B. RELATIVELY LOCALIZED SYNDROMES OF THE HEAD AND NECK

GROUP II: NEURALGIAS OF THE HEAD AND FACE

Trigeminal Neuralgia (Tic Douloureux) (II-1)

Definition
Sudden, usually unilateral, severe brief stabbing recurrent pains in the distribution of one or more branches of the Vth cranial nerve.

Site
Strictly limited to the distribution of the Vth nerve; unilateral in about 95% of the cases. Usually involves one branch; may involve two or, rarely, even all three branches. The second, third, and first branches of the Vth cranial nerve are involved in the foregoing order of frequency. The pain is more frequent on the right side.

System
Nervous system.

Main Features
Prevalence: relatively rare. *Incidence:* men 2.7, women
5.0 per 100,000 per annum in USA. Most patients have a lesion compressing the nerve where it leaves the brain stem. In patients with multiple sclerosis, there is also an increased incidence of tic douloureux. *Sex Ratio:* women affected perhaps more commonly than men. *Age of Onset:* after fourth decade, with peak onset in fifth to seventh decades; earlier onset does occur, but onset before age 30 is uncommon. *Pain Quality:* sharp, agonizing electric shock-like stabs or pain felt superficially in the skin or buccal mucosa, triggered by light mechanical contact from a more or less restricted site (trigger point or trigger zone), usually of brief duration-a few seconds (but reportedly occasionally up to 1-2 minutes followed by a refractory period of up to a few minutes. *Time Pattern:* paroxysms may occur at intervals or many times daily or, in rare instances, succeed one another almost continuously. Periodicity is characteristic, with episodes occurring for a few weeks to a month or two, followed by a pain-free interval of months or years and then recurrence of another bout. *Intensity:* extremely severe, probably one of the most intense of all acute pains.

Precipitation
Pain paroxysms can be triggered by trivial sensations from various trigger zones, that is, areas with increased sensitivity, which are located within the area of trigeminal innervation. The trigger phenomenon can be elicited by light touch, shaving, washing, chewing, etc.

Associated Symptoms and Signs
Occasionally, a mild flush may be noted during paroxysms. In true trigeminal neuralgia, apart from the trigger point, gross neurological examination is usually negative; in many patients, however, careful sensory testing to light touch will show a subtle sensory loss. No particular aggravating factors.

Relief
From carbamazepine, diphenyl hydantoin, and baclofen. If medical measures fail, radio-frequency treatment of the ganglion or microsurgical decompression of the trigeminal root are appropriate.

Usual Course

Recurrent bouts over months to years, interspersed with more or less prolonged asymptomatic phases.

Complications
Usually none. During exacerbations, nourishment may be a (transitory) problem.

Social and Physical Disability
Only as related to the recurrent pain episodes.

Pathology
When present, always involves the peripheral trigeminal (primary afferent) neuron. Impingement on the root by vascular loops, etc., appears to be the most common cause. Demyelination and hypermyelination on electron microscopy.

Essential Features
Unilateral, sudden, transient, intense paroxysms of superficially located pain, strictly confined to the distribution of one or more branches of the trigeminal nerve, usually precipitated by light mechanical activation of a trigger point. No sensory or reflex deficit detectable by routine neurologic testing.

Differential Diagnosis
Must be differentiated from symptomatic trigeminal neuralgia due to a small tumor such as an epidermoid or small meningioma involving either the root or the ganglion. Sensory and reflex deficits in the face may be detected in a significant proportion of such cases. Differential diagnosis between trigeminal neuralgia of mandibular division and glossopharyngeal neuralgia may, in rare instances, be difficult. Jabs and Jolts syndrome ("multiple jabs," "ice-pick pain"). SUNCT syndrome.

Code
006.X8a

References
Fromm GH, editor. The Medical and Surgical Management of Trigeminal Neuralgia. Mount Kisco, NY: Future Publishing Company; 1987.

Loeser JD. Tic douloureux and atypical face pain. In: Wall PD, Melzack R, editors. Textbook of Pain, 3rd ed. Edinburgh: Churchill Livingstone; 1994. p. 699–710.

Rovit RL, Murali R, Jannetta PJ. Trigeminal Neuralgia. Baltimore: Williams & Wilkins; 1990.

Secondary Neuralgia (Trigeminal) from Central Nervous System Lesions (11-2)

Definition
Sudden, severe, brief, stabbing recurrent pains in the distribution of one or more branches of the Vth cranial nerve, attributable to a recognized lesion such as tumor or aneurysm.

Site
Usually limited to distribution of trigeminal nerve.

System
Nervous system.

Main Features
Prevalence: rare; probably less than 2% of cases of tic douloureux. *Sex Ratio:* not remarkable. *Age of Onset:* corresponds to that of appearance of tumors. *Pain Quality:* paroxysmal pain may be

indistinguishable from "true" tic douloureux. Nonparoxysmal pain of dull or more constant type may occur. *Time Pattern:* may mimic tic douloureux. Attack pattern may be less typical with longer-lasting paroxysms or nonparoxysmal pain. *Intensity:* may be as severe as in tic douloureux. *Usual Duration:* indeterminate.

Associated Symptoms and Signs and Laboratory Findings
Sensory changes (hypoesthesia in trigeminal area) or loss of corneal reflex. Motor deficit is difficult to detect until late phase. X-ray, CAT scan, or MRI may reveal mass lesion in Meckel's cave or in pontine cistern.

Relief
Surgical intervention directed to the underlying cause. Occasionally, partial relief from drugs for "essential" trigeminal neuralgia.

Usual Course
Progression, usually very gradual.

Complications
Related to location of tumor.

Social and Physical Disability
Related to painful episodes and neurologic deficit when present.

Pathology
Meningioma of Meckel's cave, epidermoid cyst, and less frequently vascular malformation (arterio-venous aneurysm or tortuous basilar artery) of cerebello-pontine angle are among the most frequent causes of this rare condition.

Essential Features
Paroxysmal neuralgia in the trigeminal innervation zone, with one or more atypical features such as hyperesthesia or depression of corneal reflex, or longer-lasting paroxysms.

Differential Diagnosis
"Essential" trigeminal neuralgia.

Code

006.X4	Tumor
006.X0	Aneurysm
002.X2b code only	Arnold-Chiari syndrome: congenital;

References
Rovit RL, Murali R, Jannetta PJ. Trigeminal Neuralgia. Baltimore: Williams & Wilkins; 1990.

Secondary Trigeminal Neuralgia from Facial Trauma (11-3)

Definition
Chronic throbbing or burning pain with paroxysmal exacerbations in the distribution of a peripheral trigeminal nerve subsequent to injury.

Site
Maxillofacial region.

System
Nervous system.

Main Features
Prevalence: 5-10% following facial fractures; common after reconstructive orthognathic surgery; *1-5%* after removal of impacted teeth. *Pain Quality:* biphasic with sharp, triggered paroxysms and dull throbbing or burning background pain. *Occurrence:* constant with triggered episodes. *Intensity:* moderate. *Duration:* constant.

Signs
Tender palpable nodules over peripheral nerves; neurotrophic effects.

Usual Course
Progressive for six months, then stable until treated with microsurgery, graft-repair reanastomosis; transcutaneous stimulation and anticonvulsant pharmacotherapy.

Social and Physical Disabilities
Impaired mastication and speech.

Pathology
Neuromata; deafferentation, hypersensitivity.

Differential Diagnosis
Idiopathic trigeminal neuralgia, secondary trigeminal neuralgia from intracranial lesions, postherpetic neuralgia, odontalgia, musculoskeletal pain.

Code
006.X I

Acute Herpes Zoster (Trigeminal) (11-4)

Definition
Pain associated with acute herpetic lesions in the distribution of a branch or branches of the Vth cranial nerve.

Site
Face. Pain limited to distribution of trigeminal nerve (usually first division).

System
Trigeminal nerve.

Main Features
Prevalence: infrequent. *Sex Ratio:* not remarkable. *Age of Onset:* adults, more common in middle and old age. *Pain Quality:* burning, tingling pain with occasional lancinating components felt in the skin. *Time Pattern:* pain usually precedes the onset of herpetic eruption by one or two days (preherpetic neuralgia); may develop coincident with or after eruption. *Intensity:* severe. *Usual Duration:* one to several weeks.

Associated Symptoms
May be general malaise, low fever, headaches.

Signs and Laboratory Findings
Clusters of small cutaneous vesicles, almost invariably in the distribution of the ophthalmic distribution of the trigeminal. Frequently associated with lymphoma in treatment. Elevated protein and pleocytosis in spinal fluid.

Usual Course
Spontaneous and permanent remission. In the older age group, progression to chronic (postherpetic) neuralgia is not uncommon.

Complications
Acute glaucoma and corneal ulceration due to vesicles have been reported.

Social and Physical Disability
Related to cosmetic aspects and to pain.

Pathology
Small cell infiltrates in affected skin and bullous cutaneous changes. Similar infiltrates in ganglion and root entry zone.

Summary of Essential Features and Diagnostic Criteria
Herpetic vesicular eruption in distribution of first division of trigeminal nerve. History of burning pain in the perieruptive period.

Differential Diagnosis
Syndrome is usually unmistakable. Often related to impaired resistance, e.g., in the elderly or in the presence of carcinomatous metastases.

Code 002.x2a

Postherpetic Neuralgia (Trigeminal) (11-5)

Definition
Chronic pain with skin changes in the distribution of one or more roots of the Vth cranial nerve subsequent to acute herpes zoster.

Site
Face. Usually distribution of first (ophthalmic) division.

System
Trigeminal nerve.

Main Features
Prevalence: relatively infrequent. *Age of Onset:* sixth and later decades. *Sex Ratio:* more common in males. *Quality:* burning, tearing, itching dysesthesias and crawling dysesthesias in skin of affected area. Exacerbated by mechanical contact. *Time Pattern:* Constantly present with exacerbations. May last for years but spontaneous subsidence is not uncommon. *Intensity:* usually moderate, but constancy and intractability in many instances, contribute to intolerable nature of complaint. *Usual Duration:* months to

years.

Associated Symptoms
Depression, irritability.

Signs and Laboratory Findings
Cutaneous scarring, loss of normal pigmentation in area of earlier herpetic eruption. Hypoesthesia to touch, hypoalgesia, hyperesthesia to touch, and hyperpathia may occur.

Usual Course
Chronic, intractable, may last for years. Some cases "burn out" spontaneously.

Complications
None.

Social and Physical Disability
Severe impairment of most or all social activities due to constant pain. Suicide occasionally.

Pathology
Loss of many large fibers in affected sensory nerve. Chronic inflammatory changes in trigeminal ganglion and demyelination in root entry zone.

Summary of Essential Features and Diagnostic Criteria
Chronic burning, dysesthesias, paresthesias, and intractable cutaneous pain in distribution of the ophthalmic division of the trigeminal associated with cutaneous scarring and history of herpetic eruption in an elderly patient.
Differential Diagnosis
The syndrome is usually characteristic. Other conditions, e.g., metastatic carcinoma under treatment, may promote its occurrence.
Code 003.X2b

Geniculate Neuralgia (VIIth Cranial Nerve): Ramsay Hunt Syndrome (11-6)

Definition
Severe lancinating pains felt deeply in external auditory canal subsequent to an attack of acute herpes zoster.

Site
External auditory meatus with retroauricular radiation.

System
The sensory fibers of the facial nerve.

Main Features
Prevalence: rare; few cases in world literature. *Sex ratio:* no data. *Pain Quality:* sharp, lancinating, shocklike pains felt deeply in external auditory canal. *Intensity:* severe.

Signs and Laboratory Findings
Usually follows an eruption of herpetic vesicles which appear in the concha and over the mastoid.

Complications
None.

Social and Physical Disability
Only as related to the pain episodes.

Pathology
No reported case with pathological examination.

Summary of Essential Features and Diagnostic Criteria
Onset of lancinating pain in external meatus several days to a week or so after herpetic eruption on concha.

Differential Diagnosis
Differentiate from otic variety of glossopharyngeal neuralgia, which does not have herpetic prodromata.

Code 006.X2

Neuralgia of the Nervus Intermedius (11-7)

Note: This condition is admittedly very rare and is presented as a tentative category about which there is still some controversy.

Definition
Sudden, unilateral, severe, brief, stabbing, recurrent pain in the distribution of the nervus intermedius.

Site
In ear canal, deep in ear, and in posterior pharynx.

System
Nervous system.

Main Features
Prevalence: very rare. Probably .03 per 100,000 per annum in USA. *Sex Ratio:* women equal to men. *Age of Onset:* fifth to seventh decade most common. *Pain Quality:* sharp agonizing electric shock-like stabs of pain felt in the ear canal, middle ear, or posterior pharynx, usually of brief duration, often with a refractory period after multiple jabs of pain. *Time Pattern:* paroxysms may occur at intervals or may occur in a brief flurry.

Periodicity is characteristic, with episodes occurring for weeks or months, and then months or years without any pain. *Intensity:* extremely severe; probably one of the most intense of all acute pains.

Precipitation
Pain paroxysms can be triggered by non-noxious stimulation from the posterior pharynx or ear canal.

Associated Signs and Symptoms
None.

Relief

From carbamazepine and baclofen. Or from surgical procedures: microsurgical decompression of the nervus intermedius or section of the nerve.

Usual Course
Recurrent bouts over months to years, interspersed with asymptomatic phases.

Complications
Usually none.

Social and Physical Disability
Related to recurrent pain episodes.

Pathology
Most patients have impingement on the nervus intermedius at its root entry zone.

Essential Features
Unilateral, sudden, transient, intense paroxysms of electric shock-like pain in the ear or posterior pharynx. No sensory or motor deficit is detectable by routine neurologic testing.

Differential Diagnosis
Must be differentiated from tic douloureux involving the Vth nerve, glossopharyngeal neuralgia, and geniculate neuralgia of the VIIth nerve due to herpes zoster.

Code
006.X8c

Reference
Furlow LP. Tic douloureux of the nervus intermedius. JAMA 1942;119:255

Glossopharyngeal Neuralgia (IXth Cranial Nerve) (11-8)

Definition
Sudden severe brief stabbing recurrent pains in the distribution of the glossopharyngeal nerve.

Site
Tonsillar fossa and adjacent area of fauces. Radiation to external auditory canal (otic variety) or to neck (cervical variety).

System
Peripheral and central mechanisms involving glossopharyngeal nerve fibers.

Main Features
Prevalence: very rare. *Incidence: 0.5* per 100,000 per annum in USA. Sharp, stabbing bouts of severe pain, often triggered by mechanical contact with faucial area on one side, also by swallowing and by ingestion of cold or acid fluids. *Pain Quality:* sharp, stabbing bursts of high-intensity pain, felt deep in throat or ear. *Time Pattern:* episodic bouts occurring spontaneously several times daily or triggered by any of above mentioned stimuli. *Intensity:* very severe, interferes with eating. *Usual Duration:* episodes last for weeks to a month or two and subside spontaneously. Tendency to recurrence is common.

Associated Symptoms

Cardiac arrhythmia and syncope may occur during paroxysms in some cases.

Signs and Laboratory Findings
The important and only sign is the presence of a trigger point, usually on fauces or tonsil; sometimes it may be absent.

Usual Course
Fluctuating; bouts of pain interspersed by prolonged asymptomatic periods.

Complications
Transitory cardiac arrhythmias, as noted.

Social and Physical Disability
Only as related to pain episodes.

Pathology
Unknown. Vascular loops impinging on roots may be a cause.

Summary of Essential Features and Diagnostic Criteria
Paroxysmal bursts of sharp, lancinating pain, spontaneous or evoked by mechanical stimulation of tonsillar area, often with radiation to external ear or to angle of jaw and adjacent neck. Application of local anesthetic to tonsil (or trigger point) relieves pain.

Differential Diagnosis
Usually characteristic syndrome. May be confused with trigeminal neuralgia limited to mandibular division.

Code
006.X8b

Neuralgia of the Superior Laryngeal Nerve (Vagus Nerve Neuralgia) (11-9)

Definition
Paroxysms of unilateral lancinating pain radiating from the side of the thyroid cartilage or pyriform sinus to the angle of the jaw and occasionally to the ear.

Site
Unilateral, possibly more on the left in the neck from the side of the thyroid cartilage or pyriform sinus to the angle of the jaw and occasionally to the ear.

System
Nervous system.

Main Features
Prevalence: rare. May be a variant of glossopharyngeal neuralgia, which has also been called vago-glossopharyngeal neuralgia. Combined ratio of vagoglossopharyngeal neuralgia to trigeminal neuralgia is about 1:80. *Sex Ratio:* about equal. *Pain Quality:* usually severe, lancinating pain often precipitated by talking, swallowing, coughing, yawning, or stimulation of the nerve at its point of entrance into the larynx. Mild forms do occur.

Associated Symptoms
Local tenderness. Possibly autonomic phenomena, e.g., salivation, flushing; possibly tinnitus and vertigo.

Signs
Presence of a trigger zone.

Laboratory Findings
None.

Relief
Relief from analgesic nerve block, alcohol nerve block, or nerve section.

Complications
Loss of weight.

Social and Physical Disability
As related to pain episodes.

Pathology
There may be a history of local infection. A large styloid process or calcified stylohyoid ligament may be contributory (cf. Eagle's syndrome).

Essential Features
Sudden attacks of unilateral lancinating pain in the area of the thyroid cartilage radiating to the angle of the jaw and occasionally to the ear.

Differential Diagnosis
Glossopharyngeal neuralgia, carotidynia, local lesions, e.g., carcinoma.

Code
006.X8e

Occipital Neuralgia (11-10)

Definition
Pain, usually deep and aching, in the distribution of the second cervical dorsal root.

Site
Suboccipital area, unilateral in the second cervical root distribution from occiput to vertex. May radiate still farther forward, see below.

System
Nervous system.

Main Features
Prevalence: quite common; no epidemiological data; most often follows acceleration-deceleration injuries. *Sex Ratio:* women more frequently affected, but statistical data lacking. *Age of Onset:* from second decade to old age; more common in third to fifth decades. *Pain Quality:* deep, aching, pressure pain in suboccipital area, sometimes stabbing also. Unilateral usually; may radiate toward vertex or to fronto-orbital area and/or face. *Time Pattern:* irregular, usually worse later in day. *Intensity:* from moderate to severe.

Associated Symptoms
Hyperesthesia of scalp. A variety of symptoms such as vertigo, tinnitus, tears, etc., have been described in some cases, but these are probably transitional forms to cluster headache. Nerve block may give effective relief.

Signs and Laboratory Findings
Diminished sensation to pinprick in area of C2 and tenderness of great occipital nerve may be found.

Usual Course
Chronic, recurrent episodes. May cease spontaneously on occasion.

Complications
None.

Social and Physical Disability
Only as related to pain episodes.

Pathology
Unknown. Perhaps related to increased muscle activity in cervical muscles. May be secondary to trauma, including flexion-extension (whiplash) injury.

Summary of Essential Features and Diagnostic Criteria
Intermittent episodes of deep, aching, and sometimes stabbing pain in suboccipital area on one side. Marked tendency to chronicity. Often associated with tender posterior cervical muscles. Can be bilateral.

Differential Diagnosis
Cluster headaches, posterior fossa and high cervical tumor, herniated cervical disk, uncomplicated flexionextension injury, metastatic neoplasm at the base of the skull.

Code
004.X8 or
004.X1 (if subsequent to trauma)

References
Behrman S. Traumatic neuropathy of second cervical spinal nerves Br Med J 1983;286:1312–3.

Hypoglossal Neuralgia (II-11)

Code
006.X8

Glossopharyngeal Pain from Trauma (11-12)

Code
003.X1a

Hypoglossal Pain from Trauma (1113)

Code
003.Xlb

Tolosa-Hunt Syndrome (Painful Ophthalmoplegia) (11-14)

Definition
Episodes of unilateral pain in the ocular and periocular area combined with ipsilateral paresis of oculomotor nerves (ophthalmoplegia) and of the first branch of the Vth cranial nerve. The episodes are most often circumscribed in time, but may be repetitive.

Site
Unilateral; ocular and periocular area.

System Involved
Peripheral nervous and autonomic nervous systems.

Main Features
Prevalence: rare. Sex Ratio: no sex preponderance. Age of Onset: generally in adults; mean age of onset, around 40. Pain Quality: pain usually precedes the ophthalmoplegia. Continuous moderate to severe ache in the ocular and periocular area or behind the eye, no triggering. Time Pattern: episodes last weeks or months with a continuous or intermittent pattern. Recurrences with several such episodes may occur. Intensity: moderate to severe. Usual Duration: untreated 8.4 ± 7.4 weeks (mean ± SD), according to Bruyn and Hoes (1986).

Precipitating Factors
Not known.

Associated Symptoms and Signs
Frequently strabismus. Affection of various cranial nerves, i.e., numbers III, IV, V, and VI, either alone or in various combinations. The pupil is usually spared. Nausea and vomiting are rare.

Laboratory Findings
Orbital phlebography renders positive findings in approximately 60-65% of cases. Such findings are: thin caliber, segmental narrowing, and even occlusion and opening of new vessels. Such changes are particularly present in the so-called third segment of the ophthalmic vein and in the cavernous sinus. The pathology of these findings has not been adequately demonstrated. Oculomotor nerve palsy can be ophthalmologically verified.

Relief
From corticosteroids.

Usual Course
Self-limiting, but at times rather protracted. There may be a solitary episode or a tendency to recurrence. Milder forms apparently exist; during recurrences in particular, the pattern may be less characteristic. Occasionally, residual symptoms are found.

Social and Physical Disability
As related to pain episodes.

Pathology
Fibrous tissue formation in cavernous sinus area, involving various structures, vein wall, etc.

Essential Features
Coexistence of orbital and periorbital pain and ophthalmoplegia on the same side.

Differential Diagnosis
Raeder's paratrigeminal syndrome, ophthalmoplegic migraine, other rare cavernous sinus syndromes, symptomatic forms (e.g., tumors of the orbit or base of the brain).

Code
002.X3a

References
Bruyn GW, Hoes MJ. The Tolosa-Hunt syndrome. In: Vinken PG, Bruyn GW, Klawans HL, Rose FC, editors. Handbook of Clinical Neurology 48. Amsterdam: Elsevier; 1986. p. 291–307.
Hannerz J, Ericson K, Bergstrand G. Orbital phlebography in patients with Tolosa-Hunt syndrome in comparison with normal subject. Acta Radiol (Diagn) 1984;1125:457–63.

Hunt WE, Meagher JN, LeFever HE, Zeman W. Painful ophthalmoplegia: its relation to indolent inflammation of the cavernous sinus. Neurology 1961;11:56–62.

Tolosa E. Periarteritic lesions of the carotid siphon with the clinical features of a carotid infraclinoidal aneurysm. J Neurol Neurosurg Psychiatry 1965;17:300–2.

SUNCT Syndrome (Shortlasting, Unilateral Neuralgiform Pain with Conjunctival Injection and Tearing) (11-15)

Definition
Repetitive paroxysms of unilateral short-lasting pain usually 15-120 seconds duration, mainly in the ocular and periocular area, of a neuralgiform nature and moderate to severe intensity, usually appearing only during daytime and accompanied by ipsilateral marked conjunctival injection, lacrimation, a low to moderate degree of rhinorrhea, and (subclinical) forehead sweating. SUNCT is not responsive to indomethacin or carbamazepine, and has, so far, mostly been observed in males.

Site
The ocular and periocular area, occasionally with spread to the fronto-temporal area, upper jaw, or roof of the mouth. The headache is generally strictly unilateral without change of sides, but cases with an accompanying late stage and moderate involvement of the opposite side have been observed.

System
Not firmly identified. The pain appears neurogenic, but there is also involvement of vascular factors.

Main Features
Prevalence: probably rare. *Sex Ratio: so* far, mostly males. *Age of Onset:* middle to old age. *Pain Quality:* the onset is abrupt, the discontinuation of the attack may be a little more gradual. Occasionally, some slight interparoxysmal discomfort occurs. The pain is steady and nonpulsating. Attacks may be *triggered* by various types of minor stimuli within the innervation zone of the Vth cranial nerve but also

by neck movements. *Time Pattern:* the attack frequency varies much. In circumscribed periods lasting weeks to months, there may be many attacks per hour, at other times only a few per day or even less. Attacks are short-lasting, i.e., 15-120 seconds duration. Remissions last from months to years. In the early stages, attacks appear in bouts; eventually, a chronic course develops. No neurological deficits. *Intensity:* Moderate to severe pain.

Precipitating Factors
Attacks may be triggered by minor stimuli within the distribution of the Vth cranial nerve, but also partly by neck movements.

Associated Symptoms and Signs
Conjunctival injection, lacrimation, nasal stuffiness, and to a lesser extent, rhinorrhea and forehead sweating (which is apparently always subclinical) occur on the pain side. The onset of the conjunctival injection and lacrimation may have an almost explosive character during severe attacks.

Relief
No benefit from indomethacin or carbamazepine. No really effective treatment is yet available. Cortisone may possibly be of some avail.

Usual Course
At an early stage, an intermittent pattern which may or may not be permanent.

Social and Physical Disability
During the worst periods, some patients cannot do their ordinary work.

Pathology
Unknown.

Essential Features
Shortlasting, unilateral paroxysms of ocular pain, associated with ipsilateral autonomic phenomena like conjunctival injection, lacrimation, etc. In some cases, attacks may be triggered mechanically. Male preponderance.

Differential Diagnosis
Trigeminal neuralgia, Syndrome of "Jabs and Jolts" ("multiple jabs"), chronic paroxysmal hemicrania, cluster headache, "symptomatic SUNCT," Newton-Hoyt-Taniguchi syndrome.

Code
006.X8j

References
Bussone G, Leone M, Dalla Volta G, Strada L, Gasparotti R, Di Mona V. Shortlasting, unilateral neuralgiform headache attacks with tearing and conjunctival injection: the first "symptomatic" case? Cephalalgia 1991;11:123–7.

Sjaastad O, Saunte C, Salvesen R, Fredriksen TA, Seim A, Roe OD, Fostad K, Lobben OP, Zhao JM. Shortlasting, unilateral neuralgiform headache attacks with conjunctival injection, tearing, sweating, and rhinorrhea. Cephalalgia 1989;9:147–56.

Taniguchi RM, Goree JA, Odom GL. Spontaneous carotid-cavernous shunts presenting diagnostic problems. J Neurosurg 1971;35:384–91.

Raeder's Syndrome (Raeder's Para-trigeminal Syndrome) (11-16)

Definition
Homer's syndrome of the IIIrd cranial nerve type combined with aching steady pain in the ocular and periocular area, with or without parasellar cranial nerve involvement; the Vth nerve is most often involved, but also the IInd, IIIrd, IVth, and VIth cranial nerves may be affected, all on one side. The cases with and without parasellar cranial nerve involvement have been placed in two groups, I and II, respectively (Boniuk and Schlezinger 1962). Sweating is reduced on the symptomatic side in IIIrd nerve disorders, including Raeder's syndrome, but apparently only in the medial part of the forehead (corresponding to the sympathetic fibers that follow the internal carotid and ultimately perhaps the supraorbital arteries).

Site
Unilateral pain in the ocular and periocular area, temporal and aural areas, forehead, and occasionally also the anterior vertex.

System
Autonomic nervous system. Cranial nerves.

Main Features
Prevalence: rare. *Clinical Patterns:* two forms have been described: (I) the original form (described by the Norwegian ophthalmologist Raeder [1924]) with para-sellar cranial nerve involvement and (II) a form without parasellar nerve involvement (Boniuk and Schlezinger 1962). If parasellar cranial nerve involvement is no longer an obligatory diagnostic requirement, then the localization of the underlying disorder no longer has to be the "paratrigeminal" space: It can be anywhere from the superior cervical ganglion and its rostral connections and toward the periphery. Many of the Boniuk and and Schlezinger type cases, nevertheless, probably originate in or close to the area of pathology of type I cases. *Sex Ratio:* almost only males. A few acceptable female cases have been reported. *Age of Onset:* usually middle-old age. *Pain Quality:* the pain is not excruciating, nor is it continuously severe. It rather fluctuates between the moderate and severe stages. At times, it attains the character of an attack, frequently in the early and late stages; the pain is generally aching and nonpulsatile. *Time Pattern:* there is a relatively longlasting period of moderate to severe pain with a crescendo, a plateau, and a declining phase, and this period may or may not have been preceded by a longlasting phase or rare and/or mild headaches. The period of severe pain usually lasts for weeks to months, after which time there may be a period of lingering pain. There is only a little tendency to recurrence. *Intensity:* moderate to severe; fluctuating.

Precipitating Factors
Possibly cardiovascular factors may predispose.

Associated Symptoms and Signs
Ptosis (of a mild degree), miosis, and hypohidrosis in the medial part of the forehead (but no enophthalmus) on the symptomatic side. There are no further findings in Boniuk and Schlezinger type II. In type I cases, involvement of the IInd, IIIrd, IVth, Vth, and VIth cranial nerves appears in various combinations; most frequently the Vth nerve is affected. Cases with only *a discrete* affection (hypoesthesia, dysesthesia) of the Vth nerve (first branch) seem to be the most common type.

Relief
Group I cases may need surgery for the causal condition. Group II cases benefit from analgesics. No specific therapy is known at present and no special benefit occurs with indomethacin. Whether cortisone acts beneficially (as in the Tolosa-Hunt syndrome) is not adequately documented.

Duration and Usual Course
In most cases there is a circumscribed, self-limiting headache, lasting some weeks to months. In the

occasional case, such periods may be repeated one or more times. Group II cases have a good prognosis and may not need extensive investigation. Group I cases differ from Group II cases from a diagnostic and prognostic point of view because the underlying pathology may be a tumor, skull fracture, etc. Such cases are in need of thorough investigation.

Complications
Type I: from the paratrigeminal tumor (or other pathology).

Social and Physical Disability
During the acute stage the incapacity may be considerable.

Pathology
Type I: tumor or other (serious) pathology paratrigeminally until proven otherwise. Type II: not known.

Essential Features
Painful, type III Homer's syndrome-unilaterally, with or without parasellar II-VI cranial nerve affection; the involvement of the Vth cranial nerve is usually *discrete.*

Differential Diagnosis
The Tolosa-Hunt syndrome. Other cavernous sinus syndromes, cluster headache. Hemicrania continua is also a diagnostic possibility; hypothetically, orbital space-occupying disorders (but they hardly give rise to Horner's syndrome).

Code

Type I:	002.X4 Tumor
	002.Xla Trauma
	002.X3b Inflammatory, etc.
Type II:	002.X8 Unknown

References

Boniuk M, Schlezinger NS. Raeder's paratrigeminal syndrome, Am J Ophthalmol 1962;54:1074–84.

Raeder, J.G., Paratrigeminal paralysis of oculo-pupillary sympathetic, Brain, 47 (1924) 149-158.

Salvesen R, de Souza Carvalho D, Sjaastad O. Homer's syndrome: sweat gland and pupillary responsiveness in two cases with a probable 3rd nerve dysfunction. Cephalalgia 1989;9:63–70.

GROUP III: CRANIOFACIAL PAIN OF MUSCULOSKELETAL ORIGIN

Acute Tension Headache (III-1)

Definition
Acute, relatively shortlasting, diffuse (or more localized) dull head pain related to anxiety, depression, or emotional tension.

Main Features
As for presumed chronic tension headache except as follows: Very frequent. Affects the majority of the population. Sex ratio probably equal. The pain is dull— sometimes somewhat more marked—bilateral, and non-throbbing, with gradual onset, steady rise, plateau, and then a decline in intensity. No nausea, vomiting, or photophobia. Does not usually need any therapy, unless the pattern is repetitive.

Acute, self-limiting, relatively shortlasting (for a few hours or less); repeated separate attacks with very varying frequency. Eases with the elimination of the (acute) cause.

Pathology
In spite of the fact that it shares the appellation with the chronic variety, it may well be that the two forms differ in more than just temporal and intensity respects. They may be different types of headache.

Code
034.X7a

References
Kudrow L. Muscle contraction headaches. In: Vinken PJ, Bruyn GW, Klawans HL, Rose FC, editors. Handbook of Clinical Neurology 48. Amsterdam: Elsevier; 1986, p. 343–52.

Tension Headache: Chronic Form (Scalp Muscle Contraction Headache) (111-2)

Definition
Virtually continuous, dull aching head pain, usually symmetrical and frequently global. This headache is frequently, but not in all cases, associated with muscle "tension." The term *tension* is, nevertheless, retained; tension may also be taken to indicate stress, strain, anxiety, and emotional tension. There is a frequent association between these factors and also depressive states and this headache. In the later stages, exacerbations with a tinge of pounding headache and with nausea (and, less typically, vomiting) may occasionally occur, although less typically and with less intensity than in common migraine.

Site
Frontal, orbital, fronto-occipital, occipital, nuchal, or whole scalp area. Diffuse or bandlike, usually bilateral, sometimes more on one side during exacerbations.

System
Not known. Possibly musculoskeletal, possibly central nervous system, or vessels.

Main Features
Prevalence: Often diagnosed; even approximate prevalence is unknown, mainly because of lack of precise diagnostic criteria. *Sex Ratio:* Females more than males; ratio approximately 4:1 in those who consult their physician. *Onset:* From age 8 onward, usually before age 30.

Start: Gradual emergence as mild, diffuse ache or un- pleasant feeling, intermittent at first, increasing with time to a more definite pain that gradually will become more and more chronic. Fluctuation during the day is typical. In a proportion of cases, exacerbations with additional symptoms seem to emerge after several years of lesser headache. *Occurrence and Duration:* Every day or most days, for most of the day. Occasionally, in long-standing severe cases, pain may wake the patient from sleep. *Pain Quality: Dull* ache, usually does not throb, more severe during exacerbations, and then occasionally with throbbing. Some describe tight band feeling or gripping headache.

Precipitants and Exacerbating Factors
Emotional stress, anxiety and depression, physical exercise, alcohol (which may also have the opposite effect).

Associated Symptoms
Many patients are hypersensitive and have anxiety, depression, or both. Irritability, nausea, vomiting, photophobia, phonophobia, and pulsatile pain may occur during exacerbations in the later stages of this headache. Vomiting is, however, most unusual.

Signs
Muscle tenderness of the pericranial and/or nuchal muscles occurs but may also be found in other conditions and in healthy individuals. Tension headache with and without muscle tenderness may differ both from a pathogenetic and from a therapeutic point of view (e.g., with regard to response to tricyclic antidepressants).

Relief
Treatment of emotional problems, anxiety, or depression may diminish symptoms. Relaxation and biofeedback treatment help. Anxiolytics may help but should generally be avoided since some patients become depressed and others develop dependence. Tricyclic antidepressants are frequently very useful, but their effect may possibly differ in patients with and without muscular tenderness. Some of them, e.g., amitriptyline, have independent analgesic properties which may account for some of their usefulness. Analgesics help only a little, and discontinuation of some chronically used drugs may be of some avail.

Usual Course
Chronic course. Hard to treat in most cases.

Complications
Analgesics, narcotics, and other drug abuse. Detoxification is often mandatory in order to improve the situation and end a vicious circle of withdrawal headaches and medication.

Social and Physical Disability
Reduction of activities and of work.

Pathology
Unsettled. Evidence of chronic muscle tenderness in many cases. Apparently, there is increased muscle activity, sometimes demonstrable on EMG. Both phenomena may, however, also be present in patients with migraine. There is a lack of adequate, long-term studies comparing controls with patients, in particular after appropriate exposures.

Essential Features
Bilateral, usually low-grade to mild, more or less chronic headache, with fewer accompanying features than in common migraine, starting early in life, and occurring much more frequently in the female.

Differential Diagnosis
Mainly from other *bilateral* headaches. Multiple attacks of acute tension headache, which *may* be an altogether different headache, may masquerade as chronic tension headache. Common migraine, "mixed vascular-tension headache," chronic abuse of analgesics, refractive errors, heterophoria ("eye strain"), post-traumatic headache (bilateral cases, which probably exist), cervicogenic headache (in the bilateral cases, that sooner or later may be recognized as being characteristic of this disorder), cervical spine disorders, depression, conversion hysteria, and hallucinatory headache.

The differential diagnosis vs. common migraine is particularly challenging. The occurrence of migraine or migraine-like headache in the close family, the frequently occurring unilaterality (with *change of side),* the "anterior" onset of attacks (or exacerbations), the more marked degree of nausea, vomiting, photophobia, and phonophobia, and pulsating headache, all in common migraine, are factors of crucial importance in distinguishing the two headaches. The ergotamine effect (and probably also the sumatriptan effect) is also clearly more marked in common migraine.

Code
033.X7c

References
Friedman AP, von Storch T, Merritt HH. Migraine and tension headaches: a clinical study of two thousand cases. Neurology 1954;4:773–4.

Kudrow L. Paradoxical effects of frequent analgesic use. In: Critchley M, Friedman AK, Gorini S, Sicuteri F, editors. Advances in Neurology. New York: Raven Press; 1982. p. 335–41.

Pfaffenrath V, Wermuth A, Pollmann W. Der Spannungskopfschmerz: eine Ubersicht, Fortschr. Neurol Psychiat 1988;56:403–18.

Ziegler DK, Hassanein RS. Migraine muscle-contraction dichotomy studied by statistical analysis of headache symptoms. In: Rose FC, editor. Advances in Migraine Research and Therapy. New York: Raven Press; 1982.

Temporomandibular Pain and Dysfunction Syndrome (111-3)
(also called Temporomandibular Joint Disorder)

Definition
Aching in the muscles of mastication, sometimes with an occasional brief severe pain on chewing, often associated with restricted jaw movement and clicking or popping sounds.

Site
Temporomandibular, intra-auricular, temporal, occipital, masseteric, neck, and shoulder regions.

System
Musculoskeletal system.

Main Features
Prevalence: unknown. Epidemiological studies have shown that up to 10% of people between the ages of 15 and 35 experience clicking of the jaw with dysfunction at some point in time. *Sex Ratio:* most patients are female. *Age of Onset:* patients presenting with temporomandibular pain and dysfunction have an age range of 560 years. *Pain Quality:* the pain is usually described as intermittent, unilateral, dull, and aching, but can be constant. The pain is often exacerbated by jaw movement, e.g., chewing hard food or yawning. Combinations of aching and severe exacerbations may also occur. *Time Pattern:* the pain may be

continuous by day or brief. It is often worse on waking. *Duration:* symptoms can persist for years with fluctuations.

Clicking of the joint or popping noises in the ears are frequently present. Limitations of opening, deviation of the jaw on opening, and a feeling that the teeth do not meet together properly are common.

Signs
Restricted mandibular opening with or without deviation of the jaw to the affected side on opening; tenderness to palpation of the muscles of mastication; clicking or popping at the joint on auscultation or palpation; changes in the ability to occlude the teeth fully.

Imaging
Normal temporomandibular joint radiographic structure, variable disk displacement seen on arthrography, occasional osteoarthritic changes. Magnetic resonance imaging may show disk displacement with or without reducibility. The clinical significance of disk displacement and its relationship to the syndrome are not established.

Usual Course
Variable. Because of its fluctuating course, the response to treatment is difficult to evaluate. Psychosocial factors account for a significant portion of the outcome. The effectiveness of common treatments, e.g., occlusal splints and psychotherapy, has not been shown to be superior to placebo. A high potential for morbidity makes TM joint surgery problematic.

With conservative treatment, many patients are kept reasonably comfortable and productive. Long-term outcome studies are unavailable. However, small sample studies indicate that many experience symptoms indefinitely.

Complications
Possible degenerative joint disease, depression and anxiety, drug dependence. In some intractable cases widespread diffuse aching facial pain develops.

Social and Physical Disability
Interference with mastication and social and vocational activity, development of secondary psychological changes.

Pathology and Etiology
Muscle spasm appears in most cases. Disk displacement with or without reducibility appears in some cases. The etiology is unknown. Psychological stress and bruxism are widely believed to be contributory factors, although evidence for this is lacking. Trauma is known to be related to a minority of cases.

Summary of Essential Features and Diagnostic Criteria
Muscle tenderness; temporomandibular joint clicking; difficulty in opening the jaw and sometimes deviation on opening; a dull ache or severe episodes associated with jaw opening, or both.

Differential Diagnosis
Degenerative joint disease, rheumatoid arthritis, traumatic arthralgia, temporal arteritis, otitis media, parotitis, mandibular osteomyelitis, stylohyoid process syndrome, deafferentation pains, pain of psychological origin.

Code
034.X8a

References
Griffiths RH. Report of the president's conference on the examination, diagnosis and management of temporomandibular disorders. Laskin D, Greenfield W, Gale E, Rugh J, Neff P, Ailing C, Ayer WA, editors. J Am Dent Assoc 1983;166:75–7.

Marbach JJ, Lennon MC, Dohrenwend BP. Candidate risk factors for temporomandibular pain and dysfunction syndrome: psychosocial, health behavior, physical illness and injury. Pain 1988;34:139–47.

Rudy TE, Turk DC, Zaki HS, Curtin HB. An empirical taxometric alternative to traditional classification of temporomandibular disorders, Pain 1989;36:311–20.

Schnurr RF, Brooke RI, Rollman GB. Psychosocial correlates of temporomandibular joint pain and dysfunction. Pain 1990;42:153.

Osteoarthritis of the Temporomandibular Joint (111-4)

Code
033.X6

Rheumatoid Arthritis of the Temporomandibular Joint (111-5)

Definition
Part of the systemic disorder of rheumatoid arthritis with granulation tissue proliferating onto the articular surface.

Site
Temporomandibular joint, external acoustic meatus.

System
Musculoskeletal system.

Main Features
Prevalence: Caucasian, approximately 50% occurrence with general rheumatoid arthritis. *Sex Ratio:* female predilection. *Age of Onset:* juvenile or pubertal; adult onset 40-60 years. *Start:* spontaneous onset. *Pain Quality:* boring, penetrating, aching. *Occurrence:* constant with diurnal variation. *Intensity:* moderate A.M., mild P.M. *Duration:* minutes to hours.

Signs
Preauricular erythema, crepitus, tenderness of external acoustic meatus, restriction and deformation of other joints, open bite eventually.

Laboratory and Radiological Findings
Positive latex fixation, radiographic joint space narrowing.

Usual Course
Five to nine months acute painful course followed by joint restriction and ankylosis; responsive to condyloplasty without recurrence.

Relief
Heat, joint physiotherapy, anti-inflammatory agents.

Complication
Fibrous or bony ankylosis.

Social and Physical Disability
Mastication impairment, associated orthopedic restrictions.

Pathology
Synovitis, foam cell degeneration ("Pannus Cell" formation), secondary resorption of the articular surfaces, adhesions to the articular disk, fibrous adhesions, narrowing and loss of joint space.

Diagnostic Criteria
Multiple joint involvement, radiographic joint space loss and condylar deformation, positive lab findings.

Differential Diagnosis
Includes degenerative joint disease, traumatic arthritis, inflammatory arthritis, myofascial pain dysfunction.

Code
032.X3b

Dystonic Disorders, Facial Dyskinesia (111-6)

Code
003.X8

Crushing Injury of Head or Face (111-7)

Code
032.X1

GROUP IV: LESIONS OF THE EAR, NOSE, AND ORAL CAVITY

Maxillary Sinusitis (IV-1)

Definition
Constant burning pain with zygomatic and dental tenderness from inflammation of the maxillary sinus.

Site
Upper cheek and sometimes teeth.

System
Respiratory system.

Main Features
Prevalence: common. *Sex Ratio:* no difference. *Age of Onset:* usually adults. *Onset:* spontaneous. *Pain Quality:* dull ache, unilaterally or bilaterally, sense of fullness and tenderness in the overlying cheek. *Occurrence:* usually associated with nasal cold. Other nasal disease or dental disease causes 20% of cases. *Intensity:* mild to severe. *Duration:* days.

The illness develops when swelling of the nasal mucosa blocks the ostium so that drainage can no longer occur into the nose. When the cause is a common cold, the other nasal sinuses may become involved. Dental cases arise from infection associated with the apex of one of the molar or premolar teeth. They may also be associated with operative procedures including a tooth root being pushed accidentally into the sinus during extraction, or endodontic instruments and materials being pushed too far. In chronic cases there may be no pain or only mild, diffuse discomfort from time to time.

Signs
Zygomatic area of face may be slightly flushed and slightly swollen ("puffy"). Pain exacerbated by bending. Tenderness of upper molar and premolar teeth and over cheek.

Laboratory Findings
Radiography may show fluid level or a tooth root. In chronic cases radiographic examination reveals a sinus more opaque than normal.

Usual Course
Subsides in several days to a week.

Relief
Analgesics, sometimes with drainage by lying down on the opposite side.

Pathology
Inflammation of the lining of the maxillary sinus.

Diagnostic Criteria
Pain or discomfort over the maxillary antrum coupled with evidence of infection such as local inflammation, radiographic signs of thickening or a fluid level, and relief by antibiotics or drainage.

Differential Diagnosis
Periapical disease of the associated teeth, malignant disease.

Code
031.X2a.

Odontalgia: Toothache 1. Due to Dentino-Enamel Defects (IV-2)

Definition
Shortlasting diffuse orofacial pain due to dentino-enamel defects and evoked by local stimuli.

Site
Orofacial pain.

System
Musculoskeletal system.

Main Features
Prevalence: extremely common. *Sex Ratio:* no difference. *Age of Onset:* 2 years of age to any age. *Start:* stimulus evoked, not spontaneous, heat, cold, mechanical, osmotic. *Pain Quality:* bright to dull. *Occurrence:* intermittent. *Intensity:* mild to moderate. *Duration:* less than a second to minutes.

Signs
Dental caries, fracture, crack, or lost restoration.

Laboratory and Radiological Findings
Radiographic evidence of caries.

Usual Course
If neglected, there may be mineralization within the dentine, resulting in less frequent pain or no pain; or pulpal involvement.

Relief
By protecting defective area with a dressing or restoration.

Complications
Laceration of soft tissues by sharp edge of tooth.

Pathology
Dental caries, trauma, operative procedures.

Diagnostic Criteria
Visually observed defects, or defects palpated with a probe, plus radiographic examination.

Differential Diagnosis
Pulp disease, periapical disease.

Code
034.X2b

Odontalgia: Toothache 2. Pulpitis (IV-3)

Definition
Orofacial pain due to pulpal inflammation, often evoked by local stimuli.

Site
Face, jaw, mouth.

System
Musculoskeletal system.

Main Features
Prevalence: very common. *Sex Ratio:* no difference. *Age of Onset:* after eruption of teeth. *Start:* milder cases may be started by hot or cold stimuli. In severe cases may be spontaneous (no external stimulus needed) but is exacerbated by heat and cold stimuli. *Pain Quality:* sharp or dull ache, may throb. *Occurrence:* with food intake in milder cases. Daily until treated in severe cases. *Intensity:* can be moderate, usually severe. *Duration:* bouts lasting minutes or hours; may continue for days.

Signs
Deep dental caries, seen both directly and on radiography.

Laboratory and Radiological Findings
Radiologic evidence of caries usually extending to pulp chamber.

Usual Course
If untreated, the pulp dies and infection spreads to the periapical tissues, producing acute or chronic periapical periodontitis which is likely to be acute but might be chronic. Death of the pulp ends pain from this source, but by then pain may already have started from the acute periapical periodontitis.

Relief
By analgesics, sometimes by cold fluids, extirpation of the dental pulp; extraction of the tooth.

Complication
Spread of infection to the periodontal tissues, jaws, lymph glands.

Pathology
Histopathological examination of the pulp reveals acute inflammation.

Diagnostic Criteria
Spontaneous pain confirms. Tooth not tender to percussion unless periodontitis has supervened.

Differential Diagnosis
Other forms of dental disease, rarely can mimic trigeminal neuralgia, sinusitis, vascular facial pain syndromes.

Code
031.X2c

Odontalgia: Toothache 3. Periapical Periodontitis and Abscess (IV-4)

Definition
Orofacial pain due to the causes named and having a graduated response to local stimulation.

Site
Face, jaw, mouth.

System
Musculoskeletal.

Main Features
Prevalence: common. *Sex Ratio:* no difference. *Start:* biting on the tooth makes the pain worse. In milder cases biting can start the pain. In severe cases pain occurs spontaneously. *Quality:* sharp, or dull ache, may be throbbing. *Occurrence:* with meals in milder cases; daily in severe cases. *Intensity:* moderate to severe. *Duration:* hours to days.

Signs
Ten days from onset, radiography may show resorption of bone in the periapical area.

Laboratory Findings
Various microorganisms from the exudate.

Usual Course
if untreated, pain may cease because of drainage but there are, in many cases, recurrences with further attacks of pain. Chronic abscess may also occur.

Relief
By analgesics, drainage by pulp canal therapy, extraction of the tooth. Abscess may track to mucosal surface.

Complications
Cellulitis, facial sinus, lymphadenitis, sinusitis, spread further, including cerebral abscess.

Pathology
Rarefying osteitis about apex of the tooth, abscess formation.

Diagnostic Criteria
Tenderness of tooth on palpation. Periapical resorption of bone on radiography.

Differential Diagnosis
Other dental disease.

Code
031.X2d

References
Brooke RI. Atypical odontalgia. Oral Surg 1980;49:196–9.

Marbach JJ. Phantom tooth pain. J Endodontics 1978;4:362–72.

Rees RT, Harris M. Atypical odontalgia. Br J Oral Surg 1979;16:212–8.

Odontalgia: Toothache 4. Tooth Pain Not Associated with Lesions (Atypical Odontalgia) (IV-5)

Definition
Severe throbbing pain in the tooth without major pathology.

Site
Teeth and gingivae.

System
Musculoskeletal system.

Main Features
Sex *Ratio:* female preponderance. *Age of Onset:* adults. *Pain Quality:* teeth hypersensitive to stimuli. Severe throbbing pain in teeth and gingivae usually continuous, may vary from aching mild pain to intense pain, especially with hot or cold stimuli to the teeth. May be widespread or well localized, frequently precipitated by a dental procedure. May move from tooth to tooth. *Duration:* may be from a few minutes to several hours.

Associated Symptoms
Emotional problems. May be associated with hypotensive therapy. Also complaints of temporomandibular pain and dysfunction syndrome, oral dysesthesia, and pains of psychological origin. May be a symptom of depressive or monosymptomatic hypochondriacal psychosis. Often excessive concern with oral hygiene.

Signs and Laboratory Findings
Teeth hypersensitive to heat and cold.

Relief
Antidepressants. Small doses of phenothiazines. Counseling; avoidance of unnecessary pulp extirpations and extractions.

Pathology
Possibly hyperalgesia of pulp and periodontal pain receptors due to persistent vasodilation.

Summary of Essential Features and Diagnostic Criteria
Continuous throbbing pain in the tooth, hypersensitive to temperature and pressure. No organic pathology.

Diagnostic Criteria
Patient with history of tooth pain associated with endodontic therapy and/or extractions. Remaining teeth while clinically sound and vital are tender to thermal stimuli and to percussion.

Code
034.X8b

References
Brooke RI. Atypical odontalgia. Oral Surg 1980;49:196–9.

Marbach JJ. Phantom tooth pain. J Endodontics 1978;4:362–72.

Rees RT, Harris M. Atypical odontalgia. Br J Oral Surg 1979;16:212–8.

Glossodynia and Sore Mouth (IV-6)

(also known as Burning Tongue or Oral Dysesthesia)

Definition

Burning pain in the tongue or other oral mucous membranes.

Site

Most often tip and lateral borders of tongue. Anterior hard palate, lips, and alveolar mucosa are often involved, but any mucosal area can be affected. Most often bilateral.

Main Features

Prevalence: common in postmenopausal women: 1040% of women attending postmenopausal clinics, 15% of women aged 40-49 in general dental practices, 1.52.5% of random samples of general or dental populations. *Sex Ratio:* women predominate. *Age of Onset:* mainly over 50 years of age. *Quality:* burning, tender, annoying, tiring, nagging pain; discomforting (McGill Pain Questionnaire). *Time Pattern:* usually constant once it begins, but may be variable; increases in intensity from midmorning to late evening. *Intensity:* on 150 mm VAS (visual analog scale): least, in A.M., 22 ± 25 mm; usual in afternoon, 63 ± 27 mm; and most by late evening, 105 ± 29 mm.

Associated Symptoms

Dry mouth (63% of subjects), persistent dysgeusic taste (63%), altered taste perception (35%), thirst (37%). Burning increased with tension (78%), fatigue (54%), speaking (44%), and hot foods (38%), and decreased with sleeping (69%), eating (58%), cold (52%), distraction (48%), and alcohol (27%). Many patients anxious and depressed. Topical anesthetic applied to painful sites decreases pain. Temporary relief by food or drink is almost pathognomonic. Denture intolerance can occur.

Signs and Laboratory Findings

Usually normal but there has been experimental evidence of altered taste perception, lowered heat pain tolerance of the tongue and alterations in salivary composition, although not quantity. Occasionally, there may be evidence of connective tissue disease (e.g., positive rheumatoid factor, antinuclear factor, increased sedimentation rate, decreased complement levels). Sometimes low iron, B12i folate or other vitamin B or zinc levels, but correction of nutritional factors infrequently alleviates symptoms.

Usual Course

Fifty percent spontaneous remission within 6-7 years of onset; sometimes intractable. Often responds well to tricyclic antidepressant drugs in low doses (30-60 mg). Treatment frequently more difficult in patients who have burning only when dentures in place.

Complications

Secondary emotional changes.

Pathology

Unknown, but frequently occurs around the time of menopause.

Summary of Essential Features and Diagnostic Criteria

Burning tongue or other parts of oral mucosa, usually bilateral, dysgeusic taste, altered taste perception, dry mouth, denture intolerance.

Differential Diagnosis

Atypical facial pain; atypical odontalgia; atypical trigeminal neuralgia; oral candidiasis; erosive lichen planus; geographic tongue; vitamin, iron, or zinc deficiency.

Code

051.X5	If known
051.X8	Alternative

References

Basker RM, Sturdee DW, Davenport JC. Patients with burning mouths: a clinical investigation of causative factors, including the climacteric and diabetes. Brit Dent J 1978;145:9–16.

Grushka M, Sessle BJ. Burning mouth syndrome. Dent Clin NA 1991;35:171–84.

Van der Waal I. The Burning Syndrome. Copenhagen: Munksgaard; 1990.

Cracked Tooth Syndrome (IV-7)

Definition

Brief, sharp pain in a tooth, often not understood until a piece fractures off the tooth.

Site

Mouth.

System

Musculoskeletal system.

Main Features

Prevalence: fairly common. *Sex Ratio:* no difference. *Age of Onset:* third decade onward. *Start:* brief pain on biting or chewing. *Pain Quality:* sharp. *Intensity:* moderate. *Duration:* few seconds.

Signs

It may be a visible crack. Percussion of this cusp provokes the pain. The cusp might move away from the tooth when manipulated.

Usual Course

The pain recurs with biting and chewing until the cusp finally separates completely.

Relief

It is relieved when the cracked portion of the tooth finally fractures off, or if the crack is detected by the dentist and the defective portion is restored.

Complications

None.

Social and Physical Disability

Eating is more difficult.

Pathology

A crack in the tooth allows chemicals and microorganisms to enter and make the dentine at the pulpal side of the crack hypersensitive, possibly by a mild underlying pulpitis.

Diagnostic Criteria

A sharp brief pain on biting or chewing. There is pain on percussing the affected cusp but not the other cusps. The piece finally fractures off.

Differential Diagnosis
Other forms of toothache mainly from the dentine and the pulp.

Code
034.X I

Dry Socket (IV-8)

Definition
Unilateral pain in the jaw, usually lower, usually associated with additional tenderness due to submandibular lymphadenitis following dental extraction and due to a localized osteitis.

Site
Face, jaw, mouth, upper neck.

System
Musculoskeletal system.

Main Features
Prevalence: fairly common. *Sex Ratio:* no difference. *Age of Onset:* any age from when the teeth can be extracted. *Start:* two days after a dental extraction, the pain starts without stimulation. The submandibular lymph glands soon become involved with added tenderness. *Pain Quality:* constant, dull ache, may throb, associated with severe halitosis. *Intensity:* moderate, exacerbated by mechanical stimulation. *Duration:* hours to days.

After tooth extraction, blood normally fills the socket and clots, the clot gradually becoming organized with new bone formation. Dry socket occurs when this fails to happen either because there is no bleeding due to too much adrenaline in the local anesthetic solution, or because the blood is diluted by washing the mouth out, or because the clot is broken down by infection. In such circumstances the bone in the socket is no longer protected, and there is severe pain made worse by physical interference. Food gathers in the socket and decomposes, producing a foul taste and severe halitosis.

Signs
A recent extraction socket with no clot (and therefore dry), with food debris.

Laboratory and Radiological Findings
Recent empty tooth socket.

Usual Course
Continuous unless treated. Gingiva tends to grow over the socket.

Relief
It is relieved by washing out the socket and packing it with ribbon gauze covered with Whitehead's varnish (an iodoform resinous material).

Complication
Submandibular lymphadenitis.

Social and Physical Disability Severe halitosis.

Diagnostic Criteria
Continuous ache which starts two days after tooth extraction. Socket not closed by blood clot. Food debris within. Halitosis. Pain from mechanical stimuli. Submandibular lymphadenitis.

Differential Diagnosis
Osteomyelitis, retained tooth root.

Code
031.X1

Gingival Disease, Inflammatory (IV-9)

Code
034.X2

Toothache, Cause Unknown (IV-10)

Code
034.X8f

Diseases of the Jaw, Inflammatory Conditions (IV-11)

Code
033.X2

Other and Unspecified Pain in Jaws (IV-12)

Code
03X.X8d

Frostbite of Face (IV-13)

Code
022.X I

GROUP V: PRIMARY HEADACHE SYNDROMES, VASCULAR DISORDERS, AND CEREBROSPINAL FLUID SYNDROMES

Classic Migraine (Migraine with Aura) (V-1)

Definition
Throbbing head pain in attacks, often with a prodromal state and usually preceded by an aura which frequently contains visual phenomena. The pain is typically unilateral but may be bilateral. Nausea, vomiting, photophobia, and phonophobia often accompany the pain. Clear female predominance.

Site
Typically unilateral, but may be bilateral. Pain mostly begins in the fronto-temporal area and is most marked in this area, even at maximum, when it may involve the whole hemicranium. The side typically changes in different attacks or even during single attacks.

System
Unknown: vascular disturbances have been emphasized; central nervous system changes may be fundamental. The coding below accepts the latter.

Main Features
Frequent positive family history of migraine-like type of headache. *Prevalence:* high, but less frequent than common migraine. *Sex Ratio:* females more than males. *Onset:* from childhood to about 35. In most cases, attacks have started by late puberty. Onset of solitary attacks may be associated with emotional stress, relaxation, "anxiety," dietary causes (chocolate, cheese, citrus fruits, etc.), flashing lights, atmospheric changes, etc. *"Premonitory" Phase:* may last for hours to one or two days and precedes the aura phase, often with mood changes, weight gain. *The Aura* usually precedes the pain phase but may also occur both prior to and during it, and occasionally only during it. An aura may occur without subsequent pain, probably most frequently in male patients. In approximate order of frequency, the following phenomena occur during the aura phase: blurring of vision, flickering changes in the visual field, phenomena like a curtain or mist in parts of the field, fortification figures, scotomata and a variety of other visual changes (the visual changes usually have a homonymous distribution), paresthesias, mostly in the regions of the hand and mouth, mild paresis (the two last phenomena usually with a unilateral distribution), dysarthria, and aphasic disturbances. In extremely rare cases, there may be alloesthesia, micropsia, and macropsia, or distortions of perspective. If paresis, hemianopias, and sensory loss are prominent and longlasting, they may be part of other migraine variants (V-3). *Duration of Aura Phase:* usually 20-25 minutes. *Pain:* the aura may overlap with the pain phase. Usually the pain succeeds the aura with or without a symptom-free interval. In occasional attacks in the classic migraineur, the pain starts without a preceding aura. The pain is throbbing, ranges from mild to severe in intensity, reaches a plateau, and usually lasts from 4 to 72 hours if unmodified by drugs. The pain may be global, but typically it is unilateral and alternates sides during an attack or between attacks. The pain typically starts in the frontal temporal area. It may continue in that area or involve the entire hemicranium at a later stage. The pain is generally moderate to severe. Characteristically, the pulsating quality increases with moderate physical activity or stooping. *Frequency:* varies from a couple of attacks in a lifetime to several every week. The most usual pattern in clinical practice is 1-4 per month. Exacerbations often occur during episodes of anxiety, depressive illness, or personal conflict. The tendency to attacks is frequently markedly reduced in pregnancy. *Other Characteristics:* anorexia, nausea and vomiting, photophobia, and phonophobia are characteristic features of the attack.

Precipitating Factors
Numerous, may include stress, mood changes, relaxation, dietary factors.

Associated Symptoms and Signs
Anorexia, nausea, vomiting, photophobia, and phonophobia. With "complicated migraine," various deficiency symptoms and signs (e.g., hemiplegic migraine; see V-3).

Laboratory Findings
Fall in platelet serotonin during attacks. Changes in cerebral blood flow.

Relief
From ergot preparations, beta-blocking agents, calcium blocking agents, NSAIDs, and substances interfering with serotonin activity, in particular serotonin 1D receptor agonists like sumatriptan.

Usual Course
In time, interparoxysmal psychological changes if the headache is severe. Ergotamine dependence or other dependence on medication, even analgesic medication. Detoxification may be required to end a vicious circle of withdrawal headaches and medications.

Complications
Depression and related psychological changes if severe. Dependence on ergotamine or other medication.

Social and Physical Disability
Interruption of work in severe cases. Reduced efficiency for many.

Pathology
No definite, confirmed findings.

Essential Features
Presence of an aura phase, at least during the occasional attack. Pulsating headache. Usually unilateral headache. Nausea, vomiting, photophobia, and phonophobia.

Differential Diagnosis
Common migraine, migraine variants, cerebral angioma.

Code
004.X7a

References
Bille S. Migraine in children and its prognosis, Cephalalgia 1981;I:71–5.

Blau JN, editor. Migraine. London: Chapman & Hall; 1987.

Graham JR. Seven common headache profiles. Neurology 1963;13:16–23.

Selby G. Migraine and Its Variants. Sydney: ADIS Health Science Press; 1983.

Common Migraine (Migraine without Aura) (V-2)

Common migraine generally has the same characteristics as the classic variety with some exceptions, of which the important ones are given below.

Definition
Repetitive, unilateral, and occasionally bilateral throbbing headache attacks, moderate to severe in intensity, often with a premonitory stage but without a distinct, clinically discernible aura, usually

accompanied by nausea, vomiting, photophobia, and phonophobia. The pain alternates sides between attacks or even during an attack. The pain usually starts in the frontal areas.

Main Features
Prevalence: the prevalence is probably high. Estimates range from 1% to 31% depending on the criteria for definition of headache. Common migraine occurs much more often than classic migraine (the ratio of common to classic migraine is 2:1 or 3:1, depending upon the strictness of adherence to "classic" and to "common" criteria). *Aura:* absent. If the patient has had several attacks with aura, the majority being without an aura, the patient should still be classified under classic migraine. The complaints are clearly accentuated by minimal physical activity.

Other Features
Common migraine attacks usually last 1-2 days but may last longer, and at times may last only a few hours (lower limit: perhaps around 4 hours).

Relief
See Classic Migraine (V-1).

Complications
Drug abuse of analgesics and/or ergotamine. This is a frequent phenomenon. An improvement of the situation cannot be obtained unless detoxification is carried through.

Essential Features
The aura phase is lacking. The attack may seem to last longer than the classic migraine attack. Otherwise, grossly similar to classic migraine.

Differential Diagnosis
Tension headache, cervicogenic headache. Common migraine in general seems to be characterized by the absence of features characteristic of cervicogenic headache, such as reduced range of motion in the neck; ipsilateral, vague, nonradicular shoulder-arm pain; mechanical precipitation of attacks (see V-7.1).

Code
004.X7b

References
Blau JN, editor. Migraine. London: Chapman & Hall; 1987.

Graham JR. Migraine: clinical aspects. In: Vinken PJ, Bruyn GW, editors. Handbook of Clinical Neurology. Amsterdam: North-Holland Publishing Co; 1968. p. 45–58.

Selby G, Lance JW. Observations on 500 cases of migraine and allied vascular headache. J Neurol Neurosurg Psychiatry 1960;23:23–32.

Migraine Variants (V-3)

Hemiplegic migraine, migraine accompagnée, basilar migraine, ophthalmoplegic migraine, retinal migraine.

These variants are not described in detail. The neurological symptoms and signs are more pronounced than in "ordinary" migraine. The question of the nature of the underlying neurological disturbance may be more important than that of the differential diagnosis from other headache syndromes. Some of these terms (e.g., basilar migraine and retinal migraine) may be wrongly chosen, and it is uncertain whether

they reflect separate entities.

"Migraine cervicale" is not grouped as a migraine variant, since it probably is not "migrainous" in nature. It may rather be a headache associated with neck disorders (see Cervicogenic Headache [VII-2]).

Differential Diagnosis
Classic and common migraine, Chiari malformations, arteriovenous malformations and other structural abnormalities, pseudotumor cerebri, etc., Tolosa-Hunt syndrome (painful ophthalmoplegia), and Raeder's syndrome.

Code
004.X7c

Note: See note on Cluster Headache (V-6).

Carotidynia (V-4)

Definition
Continuous dull aching pain, sometimes throbbing, near the upper portion of the carotid arteries and adjoining cranial regions, with features of migrainous exacerbation. A partly different picture has also been described, Roseman's variant, with a self-limited, relatively short-lasting course.

Site
Pain in the neck, frequently radiating to the face and head (temporal/mastoid area), usually on one side.

System
Vascular system, probably common and internal carotid arteries.

Main Features
Prevalence: occurrence unknown, depends somewhat upon the criteria used, probably rather rare. *Sex Ratio:* more prevalent in the female than the male except for Roseman's variant, where there seems to be no sex preponderance. *Age of Onset:* usually between 20 and 60 years of age. *Pain Quality:* the pain is constant and dull, aching or throbbing. *Time Pattern:* protracted course; dull, continuous neck pain with superimposed separate attacks of hours duration. Roseman's variant: 7-10 days to several weeks. Some patients seem to experience only one episode. There is, however, a tendency for the pain episodes to recur after a symptom-free interval. *Intensity:* moderate, not very severe; apparently less severe than migraine headache.

Precipitating Factors
Moving the head, swallowing, coughing, etc., may precipitate or aggravate the pain.

Associated Symptoms and Signs
Incapacity, nausea, and photophobia. Rarely vomiting. In Roseman's variant, few features in addition to the moderate pain. The carotid artery may on palpation appear enlarged, pulsating, and tender, and externally applied pressure against the common carotid artery may reproduce the pain in the neck and face. Regional muscles may also be tender.

Relief
The treatment of carotidynia is the same as that for migraine; prophylactic drugs (propranolol, methysergide) and ergotamine may help. Symptomatic treatment may also be of some avail. For

Roseman's variant, no drugs have been found to be of specific use. It should be emphasized that in this variant the pain episode is self-limited and rather shortlasting. Success in treatment may, therefore, be confounded with the natural course of the disease.

Pathology
Unknown. The nosologic status of these headaches remains obscure.

Code
004.X7d

References
Lovshin LL. Carotidynia. Headache 1977;17:192–5.

Murray TJ. Carotidynia: a cause of neck and face pain. CMAJ 1979;120: 441–3.

Raskin NH, Prusiner S. Carotidynia. Neurology 1977;27:43–6.

Roseman DM. Carotidynia. Arch Otolaryngol 1967;85:81–4.

Mixed Headache (V-5)

Mixed headache in most cases probably refers either to migraine with interparoxysmal headache or to chronic tension headache, as described above. The headache should accordingly be categorized, whenever possible, as either migraine or chronic tension headache.

Code
003.X7b

Cluster Headache (V-6)

Definition
Unilateral, excruciatingly severe attacks of pain, principally in the ocular, frontal, and temporal areas, recurring in separate bouts with daily, or almost daily, attacks for weeks to months, usually with ipsilateral lacrimation, conjunctival injection, photophobia, and nasal stuffiness and/or rhinorrhea.

Site
Ocular, frontal, temporal areas: considerably less frequent in infraorbital area, ipsilateral upper teeth, back of the head, entire hemicranium, neck, or shoulder. The maximum pain is usually in ocular, retro-ocular, or periocular areas. Unilateral pain without alternation of sides is characteristic. The side *may,* however, change (in approximately 15% of the patients), even within a given cluster period.

System
Uncertain. The autonomic nervous system is activated. The vascular system is also involved. The pain may be neurogenic. The central nervous system may play a role.

Main Features
Prevalence: approximately 7 per 10,000 population. *Sex Ratio:* 85-90% male. *Age of Onset:* most frequently, headaches start between the ages of 18 and 40. *Pain Quality:* the pain is constant, stabbing, burning, or even throbbing. Patients characteristically pace the floor, bang their heads against the walls, etc., during attacks because of the vehement pain and are usually unable to lie down. *Time Pattern:* attacks grouped in bouts ("cluster periods") of several weeks' to months' duration (most often: 4-12

weeks, with a range from less than 1 week to 12 months), with intervals of some months' duration more or less free from attacks. Usually one cluster period occurs per 6-18 months. Usually, 1-3 attacks, lasting from half an hour to 2 hours each, occur per 24 hours in the cluster period. The maximum number of attacks is ordinarily 6-8 per 24 hours. Attacks may skip a day or two or more during the cluster period. Nocturnal attacks are typical. The patients tend to smoke and drink rather heavily. Sensitivity to alcohol occurs during bouts. *Intensity:* at maximum, excruciatingly severe. Abortive or mild attacks may nevertheless occur.

Precipitating Factors
Alcohol, during the bout. Long-lasting stress may possibly predispose to bouts.

Associated Symptoms and Signs
Usually there is no nausea, but some may occur, probably with the more severe attacks or at the peak of attacks. Vomiting is less frequent than nausea. Ipsilateral miosis or ptosis associated with some attacks; occasionally they persist after attacks and sometimes permanently. Ipsilateral conjunctival injection, lacrimation, stuffiness of the nose, and/or rhinorrhea occur in most patients. Dysesthesia upon touching scalp hairs in the area of the ophthalmic division of the Vth cranial nerve and photophobia occur in most patients. A reduction in heart rate and irregular heart activity are features in some patients, especially during severe attacks.

Relief
From ergot preparations, oxygen, corticosteroids, lithium, verapamil, methysergide, etc. Serotonin 1D receptor agonists, like sumatriptan, have a convincing, beneficial effect.

Usual Course
Attacks, less than 1 to 3 per day, appearing in bouts of 412 weeks duration. Remissions last one-half to one and one-half years. The episodic form may eventually develop into a chronic form. Possibly, less activity of the disease process with age.

Complications
Suicide risk; peptic ulcer.

Social and Physical Disability
Considerable during bout. Many patients, nevertheless, manage to do their work between attacks.

Pathology
Unknown. Perhaps cavernous sinus changes or "central" changes.

Essential Features
Excruciatingly severe attacks of unilateral headache, appearing in bouts, lasting less than 1 year. Autonomic symptoms and signs on the symptomatic side. Male preponderance.

Differential Diagnosis
Sinusitis, chronic paroxysmal hemicrania, chronic cluster headache, cluster-tic syndrome, and migraine. Cervicogenic headache and tic douloureux ought not to present differential diagnostic problems.

Code
004.X8a

Note: Although cluster headache is grouped with migraine and similar disturbances, it is doubtful if vascular disturbances are the primary source of these events, and the second code digit refers to

alternative possibilities for the origin of the pain.

References

Kudrow L. Cluster Headache: Mechanism and Management. London: Oxford University Press; 1980.

Manzoni GWC, Terzano MG, Bono G, Micieli G, Martucci N, Nappi G. Cluster headache: clinical findings in 180 patients. Cephalalgia 1983;3:21–30.

Russell D. Studies of Autonomic Functions in the Cluster Headache Syndrome. Thesis. Oslo University; 1985.

Sjaastad O. Cluster Headache Syndrome. Philadelphia: WB Saunders; 1992.

Chronic Paroxysmal Hemicrania (CPH) (Unremitting Form or Variety) (V-7.1)

Definition

Multiple daily attacks of severe to excruciating unilateral head pain, more frequently occurring in females than in males, and principally in ocular, frontal, and temporal areas by day and night, usually accompanied by ipsilateral lacrimation, conjunctival injection, and nasal stuffiness and/or rhinorrhea, and with absolute relief from indomethacin. Chronicity denotes an unremitting stage that has lasted more than a year.

Site

Ocular, frontal, and temporal areas; occasionally the infraorbital, aural, mastoid, occipital, and nuchal areas. Pain may also be felt in the ipsilateral part of the neck, arm, and upper part of the chest. There are only rare exceptions to the rule of unchanging unilaterality.

System

Uncertain. The pain may be neurogenic. The vascular and autonomic nervous systems are implicated during attacks. Central nervous system changes may play a role.

Main Features

Prevalence: probably rare. *Sex Ratio:* around 70% females. *Age of Onset:* average around 35 (more than 90% are aged *11-60). Time Pattern:* at the top of the curve, attacks appear at a rate of 9 or more per 24 hours in more than 80% of the cases (range 4-40 attacks per 24 hours). Patients have attacks every day. Attacks may occur at relatively regular intervals all through day and night. Characteristically, there is marked fluctuation in the severity of attacks and their frequency. A period of 1-2 moderate attacks per day (occasionally even barely noticeable) is followed by a period with frequent, severe attacks, thus providing a “modified cluster pattern.” Attacks usually last between 10 and 30 minutes (80% are less than 30 minutes in duration). *Pain Quality:* the pain is clawlike, throbbing, and occasionally boring, pressing, or like “dental” pain. Not infrequently, the patients are awakened by the nocturnal attacks. Some patients walk around during attacks, others sit quietly, still others curl up in bed. *Intensity:* at maximum, the pain attacks are excruciatingly severe, but there is marked fluctuation in severity.

Precipitating Factors

Attacks may be precipitated in the occasional patient (around 10%) by bending or rotating the head, particularly when at the peak of the attack curve (“mechanical precipitation of attacks”).

Associated Symptoms and Signs

Ipsilateral conjunctival injection and lacrimation occur frequently, as do ipsilateral nasal stuffiness and/or rhinorrhea. Nausea is rare and vomiting very rare. Slight ipsilateral ptosis or miosis may occur during

attacks, and rarely also edema of the upper lid. Photophobia and more rarely phonophobia are occasionally present during attacks. Tinnitus, hypersensitivity in the area of the ophthalmic division of the Vth cranial nerve, bradycardia, and extrasystoles occur in some patients during severe attacks.

Laboratory Findings
Increased nasal secretion and lacrimation (and partly also forehead sweating); increased intraocular pressure and corneal indentation pulse (CIP) amplitudes on the symptomatic side during attack.

Relief
Immediate, absolute, and permanent from continuous indomethacin treatment.

Usual Course
The chronic course may be *primary* chronic or it may develop from a remitting stage. Once chronic, the headache usually *remains* chronic. One case has been observed to revert to a remitting stage after many years of indomethacin treatment, and in a few cases, headache has virtually disappeared after a short course of indomethacin. Attacks frequently disappear partly or even completely during the greater part of pregnancy, to reappear immediately postpartum.

Complications
Possibly CPH "status." Untoward effects of chronic indomethacin therapy-peptic ulcer.

Social and Physical Disability
Considerable during the nontreated stage, including suicidal thoughts. In the worst cases, the patient does not function properly socially.

Pathology
Not identified yet.

Essential Features
Unremitting presence for at least one year of relatively shortlasting repetitive unilateral attacks, associated with ipsilateral autonomic symptoms and signs. Absolute response to indomethacin.

Differential Diagnosis
CPH, remitting form. Sinusitis, chronic cluster headache, cluster headache, cluster-tic syndrome, hemicrania continua.

Code
006.X8k

Note: See note on Cluster Headache (V-6).

References
Antonaci F, Sjaastad O. Chronic paroxysmal hemicrania (CPH): a review of the clinical manifestations. Headache 1989;29:648–56.

Sjaastad O, Dale I. A new (?) clinical headache entity: "chronic paroxysmal hemicranias." Acta Neurol Scand 1976;54:140–59.

Sjaastad O. Chronic paroxysmal hemicrania (CPH). In: Vinken PJ, Bruyn GW, Klawans H, Rose FC, editors. Handbook of Clinical Neurology 48. Amsterdam: Elsevier;1986. p. 257–66.

Sjaastad O. Chronic paroxysmal hemicrania (CPH): nomenclature as far as the various stages are concerned. Cephalalgia 1989;9:1–2.

Chronic Paroxysmal Hemicrania (CPH) (Remitting Form or Variety) (V-7.2)

The features of the remitting form are the same as for the chronic ("unremitting") form of CPH. The differences mainly concern the temporal pattern. Accordingly, for other details, the section on the unremitting variety (V 7.1) should be consulted. Absolute relief from indomethacin.

Definition
Attacks of unilateral severe or excruciating headache, occurring more frequently in females than in males, in the ocular, fronto-temporal area, and with the same attack characteristics as in the unremitting form. The periods of attacks last from a few days to many months (if a period exceeds 12 months, the chronic, unremitting stage has been reached). The remitting stage may seemingly go on indefinitely.

Main Features
The remitting form seems to be more rare than the unremitting. This is partly due to the not infrequent conversion of the remitting form to the chronic one. The diagnosis of the remitting form requires a duration of less than 1 year of a period of attacks.

Relief
Immediate, absolute, and permanent effect of indomethacin.

Essential Features
Frequently occurring, relatively shortlasting attacks of unilateral headache, not present continuously for as much as one year. Female preponderance. Absolute response to indomethacin.

Differential Diagnosis
CPH, unremitting form; cluster headache; sinusitis; cluster-tic syndrome; hemicrania continua.

Code
006.X8g

References
Antonaci F, Sjaastad O. Chronic paroxysmal hemicrania (CPH): a review of the clinical manifestations. Headache 1989;29:648–56.

Sjaastad O. Chronic paroxysmal hemicrania (CPH): nomenclature as far as the various stages are concerned. Cephalalgia 1989;9:1–2.

Sjaastad O. Cluster Headache Syndrome. Philadelphia: WB Saunders; 1992.

Chronic Cluster Headache (V-8)

The main features of chronic cluster headache are the same as those for the episodic form of cluster headache, to which the reader is referred for further details (V-6). The differences mainly concern the temporal pattern.

Definition
Bouts of excruciatingly severe unilateral pain, usually in males, principally in the ocular, frontal, and temporal areas, usually occurring more frequently than twice a week and *for more than one year*.

Main Features
The chronic form may be *primary* chronic (i.e., the ordinary, episodic form has never existed) or *secondary* chronic (i.e., a further development from the episodic form). The chronic form of cluster headache is more rare than the episodic form (approximately 1:8); the diagnosis requires at least two or more attacks per week over a period of more than one year. Occasionally, however, even longer attack-free intervals may occur.

Relief
The same measures are effective as for cluster headache, but generally the chronic form is more difficult to treat. Surgical procedures (e.g., radio-frequency treatment of the Gasserian ganglion) may be more justifiable in the chronic than in the episodic case.

Essential Features
The unremitting presence of unilateral, relatively short-lasting, and excruciatingly severe attacks for at least one year. Autonomic symptoms and signs on the symptomatic side.

Differential Diagnosis
Sinusitis, chronic paroxysmal hemicrania, cluster headache (episodic form), cluster-tic syndrome, migraine.

Code
004.X8b

Note: See note on Cluster Headache (V-6).

References
Mathew N, Hurt W. Percutaneous radio-frequency trigeminal gangliorhizolysis in intractable cluster headache. Headache 1988;28:328–31.

Onofrio BM, Campbell JK. Surgical treatment of chronic cluster headache. Mayo Clin Proc 1986;61:537–44.

Sjaastad O. Cluster Headache Syndrome. Philadelphia: WB Saunders; 1992.

Cluster-Tic Syndrome (V-9)

Definition
The coexistence of the features of cluster headache and tic douloureux (trigeminal neuralgia), whether the two entities occur concurrently or separated in time.

Site
Pain limited to the head and face; the two parts of the syndrome generally appear on the same side. The cluster headache element is located in the ocular area as is usual in cluster headache. The most common site of the tic pain is the second or third divisions of the trigeminal nerve.

System
Nervous system.

Main Features
Prevalence: rare. *Sex Ratio:* approximately equal. *Age of Onset:* usually middle age; more rarely in the elderly. *Quality:* a combination of the following: cluster headache pain which includes agonizingly severe, longlasting, burning or throbbing pain, and, concurrently or separated in time, sharp, agonizing,

electric shock-like stabs of pain felt superficially in the skin or buccal mucosa, triggered by light tactile stimuli from a restricted trigger point (the features of trigeminal neuralgia). *Time Pattern:* Paroxysms of brief pains occur many times a day with periods of freedom from pain. The attack is often precipitated by speaking, swallowing, washing the face, or shaving. This happens concurrently with, or temporally separated from, the features of cluster headache. The latter comprises severe episodes of steady pain lasting 10-120 minutes, frequently occurring at night, and characteristically occurring in cluster periods lasting 4-8 weeks, once or twice a year, but at times entering a more chronic phase and occurring daily for months. *Intensity:* Extremely severe; both elements of the combined syndrome are among the most severe pains.

Precipitating Factors
For the "tic component," a "trigger phenomenon," as with tic douloureux (see II-1). For the "cluster component," alcohol.

Associated Symptoms
Prominent autonomic features with the cluster-type pain, i.e., ipsilateral nasal obstruction or discharge, or both, ipsilateral lacrimation and conjunctival injection, facial flushing, facial diaphoresis, and agitation.

Signs and Laboratory Findings
Occasionally the presence of a Homer's syndrome is noted, presumably as a residuum from the attacks of cluster headache. No sensory deficit is present over the face.

Relief
The most successful treatment appears to be the use of carbamazepine or baclofen, or both, rather than the conventional drugs used for cluster headache.

Usual Course
The attacks of cluster headache and tic douloureux may start concurrently, or the attacks of tic douloureux may precede those of cluster headache. Cluster headache seems to precede tic douloureux only rarely.

Complications
Depression.

Social and Physical Disabilities
Usually profound during the attacks.

Pathology
Unknown.

Essential Features
Coexistence of features of cluster headache and tic douloureux. These two components of the syndrome may appear simultaneously or separated in time.

Differential Diagnosis
Sinusitis, chronic paroxysmal hemicrania. A careful neurological examination and appropriate tests such as CT scans may be necessary to rule out tumors in the cerebello-pontine region.

Code
006.X8h

References

Green M, Apfelbaum RJ. Cluster-tic syndrome. Headache 1978;18:112.

Solomon S, Apfelbaum RI, Guglielmo K. The cluster-tic syndrome and its surgical therapy. Cephalalgia 1985;25:123–6.

Post-traumatic Headache (V-10)

Definition
Continuous or nearly continuous diffusely distributed head pain associated with personality changes involving irritability, loss of concentration ability, dizziness, visual accommodation problems, change in tolerance to ethyl alcohol, loss of libido, and depression, and with or without post-traumatic stress disorder, following head injury.

Site
Head.

System
Nervous system.

Main Features
Prevalence: unknown. *Sex Ratio:* males more than females. *Onset:* difficult to recognize in children, particularly during rebellious age. *Pain Quality:* nonspecific, generalized, nonthrobbing, without aura, and without autonomic dysfunction such as nausea, vomiting, or diarrhea. *Time Pattern:* nearly constant. *Intensity:* mild (relative to migraine), but can be severe.

Associated Symptoms
Personality change involving irritability, inability to concentrate on relatively trivial matters such as balancing a checkbook, lightheadedness or vertigo, intermittent visual accommodation error, change in tolerance, usually intolerance of ethyl alcohol, and loss of libido with or without depression and with or without post-traumatic stress disorder.

Signs and Laboratory Findings
Any objective abnormality including MMPI changes, EEG abnormalities, clinical convulsions, focal neurologic findings, and organic brain syndrome usually absent and if present markedly limits the prognosis.

Usual Course
Without treatment, weeks to months, and in the presence of focal neurologic abnormalities, convulsions, or organic brain syndrome, indefinite.

Complications
Loss of victim's will to combat the illness.

Social and Physical Disabilities
At worst, left untreated, loss of gainful employment and family and social status to the point of complete destitution.

Pathology
Disruption of central axons and boutons due to angular positive or negative acceleration of the brain (unproven hypothesis). Damage to labyrinth is often postulated as well, and soft-tissue lesions from cervical sprain syndrome.

Differential Diagnosis
The word concussion is to be avoided because of lack of agreement in definition of term. Confusion with possible accompanying depression, post-traumatic stress disorder, and other accompanying or complicating psychiatric organic brain dysfunction disorders is to be avoided. In the presence of focal neurologic findings, convulsions, or organic brain syndrome, it is necessary to rule out subdural hematoma and other space-occupying lesions. It is difficult or impossible to distinguish from tension headache. The spouse or family is much more likely to be aware of the irritability of the victim.

Code
002.X l **b**

References
Brenner C, Friedman AP, Merritt HH, Denny-Brown DE. Post-traumatic headache. J Neurol 1944;1:379.

Merskey H. Psychiatry and the cervical sprain syndrome (editorial). CMAJ 1984;130:1119–21.

Trimble MR. Post-traumatic Neurosis. Chichester: John Wiley & Sons; 1981.

Tyler GS, NcNeely HE, Dick ML, Treatment of posttraumatic headache with amitriptyline. Headache 1980;20:213.

The Syndrome of "Jabs and Jolts" (V-11)

("Ice-Pick Pain" [Raskin]; "Multiple Jabs" [Mathew]; "Idiopathic Stabbing Headache" [nomenclature of the International Headache Society])

Definition
Shortlasting (mostly "ultra-short") paroxysms of head pain, with varying localization, even in the same patient; most often unilateral; in one or more locations. Highly varying frequency even in the same person, usually of moderate severity.

Site
In any region of the head. During one period, the pain may be situated in one area, only to move to another one during another period. Usually unilateral at a given time; in the rare case, bilateral. When associated with hemicrania continua, etc., it frequently occurs in the painful area. In the preheadache phase of chronic paroxysmal hemicrania, it may appear on the side opposite that of the pain.

System
Nervous system.

Main Features
Prevalence: probably common, since it appears both on its own and in many combinations. Frequently associated with various types of unilateral headache, such as chronic paroxysmal hemicrania, cluster headache, migraine, temporal arteritis (giant cell arteritis), hemicrania continua, and probably also tension headache. Sex Ratio: both sexes. Age of Onset: any age (except perhaps childhood). Since several of the headache forms with which it is combined have a clear female preponderance (see above), it is likely that within some of them there is a female preponderance also of Jabs and Jolts. Pain Quality: Sharp, shortlasting, superficial, neuralgiform ("knifelike") pain, superimposed upon the preexisting pain if it occurs in conjunction with another specific headache. Under such circumstances jabs and jolts seem to increase at the time of the symptomatic episodes and in the related areas. The Syndrome of Jabs and Jolts also seems to be a headache per se, unassociated with any of the above-mentioned headaches. Can usually not be triggered from any palpable trigger point. May occasionally be triggered by neck movements, change of position, etc. Most paroxysms occur unprovoked. Time Pattern: Extremely unpredictable

paroxysms from a temporal point of view, but may appear in bouts (cycles); even within such periods, irregular appearance, from less than once per day to multiple times per hour; the jabs usually appear together with the associated headache. May appear as solitary paroxysms or in volleys. Each paroxysm may last 1-2 seconds, but may occasionally last up to 1 minute (partly as lingering pain after the severe pain). A bout may last a day or two or months. Usually no nocturnal appearance. Intensity: Usually moderate, but can in periods be more severe. It may be so severe as to cause a jolt.

Precipitating Factors
Neck movements, change of body position, etc. Underlying mechanism: occasionally perhaps, mechanical irritation from enlarged lymph nodes.

Associated Symptoms and Signs
Few, if any, except for those of accompanying conditions.

Relief
Usually self-limiting. In some patients there is a good, incomplete effect from indomethacin (150 mg a day). The erratic spontaneous course of this headache makes the assessment of drug therapy a most difficult task.

Usual Course
Sporadic paroxysms, or bouts with accumulation of paroxysms, the bouts being of extremely varying duration, from less than one per day to many daily for months. Most frequently bouts recur.

Complications Probably none.

Social and Physical Disability
In periods with accumulated jabs, the patient may be transitorily handicapped.

Pathology
Unknown, but nerve fibers are the likely source.

Essential Features
Ultrashort paroxysms in the cephalic area, in multiple sites, with no fixed location, and with very varying frequency, often occurring in bouts. Occurs sporadically or in conjunction with other headaches, such as chronic paroxysmal hemicrania, migraine, etc.

Differential Diagnosis
Trigeminal neuralgia, SUNCT syndrome.

Code
006.X81

References
Lance JW, Anthony M. Migrainous neuralgia or cluster headache? J Neurol Sci 1971;13:401–4.

Mathew N. Indomethacin responsive headache syndromes, Headache 1981;21:147–50.

Raskin NH, Schartz RK. Icepick-like pain, Neurology 1980;30:203–5.

Sjaastad O. Chronic paroxysmal hemicrania (CPH): the clinical picture. Proc Scand Migraine Soc 1979;10.

Temporal Arteritis (Giant Cell Arteritis) (V-12)

Definition
Unilateral or bilateral headache, mainly continuous with aching or throbbing pain, sometimes very intense, usually in the elderly, with signs of temporal artery involvement-and occasionally more extensive cranial arterial involvement. Commonly associated with muscular aching ("polymyalgia rheumatica") and systemic disturbances like malaise, low-grade fever, and weight loss.

Site
The pain is maximal in the temporal area on one or both sides, from which it may spread to neighboring areas.

System
Vascular system.

Main Features
Prevalence: relatively rare; annual incidence 3-9 per 100,000. Sex Ratio: more common in the female. *Age of Onset:* mostly after fifth decade. *Pain Quality:* varying severity from dull aching to intense pain, more or less continuous, at times pulsating headache. *Time Pattern:* usually a rather protracted course if untreated. The disorder may manifest itself with a repetitive pattern. May be particularly severe at night. *Intensity:* Moderate to severe, probably never excruciatingly severe.

Precipitating Factors
Mastication may produce an effect of intermittent claudication.

Associated Symptoms and Signs
The temporal artery on the symptomatic side may be bulging and irregular in its appearance. The eyesight may fail on the symptomatic side or both sides, and chewing may become deficient during the later part of meals. No deficiency signs from the Vth cranial nerve at rest.

Laboratory Findings
The temporal artery may be pulseless, tender to palpation, and clearly irregular in its shape. A temporal artery biopsy may reveal giant cell arteritis; to some extent this depends upon the stage of disease and whether or not the biopsy is representative.

Visual acuity may fade as a consequence of the disease process. This usually occurs in the early stage of disease; it is an "alarm" situation and necessitates immediate therapeutic action (corticosteroid therapy). Involvement of the other eye may occur after a short time. Arterial involvement is demonstrable with, for example, angiography and ocular dynamic tonometry (reduced corneal indentation pulse [CIP] amplitudes). The erythrocyte sedimentation rate is frequently clearly raised.

Relief
From corticosteroid and immunosuppressive therapy, e.g., azathioprine therapy.

Usual Course
The prognosis has changed drastically with the advent of corticosteroids. The early start of steroid therapy is essential. Once blindness has appeared, the prognosis for this phenomenon is poor. Relapse may occur in the early stage. Late deaths are more likely to be due to complications of steroid therapy than to the arteritis.

Complications
Fading vision or blindness (see Laboratory Findings). Impaired chewing in late phase of meals-probably due to masticatory muscle ischemia, caused by the same disease process in the appropriate arteries.

Ocular palsy; arteritis in other vessels, e.g., coronary arteries and aorta; cerebral infarction. Complications may also arise as a consequence of steroid therapy.

Social and Physical Disability
Considerable during the acute stage, and in the case of complications like blindness.

Pathology
Fibrous tissue formation (giant cell arteritis) in the arterial wall. Relationship to polymyalgia rheumatica.

Essential Features
Acute pain, not infrequently unilateral, in the temporal area in an elderly person, with tenderness and irregular shape of the ipsilateral temporal artery and, usually, raised erythrocyte sedimentation rate. Various complications may arise, such as blindness.

Differential Diagnosis
Other acute unilateral headaches, such as the Tolosa-Hunt syndrome and Raeder's paratrigeminal neuralgia *in the early stages;* carotidynia; hemicrania continua; temporomandibular joint dysfunction (Costen's syndrome); auriculotemporal nerve neuralgia; polymyalgia rheumatica.

Code
023.X3

References
Horton BT, Magath TB. An undescribed form of arteritis of the temporal vessels. Mayo Clin Proc 1923;7:700–1.

Ross Russell RW. Giant cell (cranial) arteritis. In: Vinken PJ, Bruyn GW, Klawans HL, Rose FC, editors. Handbook of Clinical Neurology 48. Amsterdam: Elsevier; 1986. p. 309–28.

Headache Associated with Low CSF Pressure (V-13)
(Spontaneous Low CSF Pressure Headache)

Definition
Dull aching or throbbing headache associated with low CSF pressure occurring spontaneously or after a minor incident.

Site
May be frontal, occipital, or global, and not infrequently unilateral.

System
Probably vascular or meningeal, or both.

Main Features
Prevalence: probably rare. *Sex Ratio:* probably a female preponderance. *Age of Onset:* most cases described have been more than 30 years old. *Pain Quality:* usually dull or aching, but may be throbbing. *Intensity:* from mild to rather severe, probably never excruciating. *Precipitating Factors:* the pain is positional, markedly exacerbated or only present when the patient is sitting or standing, and usually relieved by lying down. *Time Pattern:* onset is usually insidious, but may occur after a mild trauma, sneezing, sudden strain, or orgasm. Individual headache episodes usually last as long as the patient remains in the upright position.

Associated Symptoms and Signs Pain and stiffness in the neck, nausea, vomiting, tinnitus, dizziness, blurred vision, and VIth cranial nerve palsy have all been reported.

Laboratory Findings
A low CSF pressure, usually <_ 60 mm H2O, is found on lumbar puncture with the patient lying horizontally.

Usual Course
Most cases improve spontaneously after a few weeks and within three months. Recurrences seem to be rare. In some cases, the headache may last for years.

Relief
Lying down. *Treatment:* Epidural blood patch, epidural saline infusion, high dose corticosteroids have been used with success in a few patients.

Complications
Usually none.

Social and Physical Disability Inability to sit or stay in the upright position because of the pain.

Pathology
Low CSF pressure demonstrated during spinal tap is essential for diagnosis. Lumbar isotope cisternography has given indications of a leakage through a nerve root sheath tear or hyperabsorption of CSF as possible causes of the low CSF pressure in a few patients. However, this can not be used as a diagnostic test.

Essential Features
Positional headache due to low CSF pressure occurring spontaneously or after mild incidents.

Differential Diagnosis
Low CSF pressure due to CSF leaks after major head trauma.

Code
023.X1 a

References
Fernandez E. Headaches associated with low spinal fluid pressure. Headache 1990;30:122–38.

Gaukroger PB, Brownridge P. Epidural blood patch in the treatment of low CSF pressure headache. Pain 1987;29:119–22.

Molins A, Alvarez J, Titus F, Codina A. Cisternographic pattern of spontaneous liquoral hypotension. Cephalalgia 1990;10:59–65.

Post-Dural Puncture Headache (V-14)

Definition
Dull, aching, or throbbing positional pain in the head occurring after dural puncture, most often in the

lumbar region

Site
Frontal, occipital, or global. May be unilateral.

System
Probably vascular and/or meningeal.

Main Features
Prevalence: occurs in 15-30% of patients who have been subject to lumbar puncture. *Sex Ratio:* women are affected twice as often as men. *Age of Onset:* relatively reduced frequency under 13 years and over 60 years. *Pain Quality:* usually dull or aching, but may be throbbing. *Precipitating Factors:* the pain is positional, markedly exacerbated or only present when the patient is sitting or standing, usually relieved by lying down. *Intensity:* from mild to rather severe, probably never excruciating. *Time Pattern:* headache usually starts within 48 hours after lumbar puncture, but it may be delayed up to 12 days.

Associated Symptoms and Signs
Frequently, the patient will have pain and stiffness in the neck and the low back. Nausea is also fairly common, whereas blurred vision, tinnitus, and vomiting occur more rarely.

Laboratory Findings
Often, but not invariably, a low CSF pressure (<_ 60 mm H2O) is found, provided a second lumbar puncture with the patient lying horizontally is carried out during a symptomatic period.

Usual Course
On average, symptoms persist for four days, but in some cases, the headache may be protracted (lasting even up to years).

Relief
Lying down. *Treatment:* Intravenous caffeine sodium benzoate, epidural blood patch, epidural saline infusion, surgical closure of dural leak.

Complications
Subdural hematoma or hygroma may rarely occur.

Social and Physical Disability
The patient may be unable to sit or stay in the upright position because of the pain.

Pathology
Sudden drop in CSF volume, usually, but not always, resulting in a low CSF pressure. Continuous leakage of CSF probably also plays a role.

Essential Features
Positional headache occurring after lumbar puncture.

Differential Diagnosis
Meningitis (bacterial or aseptic) occurring after lumbar puncture.

Code
023 .X l b

References

Tourtellotte WW, Haerer AF, Heller GL, Somers JE. Post-Lumbar Puncture Headaches. Springfield: CC Thomas Publisher; 1964.

Vilming ST, Schrader H, Monstad I. The significance of age, sex and cerebrospinal fluid pressure in post-lumbarpuncture headache. Cephalalgia 1989;9:99–106.

Hemicrania Continua (V-15)

Definition

Unilateral dull pain, occasionally throbbing, initially intermittent but later frequently a continuous headache of moderate to severe degree, sometimes with superimposed stabbing pains. Usually, there are some autonomic symptoms and signs. There is a clear female preponderance, and the headache responds completely to indomethacin.

Site

The headache is strictly unilateral, and in general without change of side. The maximum pain is usually in the ocular and fronto-temporal areas.

System

Unknown.

Main Features

Prevalence: not known, probably not frequent but may be more frequent than the other headache, completely responsive to indomethacin, i.e., chronic paroxysmal hemicrania (CPH). *Sex Ratio:* female to male about 5:1. *Age of Onset:* mean about 35, range 11-57 years of age. *Pain Quality:* dull, during exacerbations, occasionally throbbing. Considerable fluctuations in pain, even during the late, nonremitting stage. Most patients experience occasional or more frequent "jabs and jolts." *Time Pattern:* the chronic, nonremitting stage so typical of this headache is frequently preceded by a remitting stage (in approximately half the cases) of varying duration. During the remitting stage, there may be repetitive, separate attacks lasting hours or days. During the nonremitting stage, when the pain is more or less continuous, exacerbations occur, lasting a few hours to 4-5 days. *Intensity:* usually moderate to severe, with rather marked fluctuations; patients are usually able to cope with daily chores. Occasional nighttime awakening due to pain.

Precipitating Factors

Attacks or exacerbations are not known to be precipitated mechanically.

Associated Symptoms and Signs

Photophobia, phonophobia, nausea, conjunctival injection, and lacrimation (the last two on the symptomatic side) occur in up to half the cases, but these symptoms and signs generally are mild and usually only become clinically apparent during exacerbations.

Relief

Immediate, absolute, and permanent relief from continued indomethacin administration in adequate dosages.

Usual Course

The unremitting course may apparently continue for a long time, perhaps indefinitely. Once the chronic stage has been reached, no exceptions to this rule have been observed so far.

Complications
In a few instances, suicide attempts due to headache.

Social and Physical Disability
Considerable during exacerbations.

Pathology
Not known. "Symptomatic" cases have been observed, e.g., with tumor of osseous structures. When atypical features occur or when the indomethacin effect is incomplete or fading, such a possibility should be suspected.

Essential Features
Remitting or nonremitting unilateral headache, occurring mostly in the female, with the pain maximum in the oculo-fronto-temporal area, the pain being of moderate to severe degree. There may be moderate autonomic signs. Absolute and permanent indomethacin effect.

Differential Diagnosis
The other unilateral headache with absolute indomethacin response, CPH; other unilateral headaches such as Costen's syndrome, sinusitis, dental pain, and earache (in the remitting stage of hemicrania continua); cervicogenic headache. (Note the following points of differential diagnostic importance. HC: *complete* indomethacin response. Cervicogenic headache: reduced range of motion in the neck; ipsilateral, diffuse, nonradicular shoulder/arm symptoms; mechanical precipitation of attacks; absolute effect of major occipital nerve blockade.)

Code
093.X8

References
Bordini C, Antonaci F, Stovner LJ, Schrader H, Sjaastad O. Hemicrania continua: a clinical review. Headache 1991;31:20–6.

Sjaastad O, Spierings ELH. "Hemicrania continua": another headache absolutely responsive to indomethacin. Cephalalgia 1984;4:65–70.

Headache Not Otherwise Specified (V-16)

Code
OOX.X8f

HEADACHE CROSSWALK

The classification of headache of the International Headache Society appeared in 1988 (International Headache Society, Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain, Cephalalgia, 8, Suppl. 7 [1988]). That system differs from the IASP classification in several respects. This list, which follows the first six sections (Groups II through VII) in which headache specifically appears in this volume, refers also to Groups IX-1 (IX-1.7 to IX-1.11) and IX-8. It is intended to provide a statement, where possible, of the correspondence between the categories of the IHS system and the IASP system. Because the structures of the two systems differ significantly, correspondence is often not easy to determine or is definitely not available. The principal feature of the structures which provides this problem is that the IASP system for head, face, and neck, follows the same pattern as that used in other parts of the body, i.e., proceeding through neurological, musculoskeletal, and visceral disorders as well as miscellaneous conditions. Some phenomena are also described in relation to the cervical spine. The IHS system also includes a number of acute categories that are lacking by design in the IASP system, and the IASP system contains categories that were not adopted by the IHS in 1988, but which should be adopted at this point and have no exact IHS equivalent.

All the IASP categories are printed in bold, as are those IHS syndromes for which the correspondence appears to be fairly good. The crosswalk is from the IASP system to the IHS system and not in reverse. Where the only corresponding item is a "catch-all" or residual category, an entry is not necessarily made.

IASP		HIS	
1-6	**Central Pain (if confined to head and face)**	**12.7.2**	**Thalamic pain**
II-1	**Trigeminal neuralgia (tic douloureux)**	**12.2.1**	
11-2 or	**Secondary neuralgia (trigeminal) from central nervous system lesions (tumor aneurysm)**	**12.2.2.2**	**Symptomatic trigeminal neuralgia: central lesions**
11-3	**Secondary trigeminal neuralgia from facial trauma**	**12.2.2**	**Symptomatic trigeminal neuralgia**
11-4	**Acute herpes zoster (trigeminal)**	**12.1.4.1**	**Herpes zoster**
11-5	**Postherpetic neuralgia (trigeminal)**	**12.1.4.2**	**Chronic postherpetic neuralgia**
11-6	**Geniculate neuralgia (VIth cranial nerve): Ramsay Hunt syndrome**	**12.1.4.1**	**Herpes zoster**
11-8	**Glossopharyngeal neuralgia (IXth cranial nerve)**	**12.3.1** **neuralgia** **12.3.2** *neuralgia*	**Idiopathic glossopharyngeal** *Symptomatic glossopharyngeal*
11-9	**Neuralgia of the superior laryngeal nerve (vagus nerve neuralgia)**	**12.5**	**Superior laryngeal neuralgia**
11-10	**Occipital neuralgia**	**12.6**	**Occipital neuralgia**
11-11	**Hypoglossal neuralgia**	**12.1.7** *cranial*	*Other causes of persistent pain of nerve origin*
11-12	**Glossopharyngeal pain from trauma**	**12.3.2** **neuralgia**	**Symptomatic glossopharyngeal**
11-12	**Hypoglossal pain from trauma**	**12.1.7** *cranial*	*Other causes of persistent pain of nerve origin*

11-14 Tolosa-Hunt syndrome (painful ophthalmoplegia)	**12.1.5 Tolosa-Hunt syndrome**

IASP	HIS
111-2 Tension headache: chronic form (scalp muscle contraction headache	**2.2 Chronic tension-type headache** **2.3 Headache of the tension type**
111-3 Temporomandibular pain and dysfunction syndrome	**2.3.2** *Headache of the tension type with oromandibular dysfunction*
111-5 Rheumatoid arthritis of the temporomandibular joint	***11.7*** *Temporomandibular joint disease*
IV-1 Maxillary sinusitis	**11.5.1 Acute sinus headache**
IV-2 through IV-5 **Types of odontalgia**	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-6 Glossodynia and sore mouth	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-7 Cracked tooth syndrome	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-8 Dry socket	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-9 Gingival disease, inflammatory	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-10 Toothache, cause unknown	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-11 Diseases of the jaw, inflammatory conditions	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
IV-12 Other and unspecified pain in jaws	**11.6** *Headache or facial pain associated with disorder of teeth, mouth, or other facial* *or* *cranial structures*
V-1 Classic migraine (migraine with aura)	**1.2.1 Migraine with aura** **1.2.2** **1.2.6** **1.6.1**
V-2 Common migraine (migraine without aura)	**1.1 Migraine without aura**
V-3 Migraine variants	**1.2.3 Familial hemiplegic migraine** **1.2.4 Basilar migraine** **1.3 Ophthalmoplegic migraine**

	1.4 Retinal migraine
V-5 Mixed headache	**1.1 Migraine without aura** **2.2 Chronic tension type headache**
V-6 Cluster headache	**3.1.1 Cluster headache, periodicity undetermined** **3.1.2 Episodic cluster headache**
V-7.1 Chronic paroxysmal hemicrania: unremitting form or variety	**3.2 Chronic paroxysmal hemicrania**
IASP	**HIS**
V-7.2 Chronic paroxysmal hemicrania: remitting form or variety	***3.2 Chronic paroxysmal hemicrania***
V-8 **Chronic cluster headache**	**3.1.3 Chronic cluster headache**
V-10 Post-traumatic headache	**5.2.1 Chronic post-traumatic headache with significant head trauma or confirmatory signs 5.2.2 Minor head trauma with no confirmatory signs**
V-11 Syndrome of "jabs and jolts"	**4.1** *Idiopathic stabbing headaches*
V-12 Temporal arteritis (giant cell arteritis)	**6.5.1 Giant cell arteritis**
V-13 Headache associated with low cerebrospinal fluid pressure	***7.2.2*** *Cerebrospinal fluid fistula headache*
V-14 Post-dural puncture headache	**7.2.1 Post lumbar puncture headache**
V-16 Headache not otherwise specified	**13.0 Headache, not classifiable**
VI-2 Hysterical or hypochondriacal pain in the head, face, and neck	**2.3.3** ***Headache of the tension type, not fulfilling above criteria***
VI-3 Headache of psychological origin in the head, face, and neck associated with depression	**2.3.3** ***Headache of the tension type, not fulfilling above criteria***
VII-2 Cervicogenic headache	**11.2.1** ***Headache or facial pain associated with disorder of cranium, neck, etc.***
IX-1 **Cervical spinal or radicular pain attributable to a fracture**	**11.2.1** ***Headache or facial pain associated with disorder of cranium, neck, etc.***
IX-1.7 Fracture of lamina	**11.2.1** ***Headache or facial pain associated with disorder of cranium, neck, etc.***
IX-1.9 **Fracture of the anterior arch of the atlas**	**11.2.1** ***Headache or facial pain associated with disorder of cranium, neck, etc.***
IX-1.10 Fracture of the posterior arch of the atlas	**11.2.1** ***Headache or facial pain associated with disorder of cranium, neck, etc.***
IX-1.11 Burst fracture of the atlas	**11.2.1** ***Headache or facial pain associated with disorder of cranium, neck, etc.***
NOTE: Other items in the neck are not included, although they may potentially cause headache; if they do, they can be entered in the relevant section of the cervical spinal items	
IX-8 **Acceleration-deceleration injury of the neck (cervical sprain)**	**5.2.2 Minor head trauma with no confirmatory signs**

GROUP VI: PAIN OF PSYCHOLOGICAL ORIGIN IN THE HEAD, FACE, AND NECK

As for 1-16 with local distribution.

Delusional or Hallucinatory Pain (VI-1)

Differential diagnosis from local and general conditions.

Code	
O1X.X9e	Head or face
11X.X9e	Neck

Hysterical, Conversion, or Hypochondriacal Pain (VI-2)

Distribution possibly more often on the left, except in cases with lesions or compensation claims. Differential diagnosis from local conditions (see above) and general conditions, e.g., hypothyroidism, polyarthralgia, etc., which cause diffuse symptoms.

Code	
O1X.X9f	Head or face
11X.X9f	Neck

Associated with Depression (VI-3)

Code	
O1X.X9g	Head or face
11X.X9g	Neck

GROUP VII: SUBOCCIPITAL AND CERVICAL MUSCULOSKELETAL DISORDERS

(See also Group IX; for Cervical Sprain see IX-8, Acceleration-Deceleration Injury of the Neck.)

Stylohyoid Process Syndrome (Eagle's Syndrome) (VII-1)

Definition
Pain following trauma in the region of a calcified stylohyoid ligament.

Site
Mandible, floor of mouth, lateral pharynx.

System
Musculoskeletal system.

Main Features
Prevalence: among patients with calcified stylohyoid ligament and history of trauma to mandible and/or neck. *Sex Ratio:* no predilection. *Age of Onset: 40-50* years. *Start:* evoked by swallowing, opening mandible, turning head toward pain and down, with palpation of stylohyoid ligament. *Pain Quality:* throbbing, deep. *Occurrence:* with function. *Intensity:* mild to moderate. *Duration:* seconds to minutes.

Associated Symptoms
Dizziness, tenderness on palpation of the carotid trunk and branches.

Signs
Carotid bruit, transient ischemic episodes.

Radiologic Findings
Calcified stylohyoid process.

Usual Course
Benign, intractable if styloid process not excised or fractured, partial relief from stellate ganglion local anesthetic infiltration, and acetylsalicylic acid.

Complication
Secondary carotid arteritis and cerebral ischemia.

Social and Physical Disability
Interference with speech and mastication.

Pathology
Calcified stylohyoid ligament, carotid-external carotid branch arteritis.

Summary of Essential Features and Diagnostic Criteria
Presence of calcified stylohyoid ligament, tenderness of superficial vessels, history of trauma.

Differential Diagnosis
Myofascial pain dysfunction, carotid arteritis, glossopharyngeal neuralgia, tonsillitis, parotitis, mandibular osteomyelitis.

Code
036.X6

Cervicogenic Headache (VII-2)

Definition
Attacks of moderate or moderately severe unilateral head pain without change of side, ordinarily involving the whole hemicranium, usually starting in the neck or occipital area, and eventually involving the forehead and temporal areas, where the maximal pain is frequently located. The headache usually appears in episodes of varying duration in the early phase, but with time the headache frequently becomes more continuous, with exacerbations and remissions. Symptoms and signs such as mechanical precipitation of attacks imply involvement of the neck.

Site
Whole hemicranium. The pain usually starts in the neck or back of the head but soon moves to the frontal and temporal areas. It occasionally extends into the infraorbital area. Unilaterality without alternation of sides is typical, but occasionally moderate involvement of the opposite side occurs during the most severe attacks. Bilateral cases certainly exist and may be quite frequent. At the present time, however, scientific studies should preferably include only unilateral cases. Frequently, diffuse ("nonradicular") pain or discomfort occurs in the ipsilateral shoulder and arm.

System
Probably the peripheral nervous system. Musculoskeletal system is probably also involved.

Main Features
Prevalence: probably rather frequent, but exact figures are lacking. *Sex Ratio:* probably less than 3/4 of the patients are female. *Age of Onset:* young adult or middle age. Many of the patients have sustained neck trauma a relatively short time prior to the onset. *Pain Quality:* constant, deep, dull, steady, not excruciating pain. Pain seemingly identical, may be triggered by neck movements or by external pressure over the greater occipital nerve (GON). *Time Pattern:* pain episodes are of greatly varying duration, from hours to weeks, even intraindividually, the usual duration being one to a few days. The varying duration of attacks is a characteristic feature of this headache. Interval between pain episodes: days to weeks. In the later phase, there is characteristically a protracted or continuous, low-intensity pain, with superimposed exacerbations. *Intensity:* moderate to severe pain.

Precipitating Factors
Pain similar to that of the "spontaneous" pain episodes or even attacks may be precipitated by awkward neck movements or awkward positioning of the head during sleep. Also by external pressure over the GON on the symptomatic side.

Associated Symptoms
More rarely the symptoms include: nausea, vomiting, phonophobia and photophobia (usually of a low degree), dizziness, "blurred vision" (longlasting) on the symptomatic side, and difficulties in swallowing.

Signs
Reduced range of motion in the neck, in one or more directions. Occasionally, edema and redness of the skin below the eye on the symptomatic side.

Tests and Laboratory Findings
A blockade of the greater occipital nerve (GON), the minor occipital nerve, the so-called IIIrd occipital

nerve, or the cervical nerve roots should be carried out on the symptomatic side. Such blockades reduce or take away the pain transitorily, not only in the anesthetized area (the innervation area of the respective nerve) but also in the nonanesthetized, painful Vth nerve area. This represents a diagnostic test.

Relief
Repeated corticosteroid injections along the GON may provide relief of some duration. Neurolysis ("liberation operation") of GON may provide longlasting relief (1/3 to more than 2 years), but it rarely, if ever, provides *permanent* relief. There are reasons to believe that denervation of the periosteum of the occipital area on the symptomatic side may provide permanent relief in a high percentage of the cases.

Usual Course
Persistence and intensification of the pain syndrome over time.

Complications
Combination with root pain into shoulder/arm.

Social and Physical Disability
Patients can frequently do some routine work during symptomatic periods. In the worst periods, total disability.

Pathology
Probably related to various structures in the neck or posterior part of the scalp on the symptomatic side (C2/C3 innervation area), but cannot at present be precisely identified. Although the clinical picture is identifiable and rather stereotyped, the pathology varies in that pathology in the lower part of the neck may also be the underlying cause.

Essential Features
Combination of unilateral headache, ipsilateral diffuse shoulder or arm pain, reduced range of motion in the neck, presence of mechanical precipitation mechanisms, and discontinuation of the pain upon anesthetic blockades (GON, C2 etc.) in the typical case. Frequently there is a history of neck injury.

Differential Diagnosis
Common migraine, hemicrania continua, spondylosis of the cervical spine. Other unilateral headaches, such as cluster headache, are less important in this respect. Tension headache (as regards the bilateral variant of cervicogenic headache).

Code
033.X6b

References
Bogduk N, Marsland H. On the concept of third occipital headache. J Neurol Neurosurg Psychiatry 1986;49:775–80.

Fredriksen, TA. Studies on Cervicogenic Headache: Clinical Manifestation and Differentiation from Other Unilateral Headache Forms (thesis). Trondheim, Norway: Tapir; 1989.

Sjaastad O, Fredriksen TA, Pfaffenrath V. Cervicogenic headache: diagnostic criteria. Headache 1990;30:725–6.

Sjaastad O, Sante C, Hovdal H, Breivik H, Gronbaek E. "Cervicogenic" headache: an hypothesis. Cephalalgia 1983;3:249–56.

Superior Pulmonary Sulcus Syndrome (Pancoast Tumor) (VII-3)

Definition

Progressively intense pain in the shoulder and ulnar side of the arm, associated with sensory and motor deficits and Homer's syndrome due to neoplasm.

Continuous aching pain in the paraspinal region, shoulder, or elbow, in time expanding to the whole ulnar side of the arm. Exacerbations of sharp lancinating pain in the region of the lower brachial plexus. Often radiological evidence of a tumor in the apex of the lung.

Site
Shoulder and upper limb.

System
Nervous system.

Main Features
Sex Ratio: males more than females. *Age of Onset:* usually in the decades corresponding with the occurrence of carcinoma of the lung. *Pain Quality:* the pain is continuous, involving the root of the neck and ulnar side of the upper limb. It is usually progressive, requiring narcotics for relief, and becomes excruciating unless properly managed. The lesion is involvement of the VIIIth cervical and Ist thoracic roots. The pain is a severe aching and burning associated with sharp lancinating exacerbations. There is paralysis and atrophy of the small muscles of the hand and a sensory loss corresponding to the pain distribution.

Associated Symptoms
The cervical sympathetic is involved with a Homer's syndrome.

Signs and Laboratory Findings
Atrophy of the small muscles of the hand, ulnar sensory loss, ulnar paresthesias and pain, and Homer's syndrome. The diagnosis is made on chest X-ray by the appearance of a tumor in the superior sulcus. Electromyography will demonstrate denervation in the appropriate distribution.

Usual Course
The course is generally relentless and the prognosis poor.

Complications
Occasional infiltration of spinal cord with compression. Occasional hoarseness from infiltration of the laryngeal nerves.

Social and Physical Disability
Those related to the neurological loss, unemployment, and family stress.

Pathology or Other Contributory Factors
Virtually always carcinoma of the lung, though any tumor metastatic to the area may give identical findings.

Summary of Essential Features and Diagnostic Criteria
The essential features are unremitting, aching pain of increasing severity, in time expanding to the ulnar side of the arm with exacerbations of sharp lancinating pain in the distribution of the lower brachial plexus. Homer's syndrome occurs associated with damage to T1 and C8 and occasional neurological loss; the diagnosis is made by chest X-ray demonstrating tumor at the apex of the lung, and the biopsy is made by tumor.

Code

102.X4a

Reference
Bonica JJ, Ventafridda V, Pagni CA. Management of superior pulmonary sulcus syndrome (Pancoast syndrome). In: Bonica JJ, Ventafridda V, Pagni CA, editors. Advances in Pain Research and Therapy, Vol. 4. New York: Raven Press; 1982.

Thoracic Outlet Syndrome (VII-4)
(includes Scalenus Anticus Syndrome, Cervical Rib Syndrome)

Definition
Pain in the root of the neck, head, shoulder, radiating down the arm into the hand. Due to compression of the brachial plexus by hypertrophied muscle, congenital bands, post-traumatic fibrosis, cervical rib or band, or malformed first thoracic rib.

Site
Ipsilateral side of head, neck, arm, and hand.

System Involved
Musculoskeletal system.

Main Features
Sex Ratio: there is no sexual predilection. *Age of Onset:* the thoracic outlet syndrome is characteristically found in young to middle-aged adults but may affect older adults also. *Pain Quality:* typically, pain begins in the root of the neck, or shoulder, and radiates down the arm, but it may also affect the head. The ulnar aspect of the arm is the most commonly involved, but the pain may affect the entire arm. Paresthesias are common in the same distribution. The pain occurs irregularly, usually with activity. The pain in the hand or the arm is not usually intense, but the associated headache may be severe. When the pain occurs, it usually diminishes with rest.

The distribution of the paresthesias or pain in the shoulder or arm is varied and can be associated with a particular nerve root, or with many nerve roots. Often it is rather baffling in that it cannot readily be related to specific nerves or nerve roots.

Associated Symptoms
Raynaud's phenomenon involving the same extremity is common. Hemiplegia from stroke secondary to vascular thrombosis and propagation of the clot may occur. The pain is generally aggravated by exercise and relieved by rest. A dystrophic sympathetic change may also occur.

Rarely, peripheral vascular insufficiency syndromes are found, and occasionally, the subclavian axillary vein complex can be compressed, and the patient presents with swelling and blueness consistent with symptoms of venous obstruction.

Signs and Laboratory Findings
Postural abnormalities are common. Three physical findings are frequent: pain on pressure over the brachial plexus, just lateral to the scalenus anticus muscle; pain mimicked by abduction and external rotation of the arm; and pain when the brachial plexus is stretched by tipping the head to the opposite side. Color change may also appear with other maneuvers, e.g., bracing back the shoulders. The classic sign is Adson's maneuver. This is performed by maximal extension of the chin and deep inspiration with the shoulders relaxed forward and the head turned towards the suspected side of abnormality. Obliteration

of the pulse, or at least diminution, should occur. This sign is not always found and may occur in normal individuals also.

Laboratory findings are often not helpful. Angiograms are indicated when there is an arterial or venous obstruction but are very poor diagnostic maneuvers, the milder forms of the thoracic outlet syndrome only affecting neurological symptoms. Electromyography may demonstrate evidence of nerve root compression across the thoracic outlet and denervation distally in the arm, but often fails to do so.

Usual Course
The usual course is one of continued persistent discomfort. Physiotherapy may strengthen the shoulder girdle and relieve symptoms, and this should be tried at first, but ordinarily symptoms will persist until the entrapment of the plexus is relieved.

Complications
Complications include arterial compression with thrombosis and an ischemic arm. Axillary subclavian vein thrombosis may also occur separately, or in addition.

Pathology
A variety of anatomical abnormalities will compress the neurovascular bundle at the thoracic outlet and may cause this syndrome. It may be precipitated in predisposed individuals by flexion-extension injuries of the cervical spine with consequent postural or other change. This is a late sequel of such injuries.

Social and Physical Disabilities
The patients are often unable to work because of dysfunction of the extremity involved.

Summary of Essential Features and Diagnostic Criteria
Patients with this syndrome suffer from compression of the brachial plexus for which many causes exist. Characteristically, they develop pain and paresthesias in the upper extremity, sometimes associated with headache. The most common diagnostic criteria are tenderness over the brachial plexus in the neck, reproduction of the pain by the maneuver of abduction and external rotation of the arm, and pain on stretching the brachial plexus.

Differential Diagnosis
Differential diagnosis includes cervical rib, cervical osteoarthritis, Pancoast's tumor, aneurysm of the subclavian artery, tumors of the brachial plexus, cervical disk, adenopathy or tumor of other supraclavicular structures, metastatic cancer to the cervical spine.

Code
133.X6d 233.X6a

Cervical Rib or Malformed First Thoracic Rib (VII-5)

It is impossible to differentiate the scalenus anticus syndrome (VII-4) from cervical or malformed first thoracic rib, except by X-ray. The presentations are identical. The diagnosis and differential diagnoses are the same. The only variation from the scalenus anticus syndrome is the finding of the abnormal or deformed rib on X-ray. The code is the same and the reference for this syndrome is the same.

Pain of Skeletal Metastatic Disease of the Neck, Arm, or Shoulder Girdle (VII-6)

Definition
Dull aching pain in the shoulder girdle or upper extremity due to tumor infiltration of bone.

Site
Clavicle, scapula, humerus.

System
Skeletal system.

Main Feature
Age of Onset: usually in the fifth, sixth, and seventh decades-corresponding to the occurrence of carcinoma of the lung, breast, and prostate. *Pain Quality:* The pain is usually described as a continuous dull ache or a constant throb. It may radiate up into the neck or down into the anterior chest wall. An expanding lesion in the humerus may radiate into the forearm. The cardinal feature is acute exacerbation of the pain by any movement of the shoulder girdle.

Associated Symptoms
Pain at rest usually responds to nonsteroidal antiinflammatory drugs and narcotic analgesics. Pain secondary to movement is sometimes relieved by internal fixation. Both types of pain may respond to radiation therapy.

Signs and Laboratory Findings
The active range of movement of the shoulder girdle is usually much more limited than the passive range of movement. Well-localized bony tenderness is common. Neurological signs are unusual. A radioisotope bone scan is usually positive before a plain X-ray. However, both of these tests may be normal in the setting of severe pain.

Complications
The tendency to keep the upper extremity immobilized may result in a "frozen shoulder," with secondary pain on that basis. A pathological fracture in the shaft of the humerus severely exacerbates pain on movement, and this usually requires treatment with internal fixation.

Social and Physical Disability
There may be loss of use of the involved upper extremity.

Summary of Essential Features and Diagnostic Criteria
Continuous aching pain, exacerbation of the pain by movement, localized bony tenderness at the site of metastatic deposit.

Differential Diagnosis
It is important to rule out referred pain to the shoulder girdle and upper extremity due to tumor infiltration of the cervical roots and brachial plexus.

Code
133.X4j
233.X4

GROUP VIII: VISCERAL PAIN IN THE NECK

Carcinoma of Thyroid (VIII-1)

Definition
Pain in the thyroid gland, aggravated by palpation and associated with an adherent neoplastic mass.

Site
Throat and anterior neck area, spreading to the ear.

System
Endocrine system.

Main Features
Localized sharp or dull, aching or burning, occasionally stabbing if superior laryngeal nerve involved.

Associated Symptoms
Mass in neck, dysphagia, dyspnea or stridor, from secondary deposits.

Signs
Neck swelling, fixation of thyroid, stridor.

Laboratory Findings
Cold nodule on scan.

Complications
Local-dysphagia; stridor.

Code
172.X4

Carcinoma of Larynx (VIII-2)

Definition
An aching soreness in the throat, aggravated by swallowing, with hoarseness and dysphagia.

Site
Larynx and adjoining portions of neck.

System
Respiratory system.

Main Features
Initially, there is a complaint of sore throat, with irritation, which becomes a severe soreness. Later, pain may develop on swallowing. The pain spreads to the ear (otalgia), possibly because of the involvement of the vagus nerve. The pain is usually moderately severe, dull, aching, burning in character, occasionally sharp, stabbing, or lancinating if the superior laryngeal nerve is involved.

Associated Symptoms
Hoarseness; dysphagia, when local spread has occurred.

Signs
Tumor on inspection of larynx.

Complications
Stridor progressing to respiratory obstruction; dysphagia, when local spread has occurred.

Social and Physical Disability
Loss of voice following surgical treatment.

Essential Features
Persistent hoarseness, with soreness or pain supervening.

Code
122.X4

Tuberculosis of Larynx (VIII-3)

Definition
A painful irritation in the throat on air flow during breathing, coughing, and swallowing due to tuberculous lesions.

Site
Larynx and adjoining regions of neck.

System
Respiratory system.

Main Features
Now rare. Local in larynx; spreads to ear (otalgia); continuous, dull, aching, burning, stabbing, or lancinating if superior laryngeal nerve involved. Worse on swallowing. N.B.: In early stage is pain free. In advanced cases there is severe pain in the laryngeal and pharyngeal area, which may radiate to the ear.

Associated Symptoms
Hoarseness; cough; purulent sputum; night sweats and fever; weight loss.

Signs
Inflammation of larynx; ulceration of larynx; chest signs.

Pathology
Infection with *Mycobacterium tuberculosis.*

Summary of Essential Features and Diagnostic Criteria
Hoarseness in someone with tuberculosis of chest, i.e., cough, sputum, night sweats, and weight loss, with pain supervening.

Differential Diagnosis
Cancer of larynx.

Code
123.X2

Chronic Pharyngitis (VIII-4)

Code
151.X5 If known

151.X8 Alternative

Carcinoma of Pharynx (VIII-5)

Code
153.X4