

E. LOCAL SYNDROMES OF THE UPPER LIMBS AND RELATIVELY GENERALIZED SYNDROMES OF THE UPPER AND LOWER LIMBS

GROUP XI: PAIN IN THE SHOULDER, ARM, AND HAND

Tumors of the Brachial Plexus (XI-1)

Definition

Progressive aching, burning pain with paresthesias and sensory and motor impairment in the distribution of a branch or branches of the brachial plexus due to tumor.

Site

Shoulder and upper limb.

System

Nervous system.

Tumors

Benign tumors: schwannoma, neurofibroma. Malignant tumors: malignant schwannoma and fibrosarcoma, metastatic neoplasm or direct invasion from other lesion, neuroblastoma, ganglioneuroma (secondary neoplasia of peripheral nerves occurs frequently in lymphoma, leukemia, multiple myeloma). Breast, lung and thyroid neoplasia frequently involve the brachial plexus.

Main Features

Incidence: the specific tumors of peripheral nerve are extremely rare. *Sex Ratio:* there is no sex predilection. *Age of Onset:* young adulthood. They are more common with von Recklinghausen's disease. *Pain Quality:* the pain tends to be constant, gradual in onset, aching, and burning, and associated with paresthesias in the distribution of the pain, progressive wasting of muscles depending upon what groups are involved, and sensory loss. *Intensity:* severe.

Associated Symptoms

The pain is generally not affected by activity. There is associated sensory loss and muscle wasting depending upon the area of the brachial plexus involved. Pain relief is often not adequate, even with significant narcotics.

Signs and Laboratory Findings

The laboratory findings are those of the underlying disease. Signs are loss of reflexes, sensation, and muscle strength in the distribution of the involved portion of the plexus. There may be a local mass. The diagnosis is usually made promptly by X-ray or by CT scan. Electromyographic studies validate the location of the lesion, and there may be a palpable mass in the supraclavicular space.

Usual Course

These tumors are relentlessly progressive unless treated.

Social and Physical Disability

Those related to the loss of function.

Summary of Essential Features

The tumors are associated with slowly progressive pain and paresthesias, and subsequently severe sensory

loss and motor loss. The diagnostic criteria are the increasing aching, burning pain, its distribution in the brachial plexus, the associated paresthesias, motor and sensory loss, and the presence of a mass by palpation and on X-ray or CT scan.

Diagnostic Criteria

1. Burning pain of increasing severity referred to the upper extremity. Occasional superimposed lancinating pain in the same distribution.
2. Subsequent progressive motor and sensory loss and autonomic dysfunction.
3. CT scan may show tumor involvement of the brachial plexus.

Differential Diagnosis

Includes all those lesions above, the scalenus anticus syndrome, and abnormalities of the first thoracic rib or the presence of a cervical rib.

Code

102.X4b

202.X4b

Chemical Irritation of the Brachial Plexus (XI-2)

Definition

Continuous burning pain occasionally accompanied by severe paroxysms, in the distribution of the brachial plexus or one of its branches, with sensory-motion deficits due to effects of local injection of chemical irritants.

Site

Upper limb.

System

Nervous system.

Main Features

Prevalence: injections in the shoulder area with any noxious agent are extremely rare. However, the pain syndromes from these injections are quite well defined. *Incidence:* the pain begins almost immediately with the injection and is continuous. *Pain Quality:* it is usually burning in character, superficial, and unaffected by activity. Occasional paroxysms may occur. It frequently persists even after neurological loss has resolved and is not necessarily associated with paresthesias or sensory loss. There are no differences between noxious agents as to time pattern, occurrence, character, intensity, or duration.

Signs and Laboratory Findings

The signs are of brachial plexus injury. Atrophy, sensory loss, and paresthesias occur in the appropriate area depending upon the portion of the plexus injured. There are no specific laboratory findings.

Usual Course

Pain is generally acute with the injection and gradually improves. Most disappear within a few weeks. Those that persist continue unabated permanently.

Complications

The complications are those of brachial plexus injury.

Pathology

The pathology is a combination of intraneural and extra-neural scarring with focal demyelination.

Summary of Essential Features

These are those of brachial plexus injury. The diagnosis can only be made by history of injection.

Diagnostic Criteria

1. History of injection of chemical irritant.
2. Burning pain with occasional superimposed paroxysms referred to the upper extremity.
3. Pain syndrome stabilizes or actually improves after days to weeks.

Differential Diagnosis

This includes all of the muscular and bony compressions, anomalies, and tumors previously described.

Code

102.X5

202.X5

Traumatic Avulsion of the Brachial Plexus (XI-3)**Definition**

Pain, most often burning or crushing with super-added paroxysms, following avulsion lesions of the brachial plexus.

Site

Felt almost invariably in the forearm and hand irrespective of the roots avulsed. Occasionally, in avulsion of C5 root only, pain may be felt in shoulder.

System

Nerve roots torn from the spinal cord.

Main Features

Prevalence: some 90% of the patients with avulsion of one or more nerve roots suffer pain at some time. Virtually all patients with avulsion of all five roots suffer severe pain for some months at least. *Age of Onset:* vast majority of patients with this lesion are young men between the ages of 18 and 25 suffering from motorcycle accidents. The older the patient the more likely he is to suffer pain from the avulsion lesions. *Pain Quality:* the pain is characteristically described as burning or crushing, as if the hand were being crushed in a vise or were on fire. The pain is constant and is a permanent background to the patient's life, and in a high proportion of patients (in one series, 90%), there were periodic paroxysms or shots of pain lasting for a few seconds and of agonizing intensity. These paroxysms stop the patient in his tracks and may cause him to cry out and grip his arm and turn away. *Time Pattern:* frequency varies between a few an hour, a few a day, or a few a week. In a few patients they can be very frequent, with as many as 20 or 30 in an hour. There is no set pattern to the paroxysms, and the patient has no warning of their arrival. The constant pain may also be described as severe pins and needles and electric shocks, but it is most often burning or crushing. In some patients there is a gradual increase in the intensity of the pain over a period of days, building up to a very high level of pain lasting a day or more and then gradually subsiding over the next few days. In these patients the pain is particularly unpleasant and interferes seriously with their lives.

Associated Symptoms

Aggravating factors: cold weather, extremes of temperature, emotional stress, and intercurrent illness all aggravate the pain. The pain is almost invariably relieved by distraction involving absorbing work or hobbies. The pain is at its worst when the patient has nothing with which to occupy his mind. Patients often grip the anesthetic and paralyzed arm or hit the shoulder to try and relieve the pain. Drugs are singularly unhelpful and a full range of analgesics is usually tried, but very few patients respond significantly. Alcohol helps, probably by relaxing the patient and promoting sleep. A number of patients have found that smoking cannabis can markedly reduce the pain, but if so it interferes with their concentration, and very few indeed are regular cannabis smokers.

Signs

Paralysis and anesthetic loss in the territory of the avulsed nerve root, i.e., avulsion C5-T1-a totally paralyzed and insensitive arm. Most patients ask their doctors about amputation as a means of relieving the pain, and it has to be made clear to them the pain is central and amputation has no effect at all. In fact, there is a good likelihood of adding stump pain to their existing pain. Avulsion of T1 is associated with a Homer's sign, drooping of the eyelid and constriction of the pupil.

Myelography often shows evidence of meningoceles or root avulsion. Electrophysiological tests may well show the presence of sensory action potentials in anesthetic areas indicating that the lesion must be proximal to the posterior root ganglion. A flare response to intradermal histamine is occasionally useful, particularly in C5 lesions, again indicating preganglionic lesions.

Usual Course

Two-thirds of patients come to terms with their pain or say the pain is improved within three years of onset. If the pain is still severe at three years after onset, it is likely to last for the rest of their lives, and in these patients the pain steadily gets worse as they get older.

Complications

Rarely, trophic lesions of the anesthetic arm, e.g., burns or infections, or severe depression may occasionally follow prolonged pain, but it is remarkable how these young men manage to come to terms with their disability.

Social and Physical Disability

The major disability is the paralysis of the arm and the effect this has on work, hobbies, and sport. Pain itself can interfere with ability to work and can cut the patient off from normal social life.

Pathology

Avulsion is associated with spontaneous firing of deafferented nerve cells in the spinal cord at the level of the injury and may in time cause abnormal firing at higher levels of the central nervous system.

Summary of Essential Features and Diagnostic Criteria

The pain in avulsion lesions of the brachial plexus is almost invariably described as severe burning and crushing pain, constant, and very often with paroxysms of sharp, shooting pains that last seconds and vary in frequency from several times an hour to several times a week. So characteristic is the pain of an avulsion lesion that it is virtually diagnostic of an avulsion of one or more roots. Traction lesions of the brachial plexus that involve the nerve roots distal to the posterior root ganglion are seldom if ever associated with pain. Sometimes in regeneration spontaneously, or after nerve grafts for rupture of nerve roots distal to the intervertebral foramen, a causalgic type of pain develops, but this is highly characteristic of causalgia and cannot be confused with avulsion or deafferentation pain.

Code

203.X1c

Reference

Wynn Parry CB. Pain in avulsion lesions of the brachial plexus. Pain 1980;9:41–53.

Postradiation Pain of the Brachial Plexus (XI-4)

Code

203.X5

Painful Arms and Moving Fingers (XI-5)

See XXXI-9, Painful Legs and Moving Toes.

Code

202.X8

Reference

Verhagen WIM, Horstink MWIM, Notermans SLH. Painful arm and moving fingers. J Neurol Neurosurg Psychiatry 1985;48:384–98.

Brachial Neuritis (Brachial Neuropathy, Neuralgic Amyotrophy, Parsonage-Turner Syndrome) (XI-6)

Definition

Severe pain in shoulder and arm with progression to weakness and atrophy and, less frequently, numbness and paresthesias.

Site

Shoulder and upper limb.

System

Peripheral nervous system (brachial plexus).

Main Features

Severe sharp or burning nonlocalized pain in the entire upper extremity; this is usually unilateral but may be bilateral. It involves the proximal more frequently than the distal muscles.

Signs and Laboratory Findings

Diffuse weakness in nonroot and nondermatomal pattern with a patchy pattern of hypoesthesia. Laboratory tests of the spinal neuraxis are negative, but diffuse electromyographic abnormalities appear in the affected extremity with sparing of cervical paravertebral muscles.

Usual Course

Recovery is slow and may last one year or longer.

Summary of Essential Features

Onset of severe unilateral (or rarely bilateral) pain followed by weakness, atrophy, and hypoesthesia with slow recovery. The diagnosis is confirmed by positive electrodiagnostic testing and negative studies of the cervical neuraxis.

Differential Diagnosis

Avulsion of the brachial plexus; thoracic outlet syndrome.

Code

202.X8a

Bicipital Tendinitis (XI-7)**Definition**

Severe pain with acute onset due to inflammation of the long head of biceps tendon.

Site

Interior shoulder.

System

Musculoskeletal system.

Main Features

Severe pain, usually with acute onset in the anterior shoulder, following trauma or excessive exertion. It may radiate down the entire arm and is usually self-limited, but there may be recurrent episodes.

Aggravating Factors

Movement of shoulder and elbow.

Signs

Tendon palpation in the shoulder bicipital groove is painful. Pain is reproduced by resisted supination of the flexed forearm (Jergason's sign).

Usual Course

Occurs primarily after repeated use or heavy strain on tendon. It may become chronic.

Relief

Nonsteroidal anti-inflammatory agents; local steroid injection.

Complications

Frozen shoulder (adhesive capsulitis).

Pathology

Inflammation of the tendon sheath.

Essential Features

Acute pain in the anterior shoulder, aggravated by forced supination of the flexed forearm.

Differential Diagnosis

Subacromial bursitis, calcific tendinitis, rotator cuff tear.

Code
231.X3

Subacromial Bursitis (Subdeltoid Bursitis, Supraspinatus Tendinitis) (XI-8)

Definition

Aching pain in the shoulder due to inflammation of subacromial bursa.

Site

Shoulder and upper arm.

System

Musculoskeletal system.

Main Features

Age of Onset: common over 30 years of age. *Pain Quality:* the condition presents with aching pain in the deltoid muscle and upper arm above the elbow aggravated by using the arm above the horizontal level (painful abduction). The pain is aggravated by sleeping on the affected shoulder. It is usually precipitated by repeated or minor trauma.

Signs

Tenderness over the insertion of supraspinatus tendon. Painful arc of abduction, and internal rotation.

Radiologic Finding

High riding humeral head on X-ray when chronic attenuation of bursa occurs.

Usual Course

Recurrent acute episodes may produce chronic pain.

Relief

Nonsteroidal anti-inflammatory agents, local steroid injection, ultrasound, deep heat, physiotherapy.

Complications

Frozen shoulder (adhesive capsulitis).

Pathology

Chronic inflammation of bursa; tendon.

Essential Features

Aching pain in shoulder with inflammation of the subacromial bursa and exacerbation on movement as well as tenderness over the insertion of the supraspinatus tendon.

Differential Diagnosis

Calcific tendinitis, rotator cuff tear.

Code
238.X3

Rotator Cuff Tear—Partial or Complete (XI-9)

Definition

Acute severe aching pain due to traumatic rupture of supraspinatus tendon.

Site

Shoulder and upper arm.

System

Musculoskeletal system.

Main Features

Acute severe aching pain in the shoulder following trauma, usually a fall on the outstretched arm. Abduction is extremely painful or impossible. The patient is unable to sleep on the affected side.

Signs

A partial tear is distinguished from a complete tear by subacromial injection of local anesthetic; partial tears will resume normal passive range of motion. The arm may drop to the side if passively abducted to 90° (“drop arm sign”) if there is a complete tear.

Radiologic Finding

High riding humeral head on X-ray.

Complications

Frozen shoulder.

Essential Features

Acute severe pain due to trauma at the supraspinatus tendon.

Differential Diagnosis

Calcific tendinitis, subacromial bursitis.

Code

231.X1a

Adhesive Capsulitis and Frozen Shoulder (XI-10)

Code

232.X2	Infective
232.X3b	Inflammatory
232.X7	Dysfunctional

Lateral Epicondylitis (Tennis Elbow) (XI-11)

Definition

Pain in the lateral epicondylar region of the elbow due to strain or partial tear of the extensor tendon of the wrist.

System

Musculoskeletal system.

Main Features

Acute, subacute, or chronic pain of the elbow during grasping and supination of the wrist. *Age of Onset:* most common between 40 and 60 years of age. *Pain Quality:* pain radiates down the lateral forearm or to the upper arm.

Aggravating Factors

Repeated trauma.

Relief

Relieved by local steroid injection and physiotherapy.

Signs

Tenderness of the wrist extensor tendon about 5 cm distal to the epicondyle. Resisted wrist dorsiflexion reproduces pain.

Usual Course

Usually self limiting; several months duration.

Laboratory and Radiologic Findings

Negative.

Pathology

Strain or partial tear of tendon at tendoperiosteal junction.

Essential Features

Pain at the lateral epicondyle, worse on movement, aggravated by overuse.

Differential Diagnosis

Nerve entrapment, cervical root impingement, carpal tunnel syndrome.

Code

235.X1a

Medial Epicondylitis (Golfer's Elbow) (XI-12)**Definition**

Pain in the medial epicondylar region of the elbow.

Main Features

As for tennis elbow (XI-11) but much less common.

Aggravating Factors

As for tennis elbow.

Signs

Tenderness over the tendon insertion of the medial epicondyle.

Laboratory and Radiologic Findings

Negative.

Usual Course

As for tennis elbow.

Pathology

As for tennis elbow.

Differential Diagnosis

As for tennis elbow.

Code

235.X1b

DeQuervain's Tenosynovitis (XI-13)

Definition

Severe aching and shooting pain due to stenosing tenosynovitis of abductor pollicis longus or extensor pollicis brevis.

Site

Wrist.

System

Musculoskeletal system.

Main Features

Sudden onset of severe aching or shooting pains. There may be localized swelling and/or redness.

Aggravating Factors

Aggravated by pinch, grasping, or repetitive thumb and wrist movements.

Signs

Occasional tendon swelling; tenderness over the tendon in the anatomical snuff box area. Finkelstein's sign reproduces the pain; the patient's thumb is folded into a fist and then the wrist is deviated to the ulnar side.

Usual Course

May be single self-limited episode or recurrent and chronic.

Relief

Relief from local splinting or local steroid injection.

Pathology

Inflammatory lesion of tendon sheath usually secondary to repetitive motion or direct trauma.

Essential Features

Severe aching and shooting pain in the radial portion of the wrist related to movement.

Differential Diagnosis

Arthritis of the wrist, scaphoid injury.

Code
233.X3

Osteoarthritis of the Hands (XI-14)

Definition

Chronic aching pain in the fingers with degenerative changes of distal and proximal phalangeal joints of the hands.

System

Musculoskeletal system.

Main Features

The illness occurs mainly in women over 45 years of age. The pain is chronic and aching in the fingers and aggravated by use and relieved by rest. There may be mild morning stiffness for less than half an hour and subjective reduction of grip strength, worse with trauma to nodes.

Signs

Bony enlargements of the distal interphalangeal joints are called Heberden's nodes, and those of the proximal interphalangeal joints are called Bouchard's nodes. The fingers may be stiff and lose some degree of full flexion. Grip strength is usually normal when measured.

Radiologic Finding

Narrowing of joint spaces, sclerosis, and bony osteophytosis.

Relief

Analgesics, soaking in hot fluids.

Code
238.X6b

Cubital Tunnel Syndrome (XI-15)

Definition

Entrapment of the ulnar nerve in a fibro-osseous tunnel formed by a groove (trochlear groove) between the olecranon process and medial epicondyle of the humerus. The groove is converted to a tunnel by a myofascial covering, and the etiology of the entrapment is multiple.

Site

Elbow, forearm, and fingers (fourth and fifth).

System

Peripheral nervous system (ulnar nerve).

Main Features

Gradual onset of pain, numbness, and paresthesias in the distribution of the ulnar nerve, sometimes followed by weakness and atrophy in the same distribution; often seen in conjunction with a carpal tunnel syndrome ("double crush phenomenon").

Signs and Laboratory Findings

Tinel's sign at the elbow. The ulnar nerve is frequently thickened and adherent. On electrodiagnostic testing there is slowing of conduction in the ulnar nerve across the elbow, accompanied by denervation of those intrinsic muscles of the hand innervated by the ulnar nerve.

Usual Course

The course may be stable or slowly progressive; if the latter, surgery is necessary, either decompression or transposition of the nerve.

Summary of Essential Features and Diagnostic Criteria

A gradual onset of pain, paresthesias, and, at times, motor findings in the distribution of the ulnar nerve. Tinel's sign is found. The diagnosis is confirmed by slowing of conduction across the elbow and often by denervation of those intrinsic muscles of the hand innervated by the ulnar nerve.

Differential Diagnosis

Thoracic outlet syndrome, carpal tunnel syndrome.

Code

202.X6c

Carpal Tunnel Syndrome (XI-16)**Definition**

Stinging, burning, or aching pain in the hand, often nocturnal, due to entrapment of the median nerve in the carpal tunnel.

Site

One hand (sometimes bilateral), in the fingers, often including the fifth digit, often spreading into the forearm and occasionally higher; not usually well localized.

System

Peripheral nervous system.

Main Features

Prevalence: very common. *Age of Onset:* usually fourth to fifth decades. *Sex Ratio:* female to male 5:1. *Quality:* pins and needles, stinging, often aching, occasionally burning. *Time pattern:* usually nocturnal, typically awakening the patient several times and then subsiding in a few minutes; aching pain is often more constant. *Intensity:* may be severe briefly.

Associated Symptom

Aggravated by handwork such as knitting.

Signs and Laboratory Findings

Clinical examination often normal, but one may find decreased pin-prick sensation on the tips of digits I-III, a positive Tinel's or Phalen's sign, or rarely, weakness and/or atrophy of the thenar muscles (abductor pollicis brevis); nerve conduction studies showing delayed sensory and motor conduction across the carpal tunnel are diagnostic.

Usual Course

Very slow progression for years.

Social and Physical Disability

May impair ability to do handwork.

Pathology

Compression of median nerve in wrist between the carpal bones and the transverse carpal ligament (flexor retinaculum); focal demyelination of nerve fibers, axonal shrinkage and axonal degeneration.

Summary of Essential Features and Diagnostic Criteria

Episodic paresthetic nocturnal pain in the hand with electrophysiological evidence of delayed conduction in the median nerve across the wrist.

Code

204.X6

Pain of Psychological Origin in the Shoulder and Arm (XI-17)

Code

233.X7b	Tension: arm
21X.X9a	Delusional: arm
21X.X9b	Conversion: arm
21X.X9d	Associated with depression

Crush Injury of Upper Limbs (XI-18)

Code

231.X1b

Pain in a Limb or Limbs, Not Otherwise Specified (XI-19)

Code

2XX.XXz	Upper limb or limbs
6XX.XXz	Lower limb or limbs

GROUP XII: VASCULAR DISEASE OF THE LIMBS

Raynaud's Disease (XII-1)

Definition

Episodic attacks of aching, burning pain associated with vasoconstriction of the arteries of the extremities in response to cold or emotional stimuli.

Site

Predominantly in the hands, unilateral initially, later bilateral. Rarely lower limbs and exposed areas of face.

System

Cardiovascular system.

Main Features

Prevalence: Raynaud's phenomena can occur in 5% of normal females as secondary to connective tissue disease. Raynaud's disease is uncommon, with a female to male ratio of 5:1. *Onset:* most common between puberty and age 40. Exacerbations during emotional stress and possibly at time of menses. *Start:* evoked by cold, nervousness, and other stimuli which vary among patients. A typical attack occurs in three phases. Initially the digits become ashen white, then they turn blue as the capillaries dilate and fill with slowly flowing deoxygenated blood. Finally the arterioles relax and the attack comes to an end with a flushing of the diseased parts. *Pain Quality:* initially the pain is deep and aching and varies from mild to severe, changing to severe burning dysesthesias in the phase of reactive hyperemia. *Time Pattern:* recurring irregularly with changes in environmental temperature and emotional status. *Intensity:* variable from mild to severe depending upon the temperature and other stimuli. *Duration:* minutes to hours. Sometimes may last days if painful ischemia skin ulcers develop.

Associated Symptoms and Signs

Numbness or hypoesthesia are present. Progressive spasm of the vessels leads to atrophy of the tip, giving the finger a tapered appearance. The nail becomes brittle and paronychia is common. Advanced cases may develop focal areas of necrosis at the fingertip, occasionally preceded by cutaneous calcification. These areas are extremely painful and tender to palpation. Anxiety and other signs of sympathetic overactivity such as increased sweating in the limbs and piloerection develop.

Relief

Temporary relief from sympathetic block, and occasional prolonged relief from sympathectomy in the early phases. Calcium channel blocking agents may help.

Pathology

The cause of "cold sensitivity" is unknown. Abnormalities in sympathetic activity have not been proven. However, local application of cold is necessary to elicit the response of Raynaud's syndrome, and the threshold for triggering the response is lowered by any factor that increases sympathetic outflow or circulating catecholamines.

Essential Features

Color changes of digits, excited by cold or emotions, involving both upper extremities and absence of specific organic disease.

Differential Diagnosis

- Raynaud’s disease, which has no other known cause, and Raynaud’s phenomenon, which is a response occurring in other illnesses, should be distinguished. The following other diseases should be recognized:
- collagen-vascular diseases: scleroderma, rheumatoid arteritis, systemic lupus erythematosus, dermatomyositis, periarteritis nodosa;
- other vascular diseases: thromboangiitis obliterans, thrombotic or embolic occlusion, arteriosclerosis obliterans, syphilitic arteritis;
- trauma: vibration (air-hammer disease, etc.), percussion (digital pianist, typist, etc.), palmar (hypothenar hammer syndrome);
- neurovascular syndromes: thoracic outlet syndromes, spondylitis, causalgia;
- central and peripheral nervous disorders (rarely): syringomyelia, poliomyelitis, ruptured cervical disk, progressive muscular atrophy;
- cold injury: frostbite, nonfreezing cold injury, (pernio, immersion foot), cold sensitivity syndrome;
- lack of suspension stability of blood: cold agglutinins, cryoglobulinemia, cryofibrinogenemia, polycythemia vera;
- intoxication: ergot, arsenic, heavy metals (lead), nicotine, and tobacco.

Code

024.X7a	Face
224.X7a	Arms
624.X7b	Legs

Raynaud’s Phenomenon (XII-2)

Definition

Attacks like those of Raynaud’s disease but related to one or more other disease processes.

Usual Course

In accordance with the underlying disease.

Pathology

Systemic and vascular diseases such as collagen disease, arteriosclerosis obliterans, nerve injuries, and occupational trauma—for example in chain saw operators, pianists, and pneumatic hammer operators—may all contribute to the development of Raynaud’s phenomenon.

Code

024.X7c	Face
224.X7c	Arms
624.X7c	Legs

Frostbite and Cold Injury (XII-3)

Definition

Severe burning pain in digits or exposed areas of face due to cold injury.

Site

Periphery of limbs (digits) and exposed areas of face.

System

Cardiovascular system.

Main Features

Prevalence: increased incidence in elderly patients with arterial disease and in young men with hazardous exposure to cold environment, e.g., soldiers, mountaineers. *Start:* frostbite commences with an initial vasospastic phase with pallor and numbness, followed by cyanosis. Rubor only returns on rewarming. Signs and severity vary steadily with degree of cold exposure, see below. *Pain Quality:* at time of exposure, numbness and tingling of digits and severe aching pain occur. After a few days, severe burning or stinging pain, particularly after exposure to warmth. Then pain becomes a deep aching or throbbing which may persist for many weeks. *Time Pattern:* single episode after cold exposure or recurring episodes if there is a predisposition to cold injury. *Intensity:* mild initially, then severe after a few days if limb warmed. *Duration:* usually two to three weeks to eight weeks, but pain can become chronic.

Associated Symptoms

In chronic stages: sometimes hyperesthesia and increased sweating, increased sensitivity to cold, numbness, aching, paresthesias, and dysesthesias.

Signs and Usual Course

First degree frostbite: edema, erythema, and hypoesthesia lasting two to three weeks followed by superficial desquamation. *Second degree frostbite:* vesicles and blisters in superficial skin layers. In two to three weeks vesicles dry and leave thickened epithelium (in absence of infection). *Third degree frostbite:* involves full skin thickness. Hard black scar develops and separates in about eight weeks. *Fourth degree frostbite:* results in deep tissue necrosis down to bone and requires amputation of the affected area.

Complications

Infections leading to cellulitis, tetanus, and gas gangrene are unlikely unless contamination occurs after rewarming; amputation may be required for gangrenous extremities after fourth degree injury; persistent cold sensitivity; paresthesias; hyperhidrosis and burning pain which may be prevented or relieved by sympathetic block or denervation.

Social and Physical Disability

Restriction of use of limbs due to cold sensitivity, hyperhidrosis, and pain.

Pathology

On initial exposure to cold, intense vasoconstriction occurs in extremity areas and results in reduced microcirculation flow with sludging of red cells; eventually flow ceases at the onset of freezing. Frozen tissue is bloodless, hard, cold, and pale. As tissues thaw, vasodilation occurs and flow is resumed; however, interstitial edema restricts flow, and white emboli dislodge from injured vessel walls and mix with platelets to form thrombi at venular bifurcations, and this obstructive process extends through to precapillary arterioles so that within one hour most of these microvascular channels are occluded.

Tissue necrosis is attributed to mechanical effects of microvascular occlusion, to extracellular ice crystals, and to cellular dehydration.

Essential Features

Exposure to cold below 0°C followed by tissue injury a variable period after exposure.

Differential Diagnosis

Erythema pernio (chilblains), trench foot, immersion foot, cold sensitivity, cold agglutinin syndrome, cryoproteinemia. Persistent pain and hyperhidrosis following frostbite may simulate causalgia.

Code

222.X1a Arms
622.X1a Legs

Erythema Pernio (Chilblains) (XII-4)

Definition

Pain and itching in areas of extremities following exposure to cold and wet environment above 0°C and associated with pigmented or purpuric skin lesions.

Site

Digits and limbs, especially lower limbs.

Main Features

Similar to first degree frostbite except that women are more susceptible (especially those with “sensitivity to cold”). At time of exposure numbness and tingling of digits may occur. Redness and itching of the skin is a feature, together with excessive sweating. The pain is often mild but may be associated with intense itching and with burning sensations. Pernio tends to be seasonal in occurrence, associated with cold exposure. The pain is always aggravated by warmth.

Associated Symptom

Blebs filled with clear or bloody fluid may form, and pigmented or purpuric lesions may develop.

Differential Diagnosis

Erythema nodosum, erythema induratum, Raynaud’s disease, and acrocyanosis.
N.B. Acrocyanosis is painless.

Code

225. XI Arms
625.XI Legs

Acrocyanosis (XII-5)

Definition

Persistent blueness and coldness of hands and feet, sometimes with aching pain.

Site

Hands and feet, especially digits.

System

Vascular system.

Main Features

Blueness and coldness, more common in women, sometimes aching pain, often not.

Associated Features

Often chilblains. Trophic changes, ulceration, and gangrene do not occur. Symptoms are more marked in cold periods. Commences in late childhood and seems to have a hereditary background.

Code

222.X1b Arms
622.X1b Legs

References

van Nuys Allen E, Barker NW, Hines EA, Juergens JL, Spittell JA, Fairbairn JF. Peripheral vascular diseases, 5th ed. Philadelphia: W.B. Saunders; 1980.

Livedo Reticularis (XII-6)

Definition

Common, possibly vasospastic disorder in women under the age of 40; associated with persistent aching in the skin of the arms and itching of circular and reticular lesions which have a mottled cyanotic appearance.

Site

Upper or lower limbs.

Main Features

Occurring in women, the more severe form (cutis marmorata) is associated with persistent aching in the skin of the arm. Itching circular and reticular lesions with a mottled cyanotic appearance are evident.

Differential Diagnosis

Pernio, other skin changes.

Code

222.68a
622.68c

Volkmann's Ischemic Contracture (XII-7)

Code

222.X5

GROUP XIII: COLLAGEN DISEASE OF THE LIMBS

Scleroderma (XIII-1)

Definition

Intermittent vasospasm often with soreness, stiffness, or swelling of peripheral joints of the fingers and toes due to collagen disease of the skin, particularly affecting the limbs.

Main Features

Incidence: 3-5 new cases per million per annum. *Sex Ratio:* male to female 1:3. *Age of Onset:* from young adult life onward. *Pain:* is not a great problem in most cases. However, pain can occur intermittently with soreness and pain of Raynaud's phenomenon, especially aching pain in episodes ranging from mild to severe and changing to burning dysesthesias in the phase of reactive hyperemia. Numbness or hypoesthesias are present also.

Associated Symptoms and Signs

Stiffness and swelling of peripheral joints of the fingers and toes. A tight skin which may or may not be thickened. Skin temperature is often lowered, and the underlying tissues seem firm. Fingers although swollen and stiff can be moved with difficulty. The skin appears pale and waxy, skin temperature is lowered in the affected parts, and although pulses are palpable at the wrist, there is usually complete arterial obstruction in the digits. Microstomia and multiple telangiectasia may be observed over the face and hands.

Essential Features

Evidence of scleroderma with Raynaud's phenomenon.

Differential Diagnosis

See Raynaud's Disease (XII-1) and Raynaud's Phenomenon (XII-2).

Code

226.X5	Arms
626.X5	Legs

Ergotism (XIII-2)

Definition

Burning pain in the extremities, identical to Raynaud's phenomenon, associated also with systemic symptoms attributable to excessive ergot intake.

Site

Fingers and toes especially; viscera are occasionally involved also.

Main Features

Occurs in patients taking excess ergotamine tartrate or others (rarely) who have eaten rye or wheat contaminated by ergot. Uncommon in general. Presents with burning pain in the extremities identical to Raynaud's phenomenon.

Three stages can be seen in the changes in the circulation: (1) a stage of cyanosis or pallor from which recovery is rapid; (2) a stage of deep purple coloration in which blanching cannot be effected by pressure

and from which recovery may be slow or may not occur; and
(3) a stage of necrosis.

Severe cases of ergot intoxication are however sporadic. Symptoms can consist of dizziness, frontal headache, angina pectoris, Raynaud's phenomenon, coldness of the extremities, and pain. Both neurologic and vascular symptoms may produce the feeling of intense heat and cold, burning pains, known in the past as St. Anthony's fire.

Associated Symptom

Headaches, dizziness, nausea and vomiting, visual disturbances, angina pectoris, mono- or hemiplegia. May result in gangrene.

Usual Course

On discontinuation of ergot administration, pulses and signs of ischemia return to normal in 1 to 3 days. In stages 2 and 3, more vigorous therapy is needed with anticoagulant and vascular dilatation agents.

Complications

Gangrene. In some cases residual anesthesia of the skin or paralysis of the extremities may persist.

Pathology

Ergot intoxication results in constriction of the arteries. Because of the vasoconstriction, the endothelium of the vessels suffers, stasis occurs in the capillaries, and thrombosis follows. After thrombosis gangrene is inevitable. Actual intoxication is not necessary for diminution of arterial pulses. The chronic use of therapeutic doses leads to lowered foot systolic blood pressure. The degree of tolerance to the vasoconstrictive effects varies widely.

Summary of Essential Features and Diagnostic Criteria

Color changes of digits, burning pain as described, evidence of excessive ingestion of ergotamine.

Differential Diagnosis

See Raynaud's Disease (XII-1).

Code

281.X6	Arms
681.X5	Legs

References

Dukes MNG, editor. Meyler's Side Effects of Drugs, 9th ed. Amsterdam: Excerpta Medica; 1980.

Goodman LS, Gilman A. The Pharmacological Basis of Therapeutics, 7th ed, New York: Macmillan; 1985.

van Nuys Allen E, Barker NW, Hines EA, Juergens JL, Spittell JA, Fairbairn JF. Peripheral Vascular Diseases, 5th ed. Philadelphia: W.B. Saunders; 1980.

GROUP XIV: VASODILATING FUNCTIONAL DISEASE OF THE LIMBS

Erythromelalgia (XIV-1)

Definition

Episodic burning pain in the extremities accompanied by bright red discoloration in response to increased environmental temperature.

Site

Extremities of the limbs, but almost always the feet rather than the hands.

System

Vascular system.

Main Features

Primary form rare and more often bilateral than the secondary type, which is related to the frequency of the conditions. Men in the middle-age group are more often involved, but women and children may also be affected. Characterized by severe, burning pain and red discoloration. The skin temperature is often raised, the skin flushed with venous engorgement, and the surface hyperesthetic. Attacks of severe burning pain last from a few minutes to many hours.

Associated Symptoms

Arteriosclerosis, hypertension, peripheral neuritis, cold injury, polycythemia, disseminated sclerosis, infections, hemiplegia, gout, or chronic heavy metal poisoning may be present. Ulcers or gangrene rare in primary type.

Signs and Laboratory Findings

Diagnosed by reproducing symptoms after raising skin temperature to 31-36°C. Relief from cooling. Skin cyanotic or deeply red in response to rise of temperature.

Pathology

Cause of most cases unknown. Secondary erythromelalgia may result from disorders listed above.

Differential Diagnosis

Burning pain which comes in attacks and affects the foot-sole or palm of the hand, closely related to objective increased local skin temperature. Reduction of pain by elevating or cooling the affected extremity.

Code

224.X8d	Hands	<i>Note:</i> add code for secondary case according to etiology.
624.X8d	Feet	

References

Fitzpatrick TB. Dermatology in general medicine: textbook and atlas, 2nd edition. New York: McGraw-Hill; 1977.

Kappert A. Lehrbuch and atlas der angologie, 1st ed. Bern, Switzerland: Verlag Hans Huber; 1981.

van Nuys Allen E, Barker NW, Hines EA, Juergens JL, Spittell JA, Fairbairn JF. Peripheral vascular diseases, 5th ed. Philadelphia: W.B. Saunders; 1980.

Thromboangiitis Obliterans (XIV-2)

Definition

Pain in the fingers or hands or small digits of the feet, usually in males who smoke; associated with ulceration of fingertips and margins of nails; related initially to segmental inflammation of walls of medium and small arteries and veins.

Site

Fingers and hands, more often toes and feet, rarely the calf.

System

Cardiovascular system.

Main Features

Prevalence: a rare disease with a possible preponderance in Jews. Close association with smoking. *Sex Ratio:* males more than females-ratio above 9:1. *Age of Onset:* usual age of onset between 30 and 40 years. *Pain Quality, Time Pattern, Intensity:* usual onset is sharp pain in fingers or hands or more often in the foot or calf. There may be episodes of intermittent claudication in the hands or legs or constant burning in the tips of the digits (rest pain). *Intensity:* may be unbearable, often aggravated by elevation.

Associated Symptoms

Tenderness in superficial arteries, veins or nerves in affected area. Cold extremities (upper and lower), dysesthesias, and muscle weakness.

Signs

Coldness and sensitivity to cold, sensations of numbness, paresthesias, sometimes superficial thrombophlebitis. Ulceration of fingertips and margins of nails, gangrene of digits which may be wet gangrene if there is venous obstruction; edema present if there is venous obstruction. Absent ulnar or tibial artery pulsation and positive Allen test in cases affecting the arms (see Thoracic Outlet Syndrome [VII-4] for Allen test). Abnormal color of skin of digits: pale if elevated, red when first dependent, then blue. Cyclic color changes may occur in response to cold (Raynaud's phenomenon).

Laboratory Finding

Thermogram shows differences in temperature of digits. Skin plethysmography shows reduced blood flow in one or more digits, indicating local arterial disease. Tobacco sensitivity may be demonstrated. Vigorous muscle contraction of the digit may result in sufficient pressure to overcome intravascular pressure with cessation of blood flow as measured by plethysmogram. Doppler test may be helpful.

Usual Course

Gradual progression from age 30 for many decades. Favorably influenced if smoking ceases.

Complications

Gangrene and infection of digits. Osteoporosis of bones of extremities.

Pathology

Ulnar, palmar, and digital arteries affected early with segmental inflammation initially. Arteries are contracted and hard and lumens occluded by adherent mass. Arteries and veins bound together by inflammatory tissue. Acute stage: granulation tissue in all layers of affected arteries (pan-arteritis) and usually a thrombus in vessel lumen. Subacute stage: thrombus recanalized. Chronic stage: sclerotic

thrombus, dense fibrous tissue encloses arteries, veins, and nerves.

Summary of Essential Features and Diagnostic Criteria

Organic arterial disease of one or more digits, almost always in a male under 40 with a history of migrating superficial thrombophlebitis.

Differential Diagnosis

Arteriosclerosis (larger vessels and more widespread), periarteritis nodosa (veins not involved), giant cell arteritis (mainly branches of carotid), thoracic outlet syndrome.

Code

224.X3c	Arms
624.X3b	Legs

References

Haimovici M. Vascular surgery: principles and techniques. New York: McGraw-Hill; 1976.

Rutherford RB. Vascular surgery. Philadelphia: W.B. Saunders; 1977.

van Nuys Allen E, Barker NW, Hines EA, Juergens JL, Spittell JA, Fairbairn JF. Peripheral vascular diseases, 5th ed. Philadelphia: W.B. Saunders; 1980.

Chronic Venous Insufficiency (XIV-3)

Definition

Dull, aching pain in limbs, especially legs, characterized by abnormally dilated or tortuous veins.

Site

Limbs, usually the legs; especially the distal portions. Found equally on right or left sides. Bilateral in 7%.

Main Features

Prevalence: about 15% of adult population, severe in only 1%. *Sex Ratio*: more common in women. Prevalence increases with age, and there may be a hereditary predisposition. *Pain Quality*: dull, aching pain, usually associated with varicosities. Additional pain often due to thrombosis and/or thrombophlebitis acutely.

Associated Symptoms

Feelings of heaviness, numbness of the skin. Previous thrombophlebitis in a vein of the extremity, orthostasis with edema, developing during the day and disappearing during the night when the patient lies flat. After edema has been present for some time, areas of brown pigmentation (hemosiderin and melanin) may appear. Eczema is a common feature. After longer periods there is a tendency toward the development of subcutaneous fibrosis with induration and swelling.

Signs and Laboratory Findings

Edema, dilated superficial veins, varicosities, corona phlebectatica, hyper- and de-pigmentation, induration, open or healed ulcer cruris. In obscure or borderline cases phlebography is required.

Usual Course

Chronic, but dependent on stage of insufficiency and reaction on causal therapy.

Relief

Relief, even of ulcer pains, occurs gradually as a result of recumbency and more quickly if the extremity is elevated (relief after 5-30 minutes). Causal therapy for ulcers and dermatitis is indicated.

Complications

Ulceration. Neuritis of n. saphena longus is occasionally seen as a complication of chronic venous insufficiency.

Social and Physical Disability

Dependent on degree of insufficiency.

Pathology

Chronic venous insufficiency is the late consequence of extensive damage of the deep veins by thrombosis, in a given case, thrombophlebitis. The aching pain is associated with edema largely of the subcutaneous tissues. The more epicritic pain of ulcers and indurative cellulitis is usually due to secondary inflammation rather than congestion.

Etiology

Hereditary factors, blockage by thrombosis or other disease (rarely carcinoma).

Essential Features

Signs of venous insufficiency. Deep venous thrombosis in history.

Code

222.X4	Arms: neoplasm
222.X6	Arms
622.X4	Legs: neoplasm
622.X6	Legs

References

Haimovici M. Vascular surgery: principles and techniques. New York: McGraw-Hill; 1976.

Rutherford RB. Vascular surgery. Philadelphia: W.B. Saunders; 1977.

van Nuys Allen E, Barker NW, Hines EA, Juergens JL, Spittell JA, Fairbairn JF. Peripheral vascular diseases, 5th ed. Philadelphia: W.B. Saunders; 1980.

Widmer LK. Peripheral venous disorders. Bern, Switzerland. Hans Huber Publishers; 1978.

GROUP XV: ARTERIAL INSUFFICIENCY IN THE LIMBS

Intermittent Claudication (XV-1) and Rest Pain (XV-2)

Definition

Intermittent cramping pain in a muscular area produced by exercise and relieved by rest (XV-1), or constant pain in an extremity associated with hypoesthesia and/or dysesthesia and areas of skin ulceration or gangrene (XV-2).

Site

Intermittent claudication (pain after exercise) is almost always confined to the lower limbs. Pain from arterial insufficiency arising at rest may occur in lower limbs or upper limbs and may be related to gravity.

System

Cardiovascular system.

Main Features

Sex Ratio: males affected earlier than females. *Age of Onset:* over 30, increasing in later middle age and decreasing in the aged. *Pain Quality:* the intermittent pain is cramping and severe and arises, usually, after fixed and consistent amounts of exercise. Severe ischemia will result in rest pain and night pain. The pain is relieved by the dependent position, which initially causes the limb to flush red and then become cyanotic. Elevation of the limb causes blanching and increased pain.

Associated Symptoms

Hypothyroidism or myxedema, diabetes mellitus, hypercholesteremia, hyperlipidemia, xanthomatosis, and long-standing heavy smoking may be found. Occlusion leads to ulceration gangrene, etc.

Associated hypertensive ischemia ulceration: In patients with hypertension of long duration, ulceration of skin results from insufficiency of small arteries or arterioles. More common in women of age 45-70 years. Lesions usually in skin of legs but sometimes in the upper limbs.

Signs

A systolic murmur may be heard over the abdominal aorta or iliac arteries. Pain is relieved by the dependent position which initially causes the limb to flush red and then become cyanotic. Elevation of the limb causes blanching and increased pain. Absent or diminished pulses, reduced skin temperature, and coldness of the limb are characteristic.

Laboratory Finding

Arteriography demonstrates the level of arterial obstruction or obstructions.

Usual Course

May gradually progress so that the patient can walk less far. Sudden progression indicates acute occlusion of main or collateral arteries.

Relief

Relief may be provided by sympathectomy for rest pain; claudication is less often relieved by this technique.

Complications

Ulceration, gangrene.

Pathology

Claudication intermittens is a symptom that always indicates an inadequate supply of arterial blood to contracting muscle. Atherosclerosis is usually the underlying condition.

Etiology

May be due to (a) arteriosclerosis, characterized by local deposition of fat under and within the intima of arteries, most commonly the aorta, coronary, cerebral arteries; (b) arteriosclerosis obliterans, an obstructive late stage of atherosclerosis characterized by partial or complete occlusion of arteries by atheromata, often with superimposed thrombosis: more common in men, involves large vessels such as aorta and arteries of lower limb; or (c) Monckeberg's medial calcification and sclerosis, much more common in men, patients usually over 50 years of age. Changes confined to muscular media of medium-sized arteries, e.g., radial or ulnar, leading to nodularities around the artery. Only rarely leads to obstruction of arteries. Intermittent claudication and rest pain are more benign with this type of arterial disease than with others.

Essential Features

Exercise-induced pain which passes off very quickly by rest. Signs of arterial or arteriolar insufficiency.

Differential Diagnosis

Neurogenic claudication intermittens. Arterial or arteriolar vascular insufficiency by other conditions like entrapment syndromes, arteriospastic or arteritic conditions.

Code

224.X8c	Intermittent claudication: arms
624.X8c	Intermittent claudication: legs
222.X8b	Rest pain: arms
622.X8b	Rest pain: legs

References

Silva DA, Widmer LK. Peripheral venous disorders. Bern, Switzerland: Hans Huber; 1978.

van Nuys Allen E, Barker NW, Hines EA, Juergens JL, Spittell JA, Fairbairn JF. Peripheral vascular diseases, 5th ed. Philadelphia: W.B. Saunders; 1980.

Gangrene Due to Arterial Insufficiency (XV-3)

Code

222.X8d	Arms
622.X8d	Legs

GROUP XVI: PAIN OF PSYCHOLOGICAL ORIGIN IN THE LOWER LIMBS (XVI-1)

General Descriptions See section 1-16.

Pain of psychological origin in the limbs is rarely considered to be due to muscle tension. Delusional or conversion pain in these (and other) locations may be more common on the left. Recurrent or chronic limb pain due to inappropriate use of muscle groups whether or not for psychological reasons may be quite common.

Code

633.X7c	Tension: legs
61X.X9d	Delusional: legs
61X.X9e	Conversion: legs
61X.X9f	With depression: legs

References

Hall W, Clarke IMC. Pain and laterality in a British pain clinic sample. *Pain* 1982;14:63-6.

Merskey H, Watson GD. The lateralization of pain. *Pain* 1979;7:271-80.