an adjunct in the overall management of chronic pain in which the affect is a strong pain component. They may also be useful in relieving the acute stages of tension headaches and myofascial pain dysfunction because of the indirect skeletal muscle relaxation effects of these agents.

Antidepressant and psychotropic drugs. Because of the nearly universal association of chronic pain and emotional disorder, antidepressant and major tranquilizer drugs are necessary and effective treatment for many patients. Many studies have documented the therapeutic value of tricyclic antidepressants given for depression-related chronic pain, especially myofascial pain disorders. This class of drugs is contraindicated in patients with significant cardiovascular disease and inpatients with latent schizophrenia. Primary agents that have proved effective are amitryptiline, imipramine, and doxepin given in dose ranges of 75 to 150 mg daily for 4 to 6 months.

In more severe forms of psychopathology, in which facial pain may represent a conversion hysteria or a "mask" for psychosis, major tranquilizers such as chlorpromazine (Thorazine) or fluphenazine (Prolizin) have been effective.

Surgical control of maxillofacial pain

In the surgical control of maxillofacial pain, manipulations are made at four main levels of the sensory nervous pathway: (1) the peripheral nerves, (2) the sensory ganglia and

their roots, (3) the brain stem, and (4) the thalamus-cortex. Surgery can be performed by using decompression, simple nerve section, selective thermal lesions, cautery, cryosurgery, and mechanical and chemical necrosis.

Formerly, the objective of most of these techniques was denervation of the pain regions. Although rapidly effective in relieving pain in many cases, denervation, rhizotomy, and tractotomy surgeries also have the potential to result in secondary neuroma formations, causalgia, anesthesia dolorosa, and phantom pains. Recent years, therefore, have seen the emergence of less destructive and more selective thermal lesions and decompression procedures for the control of pain.

Treatment of nerve injuries. Although injuries to nerves often cannot be prevented, it is possible to decrease the incidence and severity of clinical problems by taking steps to avoid aberrant regeneration (see section on histopathology). For example, a major hindrance to proper regeneration is secondary irritation to injured nerves. Therefore, attempts should be made to avoid nerve compressions from sources such as displaced root tips, fragments of alveolar bone, or prosthetic devices that impinge on nerve branches. After facial fracture the nerves should be protected from entrapment by malplaced bone segments, irritation from transosseous wires, and shearing effects caused by mobile fractures. Other steps in the treatment of nerve injuries are encouragement of the regeneration of nerve fibers into their appropriate distal tissues and prevention of the formation of barriers to regeneration such as scar tissue.

Severed nerves that can be directly exposed should be sutured immediately by first cutting back in both the proximal and distal nerve stumps until a symmetrical cross section of nerve is obtained that bleeds freely. The the perineural sheaths of both prepared nerve cylinders are engaged by fine linen (No 10-0 Tevdek) or wire suture, with the two ends oriented properly to one another, while the remaining nerve circumference is sutured. To protect the repair site further from the ingrowth of foreign scar tissue, it is recommended that the nerve be wrapped in Millipore filter. The repaired nerve should then be stabilized by suturing it loosely into adjacent soft tissues. Although the optimum time for nerve suture is usually at the time of acute injury, if the wound has been grossly contaminated with significant tissue loss, it may be advisable to label the cut nerve ends with sutures and delay the nerve repair at least 10 days. When there has been no evidence of returning nerve function within 4 to 6 weeks after injury, it is advisable to explore the injury site. The objectives of this exploration are to decompress the nerve by evacuating hematoma, removing impinging bone fragments or foreign bodies, and resecting the neuroma segments and, finally, to rejoin the nerve segments using the suture techniques just described.

The prognosis of good function returning after nerve injury is less when motor nerves have been injured and when long segments of nerve have been lost. Although nerve grafting has been used successfully in the maxillofacial region to repair large nerve deficits, little is known at this time about the ideal conditions and the prognosis of this procedure. In all cases of nerve damage, it is important to protect and maintain the nonneurological tissues that have been denervated to assure their maximum function when regeneration does occur.

Therapeutic anesthetic blocks. Blocks of peripheral nerve, myofascial, or neurovascular trigger zones by anesthetic agents are of considerable value in managing pain syndromes. They have value as diagnostic aids as outlined previously. They are also useful as a palliative procedure to achieve instant relief for a suffering patient and to "buy time" for the clinician to establish more definitive therapy or to make a more thorough diagnosis.

Palliative blocks with long-acting anesthetics such as 0.5% bupivacaine with epinephrine, for example, may be given daily or at longer intervals to control acute triggered paroxysms of trigeminal neuralgia while effective levels of anticonvulsant drugs are being attained.

Nerve blocks have prognostic value. They may indicate whether permanent denervation or neurolysis would be effective and whether a given patient can tolerate paresthesias during everyday functions. Blocks may also be used in therapy, particularly for pain disorders of a cyclical nature in which remissions are common such as the major neuralgias, posttraumatic pain, and certain myofascial pain syndromes. Using anesthetic blocks for therapy is highly controversial because it is seen as strictly symptomatic (and not curative) treatment and because no scientific basis has been established for its effectiveness. Nevertheless, anesthetic blocks given for a wide variety of pain syndromes in many body parts have repeatedly brought pain relief for longer than the known pharmacologic action of the drug itself. Blocks are given at 48-hour to weekly intervals into nerve distributions and particularly into zones of previous trauma and neural or muscular trigger foci.

Peripheral denervation. The objectives of peripheral denervation are to give prompt and sustained relief from severe pain, either as a temporary palliative measure or to avoid the hazards of radical craniotomy procedures. It is indicated inold and debilitated patients, in cases in which the first and second divisions of the trigeminal system are involved, in neuralgias with a greater likelihood of bilateral pai such as multiple sclerosis and carcinoma, and in retarded patients who cannot cooperate tothe extent needed to perform more conservative neurolysis. It is especially indicated for the patient with short life expectancy resulting from painful invasive cancers. Interrutpion of the peripheral nerves may be brought about by injecting a 95% solution of ethanol into the affected nerve branch or by surgical exposure and sectioning of the branch. Although both techniques effectively bring about wallerian degeneration of the peripheral branches, the direct severance techniques may be preferred to alcohol injections because they are more precise and because repetitive neurectomies are more easily done. The specific objectives of peripheral neurectomy are to eliminate as much as possible of the affected nerve branch and also to attempt to block its regeneration. Therefore, after the nerve has been exposed and before it is cut, dissection should be carried distally into the terminal tissues as far as possible. Proximally the maximum amount of nerve tissue should be avulsed by rolling the nerve around a hemostat. Finally, the nerve foramen should be obliterated with sterile wooden pegs, amalgam, or bone plugs to block further nerve regeneration.

In the third trigeminal division, inferior alveolar and lingual nerves are commonly resected. The lingual nerve can most easily be exposed at the inner surface of the mandible in the third molar region by making a vertical incision along the internal oblique ridge. This same incision can be used to approach and cut the inferior alveolar nerve at the mandibular foramen. However, an extraoral approach to inferior alveolar resectin is simpler and has the advantages of avoiding damage to the lingual nerve and permitting direct obliteration of the mandibular canal. In this procedure, a 1-cm incision is made at the inferior border of the mandible in the antegonial notch region. Dissection is carried directly to bone, and, by use of a nasal speculum to retract the masseter, the outline of the mandibular canal can be seen with the aid of intraoral transillumination. The lateral bony plate of the canal is removed, the nerve is avulsed, and the canal is finally obliterated. When this procedure has been combined with mental nerve resection, the entire intramandibular length of the inferior alveolar nerve can often be removed intact.

Most clinicians prefer the intraoral approach for infraorbital neurectomy. Both the superior labial and lateral nasal branches of the nerve should be dissected free, and then the infraorbital foramen can be clearly visualized. After avulsion, canal obliteration, and suture, a firm pressure should be maintained over the infraorbital region for at least an hour to prevent the profuse bleeding that may follow this procedure.

Branches of the first trigeminal division, including the supraorbital, frontal, and supratrochlear nerves, may be exposed through an incision in the midportion of the unshaven eyebrow. Care should be taken to avoid damaging the lacrimal gland in the lateral roof of the orbit.

Peripheral neurectomies are minor operations that may be performed comfortably with local anesthesia and sedation since there is little risk even in elderly or debilitated patients. This operation may be completely effective in some cases, especially if repetitive neurectomies are done, and over 60% of patients can expect to be pain free for 4 years after neurectomies have been started. However, pain usually does recur slowly as axonal regeneration takes place, and because neurectomies may serve to delay an inevitable radical treatment, it is important that cases be selected carefully.

Selective thermal lesions. In the past 5 years the surgical treatment of choice for paroxysmal trigeminal neuralgias has become radiofrequency (RF) thermal lesions performed most often at the level of the trigeminal ganglion and sensory root. Preliminary research had shown that RF thermal lesions of 60° to 70°C, when applied to peripheral nerves, have the effect of selectively destroying small nerve fibers but at the same time retaining the larger nerve fibers. In clinical application, this has had the effect of eliminating abnormal pain and triggering functions yet preserving normal tactile sensation for the patient. It therefore represents a major improvement over previous neurectomy or alcohol or rhizotomy lesions, which produced a disturbing total loss of sensory function.

In the most common RF *thermoganglionlysis* technique a 22-gauge needle, insulated except at its tip, is inserted through the skin of the cheek, passed medial to the mandibular ramus and through the foramen ovale to come to rest at the ventral aspect of the trigeminal ganglion in Meckel's cavity. After fluoroscopic checking of location, the patient, still awake, is given a mild electrical stimulus through the needle tip to elicit parethesias and determine whether the needle is in the proper position for nerve lesion. When this is assured, a general anesthetic or deep sedative is given to the patient and one or two thermal lesions of 60° to 70°C are made for 30 seconds each. Results of these lesions have beenvery good, with a high control rate of pain, minimal complication, and a pain recurrence rate of approximately 20% per year.

Recently, RF lesions of more peripheral nerves, including occipital, inferior alveolar, infraorbital, and supraorbital branches, have produced very good results. In these techniques the same basic approach is used as described previously except that the lesions can be made in the outpatient setting, general anesthesia is not required, there are no risks of damaging vital intracranial structures, and lesions can be repeated more easily if necessary. The selective thermal lesions may, in the near future, open a new era in less destructive surgeries for controlling neuropathic pain, replacing in part the pharmacological approaches.

Craniotomy procedures. The most precise lesions of the sensory ganglia and root tissues are made by means of direct craniotomies. With these techniques, a simple incision of the dural sleeve that surrounds the ganglia and sensory roots is made, followed by a gentle

freeing and manipulation of the ganlgion and its roots. For unknown reasons, this manipulation alone has the surprising effect of pain elimination with retention of tactile and proprioceptive senses.

More recently, a major modification has been made in decompression procedures at the level of the trigeminal sensory root. In this operation, using a posterior fossa approach, the superior cerebellar arteries are elevated away from the sensory root and a Teflon sponge barrier is placed between the root and the vessels. This operation has proven highly effective in a large series of cases of neuralgia. However, the long-term recurrence rate of these central decompressions is not known.

The most definitive technique for eliminating severe pain, such as tic douloureux, continues to be the retrogasserian rhizotomy. With this technique, the sensory root fibers are selectively lesioned at some point between the ganglion and its point of root entrance into the pons. Rhizotomy is most often accomplished by a temporal approach to the middle cranial fossa, although a more hazardous posterior fossal approach is also effective. Root fibers are selectively lesioned by cutting, electrocoagulation, or cryosurgery, which results in a complete and permanent degeneration of nerve cell bodies and their centrally projecting fibers. In spite of the apparent permanence of this procedure, there is a neuralgia recurrence rate of 13% to 15% that may be a result of either incomplete nerve lesion or the action of aberrant sensory fibers in the motor trigeminal root. This operation may be complicated by painful herpes zoster lesions, corneal ulcerations, a variety of trophic lesions, and the painful anesthesia dolorosa paresthesias. In addition to neuralgia associated with the trigeminal nerve, rhizotomy is also indicated for controlling intermedius and vagoglossopharyngeal neuralgia, periodic migrainous neuralgia, and uncontrolled neoplasms of the head and neck. Rhizotomy procedures, although useful in selected cases, have largely been replaced by the more conservative radiofrequency and decompression techniques described previously.

Tractotomy procedures. The primary descending tracts of trigeminal, as well as eighth, ninth, and tenth nerve sensory fibers, may be interrupted by lesions placed 4 mm below the obex in the medulla. A desirable sensory dissociation often results from this operation, in which tactile senses are retained but pain sensitivity is lost. It is especially indicated when pain is bilateral or when many cranial nerves are involved simultaneously. However, medullary tractotomy has a significant higher mortality than rhizotomy, and it has been found that pain may recur in as many as 50% of patients within 1 year of the operation.

Tractotomy may also be performed at the level of the pons or midbrain to control maxillofacial pains such as postherpetic neuralgia. However, none of the tractotomy procedure described here have been effective in eliminating the suffering or affect component of pain. Therefore these once highly regarded procedures have gradually come into limited use in the past decade and have given way to either rhizotomy or to surgical lesions made in the thalamus or cortex.

Thalamotomy and cortical leukotomy procedures. Selective lesions in the thalamus and cortex are the only known surgical treatments for most varieties of central pain such as phantom pain, thalamic syndrome, tabes dorsalis, and postherpetic facial neuralgia. Lesions of the posterior medial thalamus are made by RF electrodes that are left implanted for a period of time to allow repetitive enlargement of the original lesion, thus adapting the lesion size to compensate for spreading of the pain disease.

Cortical leukotomy consists of white matter lesions of the frontal lobes and has the interesting result of decreasing the suffering or emotional component of pain without causing any significant change in the perception thresholds. Patients do not complain of pain spontaneously but will describe the painful stimulus in great detail if questioned about it. It is for this reason that this technique is indicated when fear and suffering are prominent features of the disease, which is often the case in patients with malignant tumors of the oral and pharyngeal regions; their pain is accompanied by fear of suffocation and exsanguination. An important danger with leukotomy procedures is the potential to induce significant personality defects and loss of "social senses". For this reason these procedures are usually reserved for cases of central suffering pain that cannot be managed by peripheral neurectomy, sensory rhizotomy, or medical psychiatric therapy.

New approaches to treatment

Even though current medical and surgical techniques are effective for many neurological problems, many of these approaches afford only partial or temporary control, and often the available techniques, such as central nervous system surgery, are too drastic to applied to the numerous milder syndromes found in the maxillofacial region. The side effects of traditional therapies may be more troublesome than the original disease problems. For these reasons there is a continuing search for new treatments of neurological disorders, especially for chronic, idiopathic pain conditions.

Physiological inhibition of pain. The use of physiological counterstimulation to inhibit chronic pathological pain grew out of the gate control concept of pain threshold. The earliest applications included the technique of dorsal column stimulation in which subcutaneous electrical transmitters have brought about pain relief in over half the cases of chronic back pain as well as in difficult cases of phantom limb pain and pain from invasive carcinoma. Daily stimulation by similar transmitter-electrode systems that have been placed adjacent to the trigeminal rootlets has also proved effective in relieving paroxysmal pains. Even peripheral nerve stimulations have shown promise by use of this technique. For example, low-voltage RF waves have been passed through the surface and needle electrodes that were implanted into the infraorbital, lingual, and auriculotemporal nerves of patients with idiopathic trigeminal neuralgia.

Transcutaneous neural stimulation. The most widely used and effective stimulation approach, however, is transcutaneous neural stimulation (TNS). With TNS, cutaneous bipolar surface electrodes are placed in the painful body regions and low-voltage electrical currents are administered by the patient. Best results have been obtained when intense stimulation is maintained for at least an hour daily for more than 3 weeks. TNS portable units are in widespread use in pain clinics throughout the world, and TNS has proved most effective against neuropathic pain such as phantom limb pain and nerve injury pain. It has been effective in a smaller percentage of patients with facial pain but, when successful, is an excellent noninvasive treatment.

Acupuncture. Serious consideration must now be given to acupuncture as a physiological parapsychological approach to pain control and neurological treatment. Acupuncture theory is based on an invisible system of communication between various organs of the body that is distinct from the circulatory, nervous, and endocrine systems known to Western medicine. The few objective reports and available statistics are impressive. In one series, surgical anesthesia by acupuncture was "successful" in 90% of 1.500 patients undergoing a wide variety of operations, including thoracotomy, orthopedic procedures, and

craniotomy for brain tumor. Thyroidectomy has been performed on 504 consecutive cases with an anesthesia success rate of 98%. As a surgical anesthetic technique acupuncture has the advantage of convenience, great safety, stability of vital functions, no interruptions of patient hydration, no nausea or vomiting, and no postoperative respiratory complications. Pathological pains have also been controlled in cases of appendicitis, peptic ulcer, hepatic abscess, and renal biliary colic, and toothaches have been completely relieved by needling at the dorsum of the wrist (the "hoku" point).

In the tehcnique of acupuncture, slender needles of any metal type are twirled and moved vertically over selected points of more than 800 acupuncture sites. Recently the need for hand needling has been eliminated by attaching phasic electrical currents of 6 to 9 V at 105 cpm to the needles. In some cases, drugs such as morphine have been injected at the needle sites, and most patients receive small amounts of intravenous meperidine.

Patients apparently sense a "numbness, distension, heaviness and hotness" at specific sites in addition to a generalized raising of the thresholds of pain. Other remarkable physical effects include increased circulation time and transient leukocytosis. In experimental animals, profound shock has been reversed and sleep EEG patterns have been induced by needling.

The reason why each of the stimulation techniques is effective in reducing pain is still not clear, although two mechanisms are most likely. First, stimulation may selectively amplify the large inhibitory nerve fibers and suppress the effects of small fibers. Second, stimulation may act indirectly through the opiate receptor-descending control system. Objective evidence of this latter mechanism has come from recent experiments when analgesia induced inexperimental animals with either TNS or acupuncture was reversed when the animals were given the narcotic antagonist, naloxone.

Psychological approaches

Significant control over pain and other sensory complains may be gained through psychophysiological techniques such as relaxation therapy, biofeedback, hypnosis, and psychotherapy.

Many of the chronic, episodic pain syndromes such as myofascial pain dysfunction, vascular headaches, and atypical burning mucosa are known to be markedly influenced by the patient's response to stress. The techniques of *progressive relaxation* originally developed by Jacobson and the related techniques of *autogenous biofeedback* are now gaining wide acceptance as pain therapies. Biofeedback techniques have been especially useful for the pain of migraine and other vascular headaches in which the patient is trained to control cranial vascular dilation through a technique of hand temperature control. The widest use of biofeedback has been in the control of painful muscle spasms, however, using electromyogram (EMG) feedback. In these techniques, microvoltage signals from muscles such as the frontal or the masseter are used to train the patient in reducing tension and relieving muscle tenseness, clenching, bruxism, and other pain-producing habits. Hypnosis is another paraphysiological technique with primary action on pain tolerance thresholds that holds promise for the control of chronic facial pains. Carefully selected patients may, with training in autosuggestion, come to effectively deny or accept their pain.

Much of the psychological management of patients with neurological problems, especially pain, must be directed toward prevention. Curable pain should be relieved promptly and not allowed to become chronic because pain that persists longer than 6 months in a

neurotic individual often becomes too valuable in interpersonal relations togive up easily. Because there is known to be a higher incidence of maxillofacial pain in paranoic and psychopathic deviates, the clinician should remain alert for signs of personality disorders. In consultation with a clinical psychologist or psychiatrist, patients with susceptible personalities can be identified, often with the help of screening techniques such as the MMPI. Patients must not be allowed to become overly dependent on a given therapist. Demands for repetitive or irrelevant treatments should be tactfully opposed. It is often helpful to discuss openly with patients the nearly univeral role of psychological components in neurological problems, especially in pain disorders, so that counseling and psychotherapy can be included in the overall treatment.

Summary

It is obvious that maxillofacial neurological problems require a wide range of professional expertise. Future treatment may rely heavily on referral centers, computer-assisted diagnostic services, and multidisciplinary pain clinics to meet the challenges in this field. The dental clinician has an important role in developing and using these new approaches to manage maxillofacial pain.