A. RELATIVELY GENERALIZED SYNDROMES

GROUP I: RELATIVELY GENERALIZED SYNDROMES

Peripheral Neuropathy (I-1)

Definition
Constant or intermittent burning, aching, or lancinating limb pains due to generalized or focal diseases of peripheral nerves.

Site
Usually distal (especially the feet) with burning pain, but often more proximal and deep with aching. Focal with mononeuropathies, in the territory of the affected nerve (e.g., meralgia paresthetica).

System
Peripheral nervous system.

Main Features
Prevalence: common in neuropathies of diabetes, amyloid, alcoholism, polyarteritis, Guillain-Barre Syndrome (for which see I-36), neuralgic amyotrophy, Fabry's disease. *Age of Onset:* variable, usually after second decade. *Pain Quality:* (a) burning, superficial, distal pain often with dysesthesia, constant. May be in the territory of a single affected nerve; (b) deep aching, especially nocturnal, constant; and (c) sharp lancinating "tabetic" pains, especially in legs, intermittent.

Associated Symptoms
Sensory loss, especially to pinprick and temperature; sometimes weakness and muscle atrophy (especially in neuralgic amyotrophy); sometimes reflex loss; sometimes signs of loss of sympathetic function; smooth, fine skin; hair loss.

Laboratory Findings
(a) Features of the primary disease, e.g., diabetes; and
(b) Features of neuropathy: reduced or absent sensory potentials, slowing of motor and sensory conduction velocities, EMG evidence of muscle denervation.

Usual Course
Distal burning and deep aching pains are often long-lasting, and the disease processes are relatively unresponsive to therapy. Pain resolves spontaneously in weeks or months in self-limited conditions such as Guillain-Barre syndrome or neuralgic amyotrophy.

Complications
Drug abuse, depression.

Social and Physical Disabilities
Decreased mobility.

Pathology
Nerve fiber damage, usually axonal degeneration. Pain especially occurs with small fiber damage (sensory fibers). Nerve biopsy may reveal the above, plus features of the specific disease process, e.g., amyloid.

Summary of Essential Features and Diagnostic Criteria
Chronic distal burning or deep aching pain with signs of sensory loss with or without muscle weakness, atrophy, and reflex loss.

Differential Diagnosis
Spinal cord disease, muscle disease.

Code

203.X2a	Arms: infective
203.X3a	Arms: inflammatory or immune reactions
203.X5a	Arms: toxic, metabolic, etc.
203.X8a	Arms: unknown or other
603.X2a	Legs: infective
603.X3a	Legs: inflammatory or immune reactions
603.X5a	Legs: toxic, metabolic, etc.
603.X8a	Legs: unknown or other
X03.X4d	Von Recklinghausen's disease

References
Asburn AK, Fields HL. Pain due to peripheral nerve damage: an hypothesis. Neurology 1984;34:1587–90.

Thomas PK. Pain in peripheral neuropathy: clinical and morphological aspects. In: Ochoa J, Culp W, editors. Abnormal Nerves and Muscles as Impulse Generators. New York: Oxford University Press; 1982.

Stump Pain (I-2)

Definition
Pain at the site of an extremity amputation.

Site
Upper or lower extremity at the region of amputation. Pain is not referred to the absent body part but is perceived in the stump itself, usually in region of transected nerve(s).

System
Peripheral nervous system; perhaps central nervous system.

Main Features
Sharp, often jabbing pain in stump, usually aggravated by pressure on, or infection in, the stump. Pain often elicited by tapping over neuroma in transected nerve or nerves.

Associated Symptoms
Refusal to utilize prosthesis.

Signs
Pain elicited by percussion over stump neuromata.

Laboratory Findings
None.

Usual Course
Develops several weeks to months after amputation; persists indefinitely if untreated.

Relief
(a) Alter prosthesis to avoid pressure on neuromata; (b) resect neuromata so that they no longer lie in pressure areas; and (c) utilize neurosurgical procedures such as rhizotomy and ganglionectomy or spinal cord or peripheral nerve stimulation in properly selected patients.

Complications
Refusal to use prosthesis.

Social and Physical Disabilities
Severe pain can preclude normal daily activities; failure to utilize prosthesis can add to functional limitations.

Pathology
Neuroma at site of nerve transection.

Essential Features
Pain in stump.

Differential Diagnosis
Phantom limb pain, radiculopathy.

Code

203.Xla	Arms
603.Xla	Legs

Phantom Pain (I-3)

Definition
Pain referred to a surgically removed limb or portion thereof.

Site
In the absent body part.

System
Central nervous system.

Main Features
Follows amputation, may commence at time of amputation or months to years later. Varies greatly in severity from person to person. Reports of prevalence vary from < 1% to > 50% of amputees. Believed to be more common if loss of limb occurs later in life, in limbs than in breast amputation, in the breast before the menopause rather than after it, and particularly if pain was present before the part was lost. Pain may be continuous, often with intermittent exacerbations. Usually cramping, aching, burning; may have superimposed shocklike components. Seems to be less likely if the initial amputation is treated actively and a prosthesis is promptly utilized. Phantom limb pain is almost always associated with distorted image of lost part.

Associated Symptoms
Aggravated by stress, systemic disease, poor stump health.

Signs
Loss of body part.

Usual Course
Complaints persist indefinitely; frequently with gradual amelioration over years.

Relief
No therapeutic regimen has more than a 30% long-term efficacy. TENS, anticonvulsants, antidepressants, or phenothiazines may be helpful. Sympathectomy or surgical procedures upon spinal cord and brain, including stimulation, are sometimes helpful.

Social and Physical Disabilities
May preclude gainful employment or normal daily activities.

Pathology
Related to deafferentation of neurons and their spontaneous and evoked hyperexcitability.

Essential Features
Pain in an absent body part.
Differential Diagnosis Stump pain.

Code

203.X7a	Arms
603.X7a	Legs

Complex Regional Pain Syndromes (CRPS), Type I (I-4) and Type II (I-5)

Definition
CRPS is a syndrome characterized by a continuing (spontaneous and/or evoked) regional pain that is seemingly disproportionate in time or degree to the usual course of pain after trauma or other lesion. The pain is regional (not in a specific nerve territory or dermatome) and usually has a distal predominance of abnormal sensory, motor, sudomotor, vasomotor edema, and/or trophic findings. The syndrome shows variable progression over time. CRPS type I develops after any type of trauma, especially fracture, soft tissue lesion (see below). CRPS type II occurs after major nerve damage.

Site
Usually the distal aspect of an affected extremity or with a distal to proximal gradient. In CRPS type II (with major nerve damage; defined below) the region of pain may initially correspond to a single peripheral nerve distribution but may become more diffuse over time.

System
Musculoskeletal system, peripheral nervous system and central nervous system.

Main Features
Pain often, but not always, follows trauma, which may be mild or may be associated with significant nerve injury in the case of CRPS type II. It may follow any type of trauma, especially fracture, soft tissue

lesion (e.g. crush injury), laceration, immobilization, or may be related to visceral disease, e.g., angina or central neurological disease such as stroke. The onset of symptoms usually occurs within one month of the inciting event. The pain is frequently described as burning and continuous and is exacerbated by movement, mechanical or thermal stimulation, or stress. The intensity of pain may fluctuate over time, and allodynia, and/or hyperalgesia may be found which are not limited to the territory of a single peripheral nerve. Abnormalities of blood flow occur, including changes in skin temperature and color. Edema is usually present and may be soft or firm. Increased or decreased sweating may appear. Dystrophic changes of skin, nails, hair, and bone may occur. Impairment of motor function and joint mobility are frequently seen and can include weakness, tremor, and, in rare instances, dystonia. The symptoms and signs may spread proximally or, rarely, spread to involve other extremities.

Associated Symptoms and Signs
Sympathetically maintained pain may be present and may be demonstrated with pharmacological blocking or provocation techniques. Affective symptoms or disorders may occur secondary to the pain and disability. Guarding of the affected part due to intense allodynia is usually observed.

Laboratory Findings
Noncontact skin temperature measurement usually indicates a side-to-side asymmetry of greater than 1 degree Celsius. Due to the unstable nature of the temperature changes in this disorder, measurements at different times are recommended. Measurements of skin blood flow may show an increase or a reduction. Testing of sudomotor function, both at rest and evoked, also may reveal side-to-side asymmetry. The bone uptake phase of a three-phase bone scan may reveal a characteristic pattern of subcutaneous blood pool changes. Radiographic examination may demonstrate patchy bone demineralization.

Usual Course
Variable.

Relief
Due to the complexity of the syndrome, the lack of information about specific mechanisms in CRPS, and its subsets as defined, and the liability of signs and symptoms, most authorities recommend a comprehensive and interdisciplinary approach. This may include physical, occupational, vocational, cognitive/behavioral, pharmacological, and anesthesiological/interventional (especially when the pain can be shown to be 'sympathetically maintained') strategies.

Complications
Phlebitis, cellulitis, atrophy, weakness, inappropriate drug use, depression and suicide. Permanent trophic changes of bone, joints and muscles as well as permanent functional disability can be seen.

Social and Physical Impairment
Inability to perform activities of daily living and occupational and recreational activities.

Pathology
Unknown. In CRPS II, the pain syndrome follows a major nerve injury, but that does not explain its pathological basis. Abnormal inflammatory responses are likely to play a role.

Diagnostic Criteria
There are two versions of the diagnostic criteria: A clinical version meant to maximize diagnostic sensitivity with adequate specificity, and a research version meant to more equally balance optimal sensitivity and specificity.

Clinical Diagnostic Criteria for CRPS

1) Continuing pain, which is disproportionate to any inciting event.

2) Must report at least one symptom in three of the four following categories:
Sensory: Reports of hyperalgesia and/or allodynia.
Vasomotor: Reports of temperature asymmetry and/or skin color changes and/or skin color asymmetry.
Sudomotor/Edema: Reports of edema and/or sweating changes and/or sweating asymmetry.
Motor/Trophic: Reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nails, skin).

3) Must display at least one sign* at time of evaluation in two or more of the following categories:
Sensory: Evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement).
Vasomotor: Evidence of temperature asymmetry and/or skin color changes and/or asymmetry.
Sudomotor/Edema: Evidence of edema and/or sweating changes and/or sweating asymmetry.
Motor/Trophic: Evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nails, skin).

4) There is no other diagnosis that better explains the signs and symptoms.

*A sign is counted only if it is observed at time of diagnosis.

**Research criteria for CRPS are recommended that are more specific, but less sensitive than the clinical criteria; they require that four of the symptom categories and at least two sign categories be present.

Subtypes of CRPS

CRPS I (old name: Reflex Sympathetic Dystrophy): As defined above.
CRPS II (old name: Causalgia): Defined as above with electrodiagnostic or physical evidence of a major nerve lesion.
CRPS-NOS* (Not Otherwise Specified): Partially meets CRPS criteria, not better explained by any other condition.
*This subtype was added to capture any patients previously diagnosed with CRPS who now do not meet criteria as elaborated above.

The evolution of terminology from two totally separate diseases, Causalgia and Reflex Sympathetic Dystrophy as in the Taxonomy of 1986, to CRPS I and CRPS II as in the Taxonomy of 1994 to the present CRPS with subtypes of I and II reflects changes in our understanding of potential mechanisms, clinical presentations and prognoses.

Differential Diagnosis
Unrecognized local pathology (e.g., fracture, strain, sprain), traumatic vasospasm, regional vascular disease, cellulitis, other regional infection, Raynaud's disease, thromboangiitis obliterans, thrombosis, specified neuropathy, erythromelalgia, specified regional motor disease, regional autoimmune process.

Code

203.Xlh	Arms
603.X1h	Legs

References

Harden RN, Bruehl S, Perez RS, Birklein F, Marinus J, Maihofner C, Lubenow T, Buvanendran A, Mackey S, Graciosa J, Mogilevski M, Ramsden C, Chont M, Vatine JJ. Validation of proposed diagnostic criteria (the ‘‘Budapest Criteria”) for complex regional pain syndrome. Pain 2010;150:268-74.

Harden RN, Bruehl SP. Diagnostic criteria: the statistical derivation of the four criterion factors. In: Wilson PR, Stanton-Hicks M, Harden RN. CRPS: Current diagnosis and therapy. Seattle: IASP Press; 2005. p. 45-58.

Central Pain (I-6)

Definition

Regional pain caused by a primary lesion or dysfunction in the central nervous system, usually associated with abnormal sensibility to temperature and to noxious stimulation.

Site

The regional distribution of the pain correlates neuroanatomically with the location of the lesion in the brain and spinal cord. It may include all or most of one side, all parts of the body caudal to a level (like the lower half of the body), or both extremities on one side. It may also be restricted simply to the face or part of one extremity.

System

Central nervous system.

Main Features

Age of Onset: all ages may be affected. The onset may be instantaneous but usually occurs after a delay of weeks or months, rarely a few years, and the pain increases gradually. *Pain Quality:* many different qualities of pain occur, the most common being burning, aching, pricking, and lancinating. Often the patient experiences more than one kind of pain. Dysesthesias are common. The pain is usually spontaneous and continuous, and exacerbated or evoked by somatic stimuli such as light touch, heat, cold, or movement. Some patients have no pain at rest but suffer from evoked pain, paresthesias, and dysesthesias. The pain can be augmented by startle stimuli (e.g., sudden sound or light), by visceral activity (e.g., micturition), or by anxiety and emotional arousal. The pain may be superficial or deep. *Intensity:* varies from mild but irritating to intolerable.

Associated Symptoms and Signs

There may be various neurological symptoms and signs such as monoparesis, hemiparesis, or paraparesis, together with somatosensory abnormalities in the affected areas. Impaired sensibility for temperature and noxious stimulation are leading signs. Increased threshold for at least one modality is most common, and this is frequently accompanied by dysesthetic or painful reactions to somatic stimuli, particularly touch and cold. Such reactions commonly meet the criteria for allodynia, hyperalgesia, and hyperpathia. In some patients it is difficult to show the altered sensibility with standard clinical tests. The threshold for tactile, vibration, and kinesthetic sensibility may be increased or normal.

Laboratory Findings

MRI or CT may show a relevant lesion.

Usual Course

In some cases improvement occurs with time, but in most patients the pain persists.

Relief

TENS may give relief in a few patients but can also transiently exacerbate the pain. Anticonvulsant drugs

help in some instances, especially carbamazepine and particularly for paroxysmal elements of the pain. Certain antidepressants (e.g., amitriptyline) seem to give the best relief, and some think that phenothiazines (e.g., chlorpromazine, fluphenazine) may be helpful.

Social and Physical Disabilities
This pain is a great physical and psychological burden to most patients. In consequence their social life and work are often much impaired. Allodynia in response to external stimuli and movements may hamper rehabilitation and prevent activities, thus making the patient physically handicapped.

Pathology
Cerebrovascular lesions (infarcts, hemorrhages), multiple sclerosis, and spinal cord injuries are the most common causes. Central pain is also common in syringomyelia, syringobulbia, and spinal vascular malformation, and may occur after operations like cordotomy. Increasing evidence indicates that central pain only occurs in patients who have lesions affecting the spino-thalamocortical pathways, which are important for temperature and pain sensibility. The lesion can be located at any level along the neuraxis, from the dorsal horn of the spinal cord to the cerebral cortex. The lesion sometimes may involve the medial lemniscal pathways.

Diagnostic Criteria
Regional pain attributable to a lesion or disease in the central nervous system and accompanied by abnormal sensibility for temperature and pain, most often hyperpathia.

Differential Diagnosis
Nociceptive, peripheral neurogenic, and psychiatric causes of pain should be excluded as far as possible. Sensory abnormalities will in most cases allow a diagnosis for positive reasons.

Code
If three or more major sites are involved, code first digit as 9:

903.X5c	Vascular
903.X l c	Trauma
903.X2c	Infection
903.X3c	Inflammatory
903.X4c	Neoplasm
903.X8c	Unknown

If only one or two sites are involved, code first digit according to specific site or sites; for example, for head or face, code 003.X5c.

Syndrome of Syringomyelia (I-7)

Definition
Aching or burning pain usually in a limb, commonly with muscle wasting due to tubular cavitation gradually developing in the spinal cord.

Site
Pain in shoulder, arm, chest, or leg, rarely in the face, occasionally bilateral.

System
Central nervous system.

Main Features
Pain is usually unilateral and continuous in an area that corresponds to the site of cavitation of spinal cord or brainstem, most frequently in the shoulder-girdle and arm. It may be a periodic diffuse dull ache but sometimes, and particularly when the pain is situated in forearm and hand, may have an intense burning quality. The pain may be severe and referred to deep structures in the limb, not responding to rest or minor sedation.

Associated Symptoms
Muscular weakness in affected region.

Signs
There is commonly muscle wasting beginning in small muscles of the hand and ascending to the forearm and shoulder-girdle with fasciculation and an early loss of tendon reflexes. Scoliosis kyphosis may occur. Characteristically, pain and temperature sensations are impaired but other sensations are intact. The area of sensory impairment typically has a shawl distribution over the front and back of the upper thorax. A Homer's syndrome may appear.

Usual Course
The disease usually begins in the second or third decade and slowly progresses.

Social and Physical Disability
The disease may be present for 15 to 20 years, progressing slowly, but still compatible with an active, self-supporting life. After 15 or 20 years the problems of pain, weakness, and general infirmity usually result in increasing invalidism, eventually leading to total dependency.

Pathology
A tubular cavitation develops slowly in the spinal cord, extending over many segments. The most common location is in the lower cervical cord near the central canal. There is loss of anterior horn cells and interruption of spinothalamic fibers. The cavity may be lined by a thick layer of glial tissue. Cavities may be bilateral and asymmetric and may communicate with an enlarged central canal. Ascent of the cavity into the brain stem produces syringobulbia. The canal may extend the entire length of the cord. Associated findings may be ectopic cerebellar tonsils, hydrocephalus, cerebellar hypoplasia, and astrocytoma or ependymoma of the spinal cord.

Essential Features
Pain in the relevant distribution of slowly progressing muscle weakness and wasting and impairment of sensation to pinprick and temperature, while other sensory modalities remain intact.

Differential Diagnosis
Other conditions which have to be considered are: (1) amyotrophic lateral sclerosis, (2) multiple sclerosis, (3) tumor of the spinal cord, (4) skeletal anomalies of the cervical spine, (5) platybasia, and (6) cervical spondylosis.

Code

007.X0	Face
207.X0	Arm
607.X0	Leg

Polymyalgia Rheumatica (I-8)

Definition
Diffuse aching, and usually stiffness, in neck, hip girdle, or shoulder girdle, usually associated with a markedly raised sedimentation rate, sometimes associated with giant cell vasculitis, and promptly responsive to steroids.

System
Musculoskeletal system.

Main Features
Incidence about 54 per 100,000 in those over 30 years of age. Deep muscular aching pain usually begins in the neck, shoulder girdle, and upper arms, but may only involve the pelvis and proximal parts of the thighs.

Morning stiffness and stiffness after inactivity are prominent features.

Associated Symptoms
Malaise, fatigue, depression, low grade fever, weight loss, and giant cell arteritis.

Aggravating Factors
Movement.

Signs
No muscle tenderness or weakness.

Laboratory Findings
Anemia of chronic disease, raised sedimentation rate (usually greater than 50 mm/hour Westergren).

Relief
Dramatic response to oral corticosteroids, usually in low doses, e.g., 5-20 mg prednisone daily.

Complications
Blindness from giant cell arteritis.

Pathology
Giant cell vasculitis.

Essential Features
Diffuse pain with malaise, elevated sedimentation rate, response to steroids.

Diagnostic Criteria
1. Symmetrical proximal limb myalgia and severe stiffness.
2. Symptoms lasting longer than two weeks.
3. Age of onset: 50 years or older.
4. Erythrocyte sedimentation rate (Westergren) 40 mm or higher.
5. Morning stiffness exceeding one hour.

The diagnosis is to be made if three or more of the above criteria are present, or if one of the above criteria and pathologic evidence of giant cell arteritis is present.

Differential Diagnosis
Polymyositis, fibrositis, hyperthyroidism.

Code
X32.X3a

References
Ayoub WT, Franklin CM, Torretti D. Polymyalgia rheumatica: duration of therapy and long-term outcome. Am J Med 1985;37:309.

Bird HA, Esselinckx W, Dixon AS, Mowat AG, Wood PH. An evaluation of criteria for polymyalgia rheumatic. Ann Rheum Dis 1979;38:434.

Fibromyalgia (or Fibrositis) (I-9)

N.B.: We consider Myofascial Pain Syndrome (diffuse or not) to have a somewhat different meaning and think it adds confusion to use the term when discussing fibromyalgia.

Definition
Diffuse musculoskeletal aching and pain with multiple predictable tender points.

Site
Multiple anatomic areas.

System
Musculoskeletal system (muscles, ligaments, tendons, joints).

Main Features
Primary fibromyalgia, without important associated disease, is uncommon compared to concomitant fibromyalgia. It may occur in childhood but is most common in the fourth and fifth decades. The sex ratio is 6:1 female to male. Concomitant fibromyalgia occurs with any other musculoskeletal condition, where it may act to intensify the pain of the associated condition. The syndrome is chronic, and remissions are uncommon. *Pain:* Widespread aching of more than three months' duration, often poorly circumscribed and perceived as deep, usually referred to muscle or bony prominences. Most common areas are cervical, thoracic, and lumbar. Although pain in the trunk and proximal girdle is aching, distal limb pain is often perceived as associated with swelling, numbness, or stiff feeling. Day-to-day fluctuation in pain intensity and shifting from one area to another are characteristic, although the pain is usually continuous. Stiffness is present in 80% and is perceived as an increased resistance to joint movement, particularly toward the end of the range of movement. Both pain and stiffness are maximal within the broad sclerotomic and myotomic areas of reference of the lower segments of the cervical and lumbar spine. *Fatigue* is present in 80%, and is often severe enough to interfere with daily activities. Sleep disturbance is present in 75%, and waking is unrefreshed or tired. *Multiple tender points:* Discrete local areas of deep tenderness widely dispersed throughout the body and involving a variety of otherwise normal tissues are a pathognomonic feature provided about 60% of examined sites are tender. Tender points are found within muscle and over tendons, muscle insertions, and bony prominences. Tender point sites are "tender" in many normal individuals but are reported as "painful," often with grimace or withdrawal when palpated, in those with fibromyalgia. The predictable location of these tender points and their multiplicity are essential features of the syndrome.

Associated Symptoms and Signs
Paresthesias: Most often involving the upper extremities, are found in 60%.

Headaches: Noted in 53%.

Irritable Bowel Syndrome: Noted in 30%. *Anxiety:* Noted in 48%.

Skinfold Tenderness: The rolling of the skin and subcutaneous tissues of the upper scapula region between the examiner's thumb and index finger elicits tenderness in 60%.

Reactive Hyperemia: Redness of the skin developing after palpation of tender points over the trapezius and contiguous regions is found in half the patients.

Autonomic Phenomena: Reactive hyperemia is the most commonly recognized feature, but temperature changes and mild soft tissue swelling involving the distal upper extremities are also frequently reported.

Aggravating and Relieving Features
Cold, poor sleep, anxiety, humidity, weather change, fatigue, and mental stress intensify symptoms in 6070%. Symptoms are typically made worse or brought on by prolonged or vigorous work activity. Warmth (78%) temporarily improves symptoms.

Signs
Tender points, widely and symmetrically distributed, are the characteristic sign of the syndrome. They are not found in other musculoskeletal syndromes.

Relief
Relief may be provided by reassurance and explanation about the nature of the syndrome and possible mechanisms of pain: anxiety may thus be reduced, expensive and hazardous investigations and treatments limited, and use of medication reduced. Low dose amitriptyline, cyclobenzaprine, and aerobic exercise have been shown, in placebo controlled double blind studies, to improve symptoms.

Pathology
Nonspecific muscle changes have been found in some biopsy studies. Blood flow during exercise is reduced, and decreased oxygen uptake in muscles has been noted. Two studies have found increased levels of substance P in the cerebrospinal fluid of patients. In general, these findings, some of which may be secondary phenomena, have been insufficient to explain the major signs and symptoms of the syndrome.

Etiology
Unknown. The syndrome may begin in childhood or early life without obvious association. It also is noted frequently following trauma, and has been known to Page 46
develop after apparent viral illness. Finally, it may appear insidiously in later life. Thus the syndrome may be the final common pathway, perhaps as hyperalgesia, for a number of causative factors. Trauma or degenerative changes in the cervical or lumbar regions might precipitate the syndrome. Intrinsic changes in levels of neurotransmitters might play a factor. A syndrome similar to fibromyalgia can be induced temporarily with experimental reduction in non-REM sleep. Low grade symptoms may be increased by mental stress or fatigue. An association with *previous* major depression in patients and families has suggested a genetic factor.

Classification Criteria for Primary and Concomitant Fibromyalgia (from Wolfe et al. 1990)

1. History of Widespread Pain

Definition
Pain is considered widespread when all of the following are present: pain in the left side of the body, pain in the right side of the body, pain above the waist and below the waist. In addition, axial skeletal pain (cervical spine or anterior chest or thoracic spine or low back) must be present. In this definition, shoulder and buttock pain is considered as pain for each involved side. "Low back" pain is considered lower segment pain.

2. Pain in 11 of 18 Tender Point Sites on Digital Palpation

Definition
Pain, on digital palpation, must be present in at least 11 of the following 18 tender point sites:

Occiput: bilateral, at the suboccipital muscle insertions. *Low Cervical:* bilateral, at the anterior aspects of the intertransverse spaces at C5-C7.

Trapezius: bilateral, at the midpoint of the upper border. *Supraspinatus:* bilateral, at origins above the scapula spine near the medial border.

Second Rib: bilateral, at the second costochondral junctions, just lateral to the junctions on upper surfaces. *Lateral Epicondyle:* bilateral, 2 cm distal to the epicondyles.

Gluteal: bilateral, in upper outer quadrants of buttocks in

anterior fold of muscle. *Greater Trochanter:* bilateral, posterior to the trochanteric prominence.

Knees: bilateral, at the medial fat pad proximal to the joint line. Digital palpation should be performed with an approximate force of 4 kg.

For a tender point to be considered "positive," the subject must state that the palpation was painful. "Tender" is not to be considered painful.

For classification purposes, patients will be said to have fibromyalgia if both criteria are satisfied. Widespread pain must have been present for at least three months. The presence of a second clinical disorder does not exclude the diagnosis of fibromyalgia.

Code
X33.X8a

References
Bennett RM, Goldenberg DL, editors. The fibromyalgia syndrome, Rheumatic Disease Clinics of North America, vol. 15, no. 1. Philadelphia: WB Saunders; 1989.

Wolfe F, Smythe HA, Yunus MB, Bennett RM, Bombardier C, Goldenberg DL, Tugwell P, Campbell SM, Abeles M, Clark P. The American College of Rheumatology 1990 criteria for the classification of fibromyalgia: report of the Multicenter Criteria Committee. Arthritis Rheum 1990;33:160–72.

Note: Specific Myofascial Pain Syndromes

Synonyms: fibrositis (syndrome), myalgia, muscular rheumatism, nonarticular rheumatism.

Specific myofascial syndromes may occur in any voluntary muscle with referred pain, local and referred tenderness, and a tense shortened muscle. The pain has the same qualities as that of the diffuse syndromes. Passive stretch or strong voluntary contraction in the shortened position of the muscle is painful. Satellite tender points may develop within the area of pain reference of the initial trigger point. Other phenomena resemble those of the diffuse syndromes. Diagnosis depends upon the demonstration of a trigger point (tender point) and reproduction of the pain by maneuvers which place stress upon proximal structures or nerve roots. This suggests that the syndrome is an epiphenomenon secondary to proximal pathology such as nerve root irritation. Relief may be obtained by stretch and spray techniques, tender point compression, or tender point injection including the use of "dry" needling.

Some individual syndromes are described here, e.g., sternocleidomastoid and trapezius. Others may be coded as required according to individual muscles that are identified as being a site of trouble.

Rheumatoid Arthritis (I-10)

Definition
Aching, burning joint pain due to systemic inflammatory disease affecting all synovial joints, muscle, ligaments, and tendons in accordance with diagnostic criteria below.

Site
Symmetrical involvement of small and large joints.

System
Musculoskeletal system and connective tissue.

Main Features
Diffuse aching, burning pain in joints, usually moderately severe; usually intermittent with exacerbations and remissions. The condition affects about 1% of the population and is more common in women. Diagnostic criteria of the American Rheumatism Association describe and further define the illness. They are as follows: (1) morning stiffness, (2) pain on motion or tenderness at one joint or more, (3) swelling of one joint, (4) swelling of at least one other joint, and (5) symmetrical joint swelling.

All of the above have to be of at least six weeks' duration. Further criteria include: (6) subcutaneous nodules, (7) typical radiographic changes, (8) positive test for rheumatoid factor in the serum, (9) a poor response in the mucin clot test in the synovial fluid, (10) synovial histopathology consistent with rheumatoid arthritis, and (11) characteristic nodule pathology.

Classical rheumatoid arthritis requires seven criteria to be diagnosed. Definite rheumatoid arthritis may be diagnosed on five criteria, and probable rheumatoid arthritis on three criteria.

Associated Symptoms
Morning stiffness usually greater than half an hour's duration; chronic fatigue. Inflammation may affect eyes, heart, lungs.

Signs
Tenderness, swelling, loss of range of motion of joints, ligaments, tendons. Chronic destruction and joint deformity are common.

Laboratory Findings
Anemia, raised ESR (erythrocyte sedimentation rate), rheumatoid factor in the serum in the majority of

cases.

Relief
Usually good relief of pain and stiffness can be obtained with nonsteroidal anti-inflammatory drugs, but some patients require therapy with gold or other agents.

Pathology
Chronic inflammatory process of synovium, ligaments, or tendons. There may be systemic vasculitis.

Essential Features
Aching, burning joint pain with characteristic pathology.

Diagnostic Criteria
1. Morning stiffness in and around joints lasting at least one hour before maximal improvement.
2. Simultaneous soft tissue swelling or fluid in at least three joint areas observed by a physician. The 14 possible areas are right or left proximal interphalangeal joints (PIP), metacarpal phalangeal (MCP), wrist, elbow, knee, ankle, and metatarsal phalangeal joints (MTP).
3. At least one area of soft tissue swelling or effusion in a wrist, MCP, or PIP joint.
4. Symmetrical arthritis. Simultaneous involvement of the same joint areas as defined in 2 above in both sides of the body (bilateral involvement of PIP, MCP, or MTP is acceptable without absolute symmetry).
5. Rheumatoid nodules.
6. Positive serum rheumatoid factor, demonstrable by any method for which any result has been positive in less than 5% of normal control subjects.
7. Radiographic changes typical of rheumatoid arthritis on posterior-anterior hand and wrist radiographs; this must include erosions or unequivocal bony decalcification which is periarticular.

A patient fulfilling four of these seven criteria can be said to have rheumatoid arthritis. Criteria 1-4 must have been present for at least six weeks.

Differential Diagnosis
Systemic lupus erythematosus, palindromic rheumatism, mixed connective tissue disease, psoriatic arthropathy, calcium pyrophosphate deposition disease, seronegative spondyloarthropathies, hemochromatosis (rarely).

Code
X34.X3a

Reference
Arnett FC, Edworthy SM, Bloch DA, McShane DJ, Fries JF, Cooper NS, Healey LA, Kaplan SR, Liang MH, Luthra HS. The American Rheumatism Association 1987 revised criteria for the classification of rheumatoid arthritis. Arthritis Rheum 1988;31:315–24.

Osteoarthritis (I-11)

Definition
Deep, aching pain due to a “degenerative” process in a single joint or multiple joints, either as a primary phenomenon or secondary to other disease.

Site
Joints most commonly involved are distal and proximal interphalangeal joints of the hands, the carpo-metacarpal thumb joint, the knees, the hips, and cervical and lumbar spines. Many joints or only a few joints may be affected, e.g., at C5 or L5, the hip or knee; proximal joints may be involved alone or only distal interphalangeal joints.

System
Musculoskeletal system.

Main Features
There is deep, aching pain which may be severe as the disease progresses. The pain is felt at the joint or joints involved but may be referred to adjacent muscle groups. Usually the pain increases in proportion to the amount of use of the joint. As the disease progresses there is pain at rest and later nocturnal pain. The pain tends to become more continuous as the severity of the process increases. Stiffness occurs after protracted periods of inactivity and in the morning but lasts less than half an hour as a rule.

There is a discrepancy between radiological prevalence and clinical complaints. Radiological evidence of osteoarthritis occurs in 80% of individuals over 55 years of age. Only about 25% of those with radiographic changes report symptoms. The incidence increases with age. There is a greater prevalence relatively in men under the age of 45 compared with women, and in women over the age of 45 compared with men.

Aggravating Features
Use, fatigue.

Signs
Clinically, joint line tenderness may be found and crepitus on active or passive joint motion; noninflammatory effusions are common. Later stage disease is accompanied by gross deformity, bony-hypertrophy, contracture. X-ray evidence of joint space narrowing, sclerosis, cysts, and osteophytes may occur.

Laboratory Findings
None specific.

Usual Course
Initially there is pain with use and minimal X-ray and clinical findings. Later pain becomes more prolonged as the disease progresses and nocturnal pain occurs. The course is one of gradually progressive pain and deformity.

Relief
Some have relief with nonsteroidal anti-inflammatory agents or with non-narcotic analgesics. Joint rest in the early stages relieves the pain. Occasional relief in the early phases may appear from intra-articular steroids.

Physical Disability
Progressive limitation of ambulation occurs in large weight-bearing joints.

Pathology
This is loosely described as a “degenerative” disease of articular cartilage.

Essential Features
Deep, aching pain associated with the characteristic "degenerative" changes in joints.

Diagnostic Criteria
No official diagnostic criteria exist for osteoarthritis, although criteria have been proposed for osteoarthritis of the knee joint.

Noninflammatory arthritis of one or several diarthrodial joints, occurring in the absence of any known predisposing cause, with loss of cartilage and/or bony sclerosis (or osteophyte formation) demonstrable by X-rays.

Differential Diagnosis
Calcium pyrophosphate deposition disease; presence of congenital traumatic, inflammatory, endocrinological, or metabolic disease to which the osteoarthritis may be secondary.

Code
X38.X6a

Calcium Pyrophosphate Dihydrate Deposition Disease (CPPD) (I-12)

Definition
Attacks of aching, sharp, and throbbing pain with acute or chronic recurrent inflammation of a joint caused by calcium pyrophosphate crystals.

Site
Usually one joint, sometimes more, often alternating. Knees, wrists, and metacarpo-phalangeal joints are most frequent sites.

System
Musculoskeletal system.

Main Features
The disorder occurs clinically in about 1 in 1000 adults, more often in the elderly, but radiology shows the presence of the disease in 5% of adults at the time of death. There are four major clinical presentations: *(1) pseudogout:* acute redness, heat, swelling, and severe pain which is aching, sharp, or throbbing in one or a few joints; the attacks last from 2 days to several weeks, with freedom from pain between attacks; *(2) pseudorheumatoid arthritis:* marked by deep aching and swelling in multiple joints, with attacks lasting weeks to months; (3) *pseudo-osteoarthritis:* see the description of osteoarthritic features; and *(4) pseudarthritis with acute attacks:* the pain being the same as in osteoarthritis but with superimposed acute painful swollen joints.

Signs
Aspiration of calcium pyrophosphate crystals from the joint is diagnostic. X-rays show calcification in the cartilage of the wrists, knees, and symphysis pubis.

Relief
Acute attacks respond well to nonsteroidal antiinflammatory drugs, with or without local corticosteroid injections.

Complications
Chronic disabling arthritis.

Associated Disorders
Hyperparathyroidism, hemochromatosis. There may be hereditary, sporadic, or metabolic causes.

Pathology
Acute and chronic inflammation or degeneration.

Diagnostic Criteria
1. Demonstration of CPPD crystals in tissues or synovial fluid by definitive means such as X-ray diffraction.
2. Crystals compatible with CPPD demonstrable by compensated polarized light microscopy.
3. Typical calcifications seen on roentgenograms.

A definite diagnosis can be made if 1 above is present, or if 2 and 3 are present. A probable diagnosis can be made if 2 or 3 is present.

Differential Diagnosis
Gout, infection, palindromic rheumatism, osteoarthritis.

Code
X38.X0 or X38.X5a

Reference
Ryan LM, McCarty DJ. Calcium pyrophosphate crystal deposition disease: pseudogout articular chondrocalcinosis. In: McCarty DJ, editor. Arthritis and Allied Conditions, 10th ed. Philadelphia: Lea & Febiger; 1985. p. 1515–46.

Gout (I-13)

Definition
Paroxysmal attacks of aching, sharp, or throbbing pain, usually severe and due to inflammation of a joint caused by monosodium urate crystals.

Site
First metatarso-phalangeal joints, midtarsal joints, ankles, knees, wrists, fingers, or elbows.

Main Features
More common in men in the fourth to sixth decades of life and in postmenopausal women. Acute severe paroxysmal attacks of pain occur with redness, heat, swelling, and tenderness, usually in one joint. The pain is aching, sharp, and throbbing. The patient is often unable to accept the weight of bedclothes on the joint and unable to bear weight on the affected joint. Attacks last two days to several weeks in duration.

Associated Symptoms
In the acute phase, patients may be febrile and have leukocytosis.

Aggravating Factors
Trauma, alcohol ingestion, surgery, starvation.

Signs
Redness, heat, and tender swelling of the joint, which may be extremely painful to move. Intracellular urate crystals aspirated from the joint are diagnostic.

Laboratory Findings
Serum urate may vary during the acute attack. Leukocytosis and raised sedimentation rate are seen during the attack.

Usual Course
Initially the disorder is monoarticular; in 50% of patients the first metatarso-phalangeal joint is involved in the great toe. Acute attacks are separated by variable symptom free intervals. Attacks may become polyarticular and recur at shorter intervals and may eventually resolve incompletely leaving chronic, progressive crippling arthritis.

Relief
Responds well to nonsteroidal anti-inflammatory agents, intravenous colchine, and local steroid injections.

Complications
Renal calculi, tophaceous deposits, and chronic arthritis with joint damage.

Pathology
Acute inflammatory response induced by uric acid crystals.

Essential Features
Paroxysmal joint pains with sodium monourate deposition.

Diagnostic Criteria
1. Demonstration of intracellular sodium urate monohydrate crystals in synovial fluid leukocytes by polarizing microscopy or other acceptable methods of identifying crystals.
2. Demonstration of sodium urate monohydrate crystals in an aspirate or biopsy of a tophus by methods similar to those in 1.
3. In the absence of specific crystal identification, a history of monoarticular arthritis followed by an asymptomatic intercritical period, rapid resolution of synovitis following Colchicine administration, and the presence of hyperuricemia.

Any one of the three above is sufficient to make the diagnosis.

Differential Diagnosis
Calcium pyrophosphate deposition disease, infection, palindromic rheumatism.

Code
X38.X5b

Reference
Holmes EW. Clinical gout and the pathogenesis of hyperuricemia. In: McCarty DJ, editor. Arthritis and Allied Conditions, 10th ed. Philadelphia: Lea & Febiger; 1985. p. 1445–80.

Hemophilic Arthropathy (I-14)

Definition
Bouts of acute, constant, nagging, burning, bursting, and incapacitating pain or chronic, aching, nagging, gnawing, and grating pain occurring in patients with congenital blood coagulation factor deficiencies and secondary to hemarthrosis.

Site
The most common joints affected initially are the knees, ankles, and elbows. Shoulders, hips, and wrist joints are affected next most often. As the first joints become progressively affected, other remaining articular and muscle areas are involved with changes of disuse atrophy or progressive hemorrhagic episodes.

System
Musculoskeletal system.

Main Features
Prevalence: hemophilic joint hemorrhages occur in severely and moderately affected male hemophiliacs. They only rarely occur in female Factor VIII and Factor IX carriers and in homozygous severely affected patients with von Willebrand's disease. Acute hemarthrosis occurs most commonly in the juvenile in association with minor trauma. In the adult, spontaneous hemorrhages and pain occur in association also with minor or severe trauma. Characteristically the acute pain is associated with such hemarthrosis, which is relieved by replacement therapy and rest of the affected limb. A reactive synovitis results from repeated hemarthroses, which may be simply spontaneous small recurrent hemorrhages. The pain associated with them is extremely difficult to treat because of the underlying inflammatory reaction. *Time Course:* The acute pain is marked by fullness and stiffness and constant nagging, burning, or bursting qualities. It is incapacitating and will cause severe pain for at least a week depending upon the degree of intra-capsular swelling and pressure. It will recur episodically from the causes indicated. Chronic pain is often a dull ache, worse with movement, but can be debilitating, gnawing, and grating. At the stage of destructive joint changes the chronic pain is unremitting and relieved mainly by rest and analgesics. These syndromes are exacerbated by accompanying joint and muscle degeneration due to lack of mobility rather than repeated hemorrhages.

Associated Symptoms
Depressive or passive/aggressive symptoms often accompany hemorrhages and are secondary to the extent of pain or to the realization of vulnerability to hemorrhage, which is beyond the control of the hemophiliac. If bleeding occurs into a muscle or potential space, e.g., retroperitoneal and iliopsoas muscle, this can mimic joint hemorrhage and can cause severe nerve compression syndromes, e.g., of the femoral nerve. Numerous psychosomatic complaints are associated with the chronic and acute pain of chronic synovitis, arthritis, and hemarthrosis.

Signs
Reactive Synovitis: There is a chronic swelling of the joint with a "boggy" consistency to the swelling, which is tender to palpation. Marked limitation of joint movement often with signs of adjacent involvement of muscle groups due to disuse atrophy. *Chronic Joint Degeneration:* Severe bony remodeling with decrease in joint movement, adjacent muscular atrophy with subsequent fixation of the joint and loss of effective use.

Laboratory Findings
X-rays with the large hemarthrosis show little except for soft tissue swelling. In reactive synovitis there is often evidence of osteoporosis accompanied by overgrowth of the epiphyses but not evidence of joint

destruction. In chronic arthropathy there is cartilage destruction and narrowing of the joint space. Gross misalignment of the joint surfaces progresses. Cysts, rarefactions, subcondylar cysts, and an overgrowth of the epiphysis are noted. This progresses through to fibrous joint contracture, loss of joint space, extensive enlargement of the epiphysis, and substantial disorganization of the joint structures. The articular cartilage shows extensive degeneration with fibrillation and eburnated bone ends.

Usual Course
Until the availability of therapy with blood clotting factor concentrate, there was an inexorable deterioration of the affected joints following the initial repeated spontaneous hemarthroses in the severely affected individual. This joint deterioration was associated with pain as described in the section regarding time course. The introduction of concentrated clotting factor transfusions has avoided the consequence of repeated acute severe hemarthroses. However, it is by no means certain whether the pain pattern of chronic synovitis and arthritis can be avoided or merely delayed using such therapy. Therapy blood clotting factor concentrate is available on a regular basis only in North America and Europe at this time.

Relief
Acute Hemarthrosis: Adequate intravenous replacement with appropriate coagulation factors with subsequent graded exercise and physiotherapy will provide good relief. Aspiration of the joint will be necessary under coagulation factor cover if there is excessive intracapsular pressure. Analgesics are required for acute pain management. *Reactive and Chronic Hemarthrosis:* Prophylactic factor replacement is required in association with analgesics and carefully selected antiinflammatory agents, e.g., steroids or ibuprofen. Pain control using analgesics and transcutaneous nerve stimulation is also useful, and physiotherapy is of considerable assistance in managing both symptoms and signs. Synovectomy may be of use for the control of pain secondary to the recurrent bleeding. *Chronic Destructive Arthropathy:* Replacement therapy is of little assistance in relieving pain and disability. Carefully selected antiinflammatory agents and rest are the major therapies of use. Physiotherapy after control of acute symptoms is useful. Joint replacement is a final choice for chronic pain management.

Complications
Analgesic abuse is a common problem in hemophilia due to the acute and chronic pain syndromes associated with hemophilic arthropathy. This problem can be avoided in the younger age group by not using narcotic analgesics for chronic pain management and relying upon principles of comprehensive hemophilia care. These include regular physiotherapy, exercise, and making full use of available social and professional opportunities.

Social and Physical Disability
Severe crippling and physical disability, with prolonged school and work absences, have traditionally been associated with this form of arthropathy. Consequently, affected individuals have not been able to achieve satisfactory school and job schedules. It is considered that the higher suicide rate is related not only to the family and psychosocial aspects of the disease but also to the chronic pain syndromes that these individuals experience.

Pathology
This depends upon the phase of the disorder. Generally two pathologic phases are associated with the hemophilic joint. Phase one involves an early synovial soft tissue reaction caused by intraarticular bleeding. Synovial hypertrophy with hemosiderin deposition and mild perivascular inflammation are present. Cartilage degeneration and joint degeneration similar to that seen in osteoarthritis and rheumatoid arthritis is seen in the second-phase joint. Associated with this type of phase two change is synovial thickening and hyperplasia which falls into numerous folds and clusters of villi. The amount of hemosiderin deposited is increased compared to phase one.

Summary of Essential Features and Diagnostic Criteria
Acute and chronic pain as the result of acute hemarthrosis with chronic synovial cartilaginous and bony degeneration is exacerbated by spontaneous and trauma-related hemorrhage.

Diagnostic Criteria
Pain associated with hemophiliac arthropathy must satisfy both 1 and 2.

1. Spontaneous intracapsular hemorrhages in an individual with an inherited hemostatic defect.
2. Demonstrable synovial bleeding with or without bony joint contour abnormalities.

Differential Diagnosis
In the presence of a severe (less than 0.01 units/ml) hemophilic factor deficiency, no other diagnosis is possible. In the mildly affected individual (greater than 0.05 units/ml), all other causes of degenerative arthritis, particularly in the older affected individual, must be considered.

Code
X34.X0a

References
Arnold WD, Hilgartner MW. Haemophilia arthropathy: current concepts of pathogenesis and management. J Bone Joint Surg 1977;59A:287–305.

Duthie RB, Matthews JM, Rizza CR, Steele WM. The Management of Musculoskeletal Problems in the Hemophiliac, 1st ed. Oxford: Blackwell; 1972.

Hilgartner MW. Hemophiliac arthropathy. Adv Paediatr 1975;21:139–65.

Hoskinson J, Duthie RB. Management of musculoskeletal problems in the hemophilias. Orthop Clin North Am 1978;9:455–80.

Burns (I-15)

Definition
Acute and severe pain at first, following bums, later continuous with exacerbations, gradually declining.

Site
Anywhere on the body surface and deep to it.

System
Usually only epidermis and/or dermis, but any system may be involved.

Main Features
Prevalence: is approximately 3 per 1000 of population. Ten percent of these will require hospital admission. Any age can be affected, but the highest incidence (18%) is between 20 and 29 years. Children are the next largest group, with 30% of these being in the 1-2 year age group. *Sex Ratio:* approximately 1:1, but 3:2 males to females in children.

Pain Quality: initially the pain is acute and intense. It is frequently described as throbbing, smarting, and stinging, and marked exacerbations of stabbing pain occur with any movement or procedure. Thus, it is particularly intense where there are skin creases or flexures or where pressure is applied, such as palms, soles, genitalia, ears, or resting surfaces. This applies especially to partial thickness bums. Despite the destruction of all cutaneous nerve endings, full thickness bums are often painful with a quality described as deep, dull, or aching.

Intensity and Duration: the pain tends to diminish in intensity as healing takes place. In addition, the quality of the pain changes, and at one to two weeks after the bum is usually described as sore, aching, tender, tiring, and tight. After three or four weeks it is described as itchy or tingling. These descriptions also apply to pain at donor sites. Pain is exacerbated by procedures such as "tanking" for the removal of eschar, and physiotherapy. In addition, frequent surgery is often necessary, with an accompanying increase in pain. Relief may be promoted by the use of opioid premedication prior to procedures, time-contingent analgesics, inhalational analgesia during procedures, ensuring that the burnt areas never dry out, protecting the bum with creams, and achieving skin cover by some means as soon as possible.

Associated Symptoms
Dyspnea may occur as a result of smoke inhalation. Disuse may lead to causalgia-like symptoms.

Usual Course
Tends to settle with skin healing. Burnt areas may be tender and sore for up to a year. Itch and irritation may continue for two or three weeks.

Complications
If healing occurs, it is unusual to have persistent pain unless deep structures (muscle, bones, major nerves) are involved. Cellulitis in burnt areas or donor sites may lead to a marked increase in the severity of pain.

Social and Physical Disability
This is most frequent where the bum is extensive, and such cases often require sustained treatment and prolonged hospitalization. Psychological treatment is also needed where scars affect the patient's ability to function socially or physically, for example, as a result of scars of the hands, face, or genitalia.

Pathology
Loss of skin integrity with consequent loss of fluid and thermoregulation and an increased likelihood of infection. Burns are classified in three degrees of severity based on burn depth. A superficial burn involves the epidermis only. A partial thickness burn involves epidermis and dermis at varying depths, and a full thickness burn involves epidermis, dermis, and at times deeper tissues. Electrical burns may cause considerable damage to deeper tissues by direct effect and by occlusion of blood vessels. The severity of damage is related to the temperature to which the area was exposed, the duration of exposure, and the thickness of the skin involved. The agents responsible may be thermal, electrical, or chemical.

Summary of Essential Features and Diagnostic Criteria
Pain with the appropriate time course following burns.

Differential Diagnosis
Possibly hysterical conversion pain or pain of psychological origin may prolong or exacerbate the original effects of the injury. This may be more important in work-related injuries or where there is litigation.

Code
X42.Xl or X82.X1

Pain of Psychological Origin: Muscle Tension Pain (I-16.1)

Definition
Virtually continuous pain in any part of the body due to sustained muscle contraction and provoked by emotional causes or by persistent overuse of particular muscles.

Site
Any region with pain reference from voluntary muscle.

System
Central nervous system (psychological and social).

Main Features
Prevalence: often diagnosed. Even approximate prevalence is unknown. *Sex Ratio:* females more than males, 4:1 in those who consult doctors. *Age of Onset:* from age 8 onward, usually before age 30. *Start:* gradual emergence intermittent at first, as mild diffuse ache or unpleasant feeling, increasing to a definite pain part of the time. Fluctuation during the day is typical. These exacerbations seem to emerge after several years of lesser headache. *Pain Quality:* dull ache, usually does not throb; severe during exacerbations, often or almost always with throbbing. Some describe tight bands or gripping headache. They may be a minority. Others describe pressure sensations. *Occurrence and Duration:* most days per week, usually every day for most of the day. Occasionally in long-standing severe cases pain may wake the patient from sleep. *Precipitants and Exacerbating Factors:* emotional stress, anxiety and depression, physical exercise, alcohol.

Associated Symptoms
Many patients have anxiety, depression, irritability, or more than one of these combined.

Signs
Muscle tenderness occurs but may also be found in other conditions and in normal individuals.

Relief
Resolution or treatment of emotional problems, anxiety, or depression often diminishes symptoms. Relaxation treatment helps. Anxiolytics may help but should be avoided since some patients become depressed and others develop dependence. Tricyclic antidepressants are frequently very useful. Analgesics help only a little.

Complications
Analgesics, narcotic, and other drug abuse.

Social and Physical Disability
Reduction of activities and of work.

Pathology
Unsettled.

Differential Diagnosis
From delusional and conversion pains; from muscle spasm provoked by local disease; and from other causes of dysfunction in particular regions, e.g., migraine, posttraumatic headache, cervical spine disorders, depression, hallucinatory headache, and conversion hysteria.

Code
X33.X7b

Note: “b” coding used to allow the “a” coding to be employed if an acute syndrome needs to be specified.

Pain of Psychological Origin: Delusional or Hallucinatory (I-16.2)

Definition
Pain of psychological origin and attributed by the patient to a specific delusional cause.

Site
Any part of the body. May be symmetrical, e.g., in a fronto-temporal-occipital ring distribution, or in one place, e.g., at vertex, precordial, genital.

Main Features
Prevalence: rare; estimated to be present in less than 2% of patients with chronic pain without lesions. *Age of Onset:* not apparently reported in children; onset in late adolescence or at any time in adult life. *Pain Quality:* may be sensory or affective or both, not necessarily bizarre; essential characteristic is attribution of the pain by the patient to a specific delusional cause, e.g., to a crown of thorns in a patient who had messianic delusions. *Time Pattern:* in accordance with the delusion. *Intensity:* from mild to severe. *Usual Duration:* in accordance with the causal psychological illness.

Associated Symptoms and Modifying Factors
May be exacerbated by psychological stress, relieved by treatment causing remission of illness. No physical signs or laboratory findings.

Complications
In accordance with causal condition; usually lasts for a few weeks in manic-depressive or schizo-affective psychoses, may be sustained for months or years in established schizophrenia if resistant to treatment. Occasionally chronic pain without any formal delusions remits to be succeeded by a paranoid or schizophrenic psychosis.

Social and Physical Disabilities
In accordance with the mental state and its consequences. Drug addiction not reported.

Etiology
Manic-depressive, schizophrenic, or possibly other psychoses.

Essential Features
Those required for diagnosis are pain, without a lesion or overt physical mechanism and founded upon a delusional or hallucinatory state.

Differential Diagnosis
From undisclosed or missed lesions in psychotic patients, or migraine, giving rise to delusional misinterpretations; from tension headaches; from hysterical, hypochondriacal, or conversion states.

Code
X 1 X.X9a

Note: X = to be completed individually according to circumstances in each case.

Pain of Psychological Origin: Hysterical, Conversion, or Hypochondriacal (1-16.3)

Definition
Pain specifically attributable to the thought process, emotional state, or personality of the patient in the absence of an organic or delusional cause or tension mechanism.

Site
May be symmetrical; if lateralized, possibly more often on the left precordium, genitals; may be at any single point over the cranium or face, can involve tongue or oral cavity or any other body region. Usually diffuse in fronto-temporo-occipital region or in maxillary area.

Main Features
Prevalence: true population prevalence unknown. Frequency increases from general practice populations to specialized headache or pain clinics or psychiatric departments. Estimates of 11% and 43% have been found in psychiatric departments, depending on the sample. *Sex Ratio:* estimated female to male ratio 2:1 or greater-particularly if multiple complaints occur. *Onset:* may be at any time from childhood onward but most often in late adolescence. *Pain Quality:* described mostly in simple sensory terms, but complex or affective descriptions occur in some cases. *Time Pattern:* Pain is usually continuous throughout most of the waking hours but fluctuates somewhat in intensity, does not wake the patient from sleep. *Duration:* usually lasts for more than six months.

Associated Symptoms
Loss of function without a physical basis (anesthesia, paralyses, etc.) may be present. Pain is often present in other areas. There may be frequent visits to physicians to obtain relief despite medical reassurance, or excessive use of analgesics as well as other psychotropic drugs for complaints of depression, neither type of remedy proving effective. The pain may have a symbolic significance, e.g., identifying the patient with someone who died of brain tumor. Psychological interpretations are frequently not acceptable to the patient, although emotional conflict may have provoked the condition. These patients tend to marry but have poor marital relationships.

La belle indifference can occur but is not common. Depressive complaints and resentment are more frequent. The personality is often of a dependent-histrionic-labile type ("hysterical personality" or "passive dependent personality"). A history of past conversion symptoms is helpful in diagnosis.

There are three overlapping types in this category. The first is largely *monosymptomatic,* is relatively rare, and consists of patients who have pain in one or two regions only, who have only recently developed pain, and who have clear evidence of emotional conflicts, perhaps with an associated paralysis or anesthesia, and a relatively good prognosis. Some patients who primarily have a depressive illness also present with pain as the main somatic symptom. Their pain may be interpreted delusionally or may be based on a tension pain, etc., or may be hysterical.

The second type is of patients with more numerous or *multiple complaints,* often of many and varied types without a physical basis. In the history these often number more than 10, including classical conversion or pseudoneurological symptoms (paralyses, weakness, impairment of special senses, difficulty in swallowing, etc.), gastrointestinal, cardiovascular (palpitations, shortness of breath), disturbances in sexual function (impaired libido, reduced potency), etc., as well as pains in different parts.

In the third, or *hypochondriacal,* subtype, the patient presents excessive concern or fear of the symptoms and a conviction that disease is present despite thorough physical examination, appropriate investigation,

and careful reassurance. There may also be other signs of preoccupation with somatic health, e.g., great anxiety over constipation, the color of the urine, etc.

As emphasized, the subtypes overlap. The most common pattern in pain clinics is the second one described. A hypochondriacal pattern may be observed either alone or with the first or the second subtype, more often with the second. In all types, physical treatments (manipulation, physiotherapy, surgery) tend to produce brief improvements which are not maintained. In the second and third types, a disorder of emotional development is often present.

Note: Depressive pain has been distributed among the above three types and also into the delusional and tension pain groups. This is done because there does not seem to be a single mechanism for pain associated with depression, even though such pain is frequent. The words "depressive pain" as indicating a particular type or mechanism should be avoided.

Aggravating Factors
Emotional stress may be a predisposing factor and is almost always important in the monosymptomatic type. Experience of physical illness or pain due to emotional stress in person or in a family member or close associate may be a predisposing factor.

Usual Course
Usually chronic in the first subtype. In relatively acute monosymptomatic conditions, environmental change and sometimes individual psychotherapy may promote recovery.

Complications
Dependence on minor tranquilizers; salicylate addiction; narcotic addiction; drug-induced confusional states; excessive investigations; unsuccessful surgery, sometimes repeatedly.

Social and Physical Disability
Often associated with marital disharmony, inability to sustain regular employment, sometimes loss of function or limbs due to surgery.

Essential Features
Pain without adequate organic or pathophysiological explanation. Separate evidence other than the prime complaint to support the view that psychiatric illness is present. Proof of the presence of psychological factors in addition by virtue of both of the following: (1) an appropriate and important relationship in time exists between the onset or exacerbation of the pain and an emotional conflict or need, and (2) the pain enables the individual to avoid some activity that is unwelcome to him or her or to obtain support from the environment that otherwise might not be forthcoming.

The condition must not be attributable to any psychiatric disorder other than the following, and it should conform to the requirements for the diagnoses of Dissociative [conversion] Disorders (F44) or Somatoform Disorder (F45) in the International Classification of Diseases, 10th edition, or to those for somatization disorder (300.81) or conversion disorder (300.11), somatoform pain disorder (307.80), or hypochondriasis (300.70) in the American Psychiatric Association Diagnostic and Statistical Manual, 3rd edition revised (DSM-III-R).

Differential Diagnosis
(1) From physical causes of pain, e.g., tumor, acromegaly, Paget's disease of bone, etc.; (2) from physical illnesses that may present with multiple, often diffuse symptoms, e.g., hypothyroidism, hyperparathyroidism, disseminated lupus erythematosis, multiple sclerosis, porphyria; (3) from schizophrenia, endogenous depression, reactive depression, or major depressive disorder according to

DSM-III-R, from pain of psychological origin associated with depression; and (4) from tension pain, particularly headache. The differential diagnosis from tension headache usually will be based on one or more of the following: (a) the level of observed anxiety is not sufficient to account for tension which might produce the symptom; (b) the personality conforms to the hysterical or hypochondriacal pattern and the complaint to an acute conflict situation or to a pattern of multiple symptoms; and (c) relaxation exercises and sedation do not provide relief.

Code
X1X.X9b

References
Diagnostic and Statistical Manual, 3rd ed., Revised. Washington, D.C.: American Psychiatric Association; 1987.

International Classification of Diseases, 10th ed. Geneva: World Health Organization; 1992.

Pain of Psychological Origin: Associated with Depression (1-16.4)

Definition
Pain occurring in the course of a depressive illness, usually not preceding the depression and not attributable to any other cause.

Site
Any part of the body; may be symmetrical, e.g., in a fronto-temporal occipital ring distribution, or in one place, e.g., at vertex, precordial, low back, genital.

Main Features
Prevalence: probably common. Likely to appear in the majority of patients with an independent depressive illness, more often in nonendogenous depression, and less often in illness with an endogenous pattern. *Sex Ratio:* more common in females. *Pain Quality:* may be sensory or affective, or both, not necessarily bizarre; worse with intercurrent stress, increased anxiety. The pain may occur at the site of previous trauma (accidental or surgical) and may therefore be confused with a recurrence of the original condition. Usually aching or throbbing, may be described as sharp. May have both sensory and affective components. *Intensity:* varies from mild to severe. *Duration* and intensity often in accordance with the length and severity of the depression.

Associated Symptoms
Anxiety and irritability are common.

Signs
Tenderness may occur, but may also be found in other conditions and in normal individuals.

Relief
Improvement in the pain occurs with the improvement of the depression. The response to psychological treatments or antidepressants is better than to analgesics.

Social and Physical Disability
Reduction of activities and work.

Etiology
A link with reductions in cerebral monoamines or monoamine receptors has been suggested.

Differential Diagnosis
Muscle tension pain with depression, delusional, or hallucinatory pain; in depression or with schizophrenia, muscle spasm provoked by local disease; and other causes of dysfunction in particular regions, e.g., migraine, posttraumatic headache, cervical spine disorders, hysterical or hypochondriacal pain.

It is important not to confuse the situation of depression causing pain as a secondary phenomenon with depression which commonly occurs when chronic pain arising for physical reasons is troublesome.

Code
X1X.X9d

Note: Unlike muscle contraction pain, hysterical pain, or delusional pain, no clear mechanism is recognized for this category. If the patient has a depressive illness with delusions, the pain should be classified under Pain of Psychological Origin: Delusional or Hallucinatory. If muscle contraction predominates and can be demonstrated as a cause for the pain, that diagnosis may be preferred. Patients with anxiety and depression who do not have evident muscle contraction may have pain in this category. Previously, depressive pain was distributed between other types of pain of psychological origin, including delusional and tension pain groups and hysterical and hypochondriacal pains. The reason for this was the lack of a definite mechanism with good supporting evidence for a separate category of depressive pain. While the evidence that there is a specific mechanism is still poor, the occurrence of pain in consequence of depression is common, and was not adequately covered by the alternative categories mentioned.

Reference
Magni G. On the relationship between chronic pain and depression when there is no organic lesion. Pain 1987;31:1–21.

A Note on Factitious Illness and Malingering (1-17)

Factitious illness is of concern to psychiatrists because both it and malingering are frequently associated with personality disorder. Physicians in any discipline may encounter the problem in differential diagnosis. No coding is given for pain in these circumstances because it will be either induced by physical change or counterfeit. In the first instance it can be coded under the appropriate physical heading. In the second case, the complaint of pain does not represent the presence of pain. ICD-10 does not appear to provide a code for malingering, which suggests that the final application of the label of malingering is a judicial (legal) process and not a medical one. The role of the doctor in this task may be limited to drawing attention to discrepancies and inconsistencies in the history and clinical findings.

Regional Sprains or Strains (1-18)

Code
X33.X1d

Sickle Cell Arthropathy (1-19)

Code
X34.X0c

Purpuric Arthropathy (1-20)

Code
X34.X0d

Stiff Man Syndrome (1-21)

Code
934.X8

Paralysis Agitans (1-22)

Code
902.X7

Epilepsy (1-23)

Code
X04.X7

Polyarteritis Nodosa (1-24)

Code
X5X.X3

Psoriatic Arthropathy and Other Secondary Arthropathies (1-25)

Code
X34.X8c

Painful Scar (1-26)

Code
X4X.X l b

Systemic Lupus Erythematosis, Systemic Sclerosis and Fibrosclerosis, Polymyositis, and Dermatomyositis (1-27)

Code
X33.X3b

Infective Arthropathies (1-28)

Code
X33.X3c

Traumatic Arthropathy (1-29)

Code
X33.Xla

Osteomyelitis (1-30)

Code
X32.X2f

Osteitis Deformans (1-31)

Code
X32.X5b

Osteochondritis (1-32)

Code
X32.X5c

Osteoporosis (1-33)

Code
X32.X5d

Muscle Spasm (1-34)

Code
X37.X7

Local Pain, No Cause Specified (1-35)

Code
X7X.XXa or X3X.X8e

Guillain-Barré Syndrome (1-36)

Definition
Pain arising from an acute demyelinating neuropathy.

Site
Back, extremities, abdomen.

System
Peripheral nervous system, musculoskeletal system.

Main Features
Deep aching pain involving the low back region, buttocks, thighs, and calves is common (> 50%) in the first week or two of the illness. Pain may also occur in the shoulder girdle and upper extremity but is less frequent. Beyond the first month, burning tingling extremity pain occurs in about 25% of patients. **Note:** While in the Guillain-Barré syndrome weakness typically occurs first in the feet and the legs and then later in the arms, the worst pain is in the low back, buttocks, thighs, and calves.

Associated Symptoms
During the acute phase there may be muscle pain and pains of cramps in the extremities associated with muscle tenderness. Constipation can produce lower abdominal and pelvic pain.

Signs
Extremity weakness and areflexia are essential features of the neuropathy. Back and leg pain are commonly exacerbated by nerve root traction maneuvers such as straight-leg raising.

Laboratory Findings
EMG evidence of demyelination (conduction block) and secondary axonal degeneration. Cerebrospinal fluid shows elevated protein with relatively normal cell count.

Usual Course
Aching back and extremity pain, sometimes of a severe nature, usually resolves over the first four weeks. Dysesthetic extremity pain persists indefinitely in 5-10% of patients.

Relief
Acetaminophen or nonsteroidal anti-inflammatory drugs for mild to moderate pain. Opioid analgesics for severe pain-continuous parenteral infusion or epidural administration may be required. Active and passive exercise program. Bowel stimulants to prevent constipation. Padding to prevent pressure palsies.

Complications
Persistent weakness and contractures from incomplete recovery. Ulnar and peroneal pressure palsies from immobilization.

Pathology
Peripheral nerve demyelination with secondary axonal degeneration.

Differential Diagnosis
Pain secondary to neuropathies stimulating Guillain Barré syndrome: porphyria, diphtheritic infection, toxic neuropathies (e.g., lead, solvent abuse such as glue sniffing).

Code
901.X3