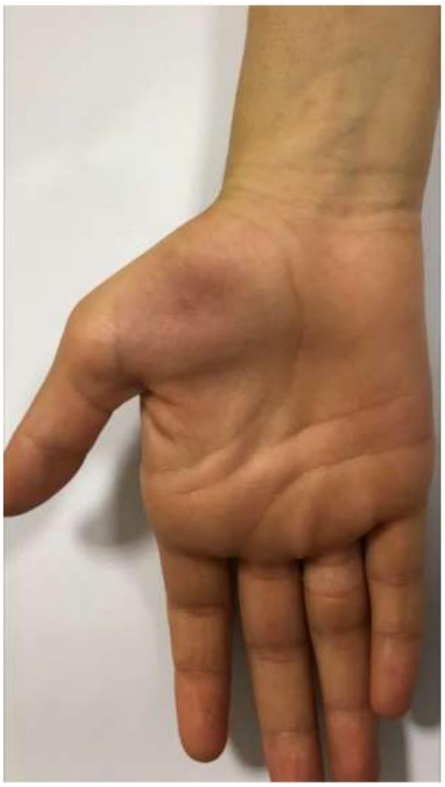
Hirayama Disease and Homoeopathy



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Hirayama disease is a focal motor neuron disease, a rare cervical myelopathy, predominantly affecting young men, and which presents with distal atrophy of the upper limbs as its first and main symptom. (Psora/ Syphilis)

Etymology

This disease was initially recognized in Japan in 1959 by Hirayama et al. and was reported under the name of juvenile muscular atrophy of unilateral upper extremity.

Synonyms

- HD
- Benign focal amyotrophy
- Cervical myelopathy related to flexion movements of the neck
- Juvenile asymmetric segmental spinal muscular atrophy
- Juvenile muscular atrophy of the distal upper extremity
- Monomelic amyotrophy (MMA)
- Non-progressive juvenile spinal muscular atrophy of the distal upper limbs

Definition

Hirayama disease is a benign motor neurone disorder with a stationary stage after a progressive course, caused by cervical myelopathy related to flexion movements of the neck (Psora/ Syphilis/ Causa occasionalis), causing non-progressive juvenile spinal muscular atrophy (Syphilis) of the distal upper limbs, characterized by the insidious onset of self-limited, unilateral or asymmetric oblique amyotrophy (Syphilis) that affects the C7, C8 and T1 myotomes, leading to atrophic weakness of the forearms and hands (Syphilis) predominantly in young males.

Causes

The relatively short and tight dura mater (Psora) seen in patients with HD is unable to compensate for the increased length of the vertebral canal during neck flexion (Causa occasionalis). The forward displacement of the posterior dura of the lower cervical dural canal during neck flexion has been postulated to lead to lower cervical cord atrophy with asymmetric flattening.

Pathophysiology

Exact pathophysiology of Hirayama disease is not known, but following changes are considered-

- Cell shrinkage and necrosis (Syphilis)
- Various degrees of degeneration of small and large nerve cells (Syphilis)
- Mild gliosis (Psora/ Syphilis)
- Some circulatory insufficiency in the anterior horns of the spinal cord from the lower cervical to upper thoracic levels, particularly at the C7 and C8 levels (Psora/ Syphilis)
- Atopy and elevated serum IgE level have also been postulated to be precipitating factors in HD (Syphilis)
- The most widely accepted hypothesis is a cervical myelopathy associated with neck flexion (Psora/ Syphilis/ Causa occasionalis)

Normally, the spinal dura mater is loosely anchored to-

- The vertebral canal by the nerve roots

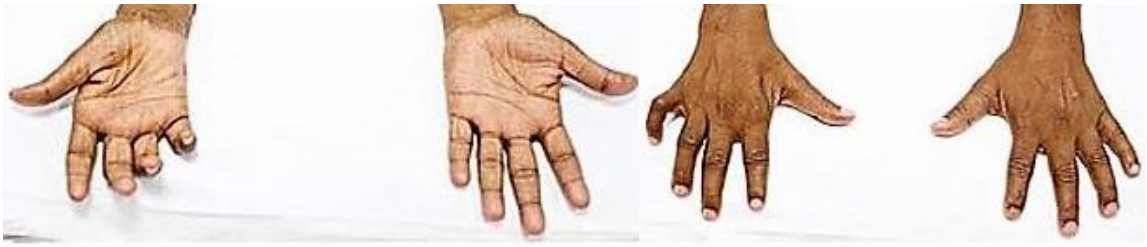
- The periosteum at the foramen magnum
- The dorsal surfaces of c2 and c3 and
- The dorsal surface at the coccyx

The comparatively short and close-fitting dura mater seen in patients with HD is unable to compensate for the increased length of the vertebral canal during neck flexion. This results in tightening of the dural canal during neck flexion, which leads to an anterior shift of the posterior dural wall, causing spinal cord compression against the vertebral body. This repeated neck flexion results in multiple incidents of ischaemia and chronic trauma to the spinal cord, which ultimately leads to myelopathy.

Signs and symptoms

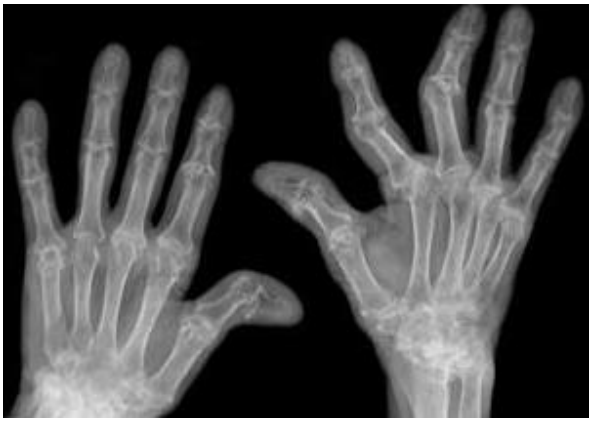
The disease usually progresses for few years (1–3) and then is followed by arrest of progression, rendering a relatively benign course. It is characterized by-

- Insidious onset asymmetrical weakness (Psora/ Syphilis)
- Wasting of muscles of upper limb, affecting predominantly C7, C8, and T1 myotomes
- Male preponderance
- Between 15 and 25 years of age
- Patient feels contracture of middle and ring finger
- Slight thinning of the sub digital Palm of the affected fingers



The clinical features may also include-

- Irregular coarse tremors, called minipolymyoclonus in the fingers of the affected hands with mild transient worsening of symptoms on exposure to cold
- The sensory, reflex, and cranial nerve examinations generally normal
- Pyramidal tract involvement in lower limb, autonomic disturbances, and cerebellar deficits rare
- EMG of the affected muscles showing chronic denervation, with or without acute denervation changes like- fasciculations, positive sharp waves, fibrillations potentials etc.
- Apparently healthy muscles may also show abnormal EMG findings
- Distal predominant muscle weakness and atrophy in forearm and hand
- Involvement of the unilateral upper extremity almost always all the time
- Insidious onset with gradual progression for the first several years, followed by stabilization
- No lower extremity involvement
- No sensory disturbance and tendon reflex abnormalities
- Sparing of brachioradialis muscle, giving the impression of an oblique atrophy
- Excessive sweating of both palms, suggestive of autonomic dysfunction
- Hypertonia of lower limbs with exaggeration of deep tendon reflexes with positive Babinski's sign, suggestive of UMN lesion



Differential diagnosis

Amyotrophic lateral sclerosis (ALS)

It is a progressive neurodegenerative disease that affects nerve cells in the brain and the spinal cord.

Anterior interosseous or deep ulnar neuropathy

Anterior interosseous nerve syndrome (also called Kiloh-Nevin syndrome) is caused by entrapment or compression of the anterior interosseous nerve in the proximal part of the forearm. Most lesions that lead to this syndrome have a location distal to that typical of lesions that cause pronator syndrome.

Brachial plexopathy

Brachial plexopathy is a neurologic affliction that causes pain or functional impairment or both of the ipsilateral upper extremity and may result from medical conditions and from violent stretching, penetrating wounds, or direct trauma.

Cervical vertebral abnormalities

Atlanto-occipital assimilation

Occipitalization is a congenital synostosis of the atlas to the occiput caused by a failure of segmentation and separation of the most caudal occipital sclerotome during the first few weeks of foetal life.

Block vertebra (Congenital Synostosis, Congenital Vertebral Fusion)

This results from the embryological failure of normal spinal segmentation. Patients are generally asymptomatic but increasing age and injury may precipitate symptoms. Premature degenerative changes at adjoining motion segments are common as this condition results in greater biomechanical stress in the adjoining segments. Discal tear, rupture of the transverse ligament, fracture of the odontoid process, and spondylosis are common consequences.

Os Odontoideum

Os odontoideum is defined as non-union of the dens with the axis body. It is associated with atlantoaxial instability, local mechanical irritation (neck pain and torticollis), progressive myelopathy, or transient neurologic symptoms secondary to vertebral artery compression.

Atlantodental Interval (ADI)

The odontoid process is supported by the transverse ligament posteriorly. If the atlantodental interval increases, it leads to rupture of the transverse ligament and other supporting ligaments.

Basilar invagination

Basilar invagination describes the condition of a relatively cephalad position of the upper cervical vertebra to the base of the skull.

Common signs and symptoms include muscle weakness, neck pain, posterior column dysfunction, bowel and bladder disturbance, and paraesthesia. Basilar impression can also lead to sudden hearing loss, pyramidal tract signs, and wasting of the upper limbs.

Distal form of spinal muscular atrophy

Spinal muscular atrophies (SMA) are clinically heterogenous group of motor system disorders characterized by progressive pure lower motor neuron involvement. ALS, or amyotrophic lateral sclerosis, is a progressive neurodegenerative disease that affects nerve cells in the brain and the spinal cord.

Motor neuron disease

It is a progressive neurological condition that attacks the motor neurons, or nerves, in the brain and spinal cord. This means messages gradually stop reaching the muscles, which leads to weakness and wasting.

Multifocal motor neuropathy

It is a rare neuropathy characterized by progressive, asymmetric muscle weakness and atrophy. Signs and symptoms include weakness in the hands and lower arms; cramping, involuntary contractions or twitching; and atrophy of affected muscles

Multifocal motor neuropathy with conduction block

It is a purely motor neuropathy characterized by motor deficits in the distribution of single nerves without associated sensory loss.

Post-polio syndrome

Post-polio syndrome is a cluster of potentially disabling signs and symptoms that appear decades after the initial polio illness.

Spinal cord tumours

It is also called an intradural tumour, that begins within the spinal cord or the covering of the spinal cord called dura. Some extramedullary tumours affecting spinal cord include- meningiomas, neurofibromas, schwannomas and nerve sheath tumours.

Syringomyelia

It is a generic term referring to a disorder in which a cyst or cavity forms within the spinal cord. This cyst, called a syrinx, can expand and elongate over time, destroying the spinal cord. The damage may result in loss of pain, paralysis, weakness, and stiffness in the back, shoulders, and extremities.

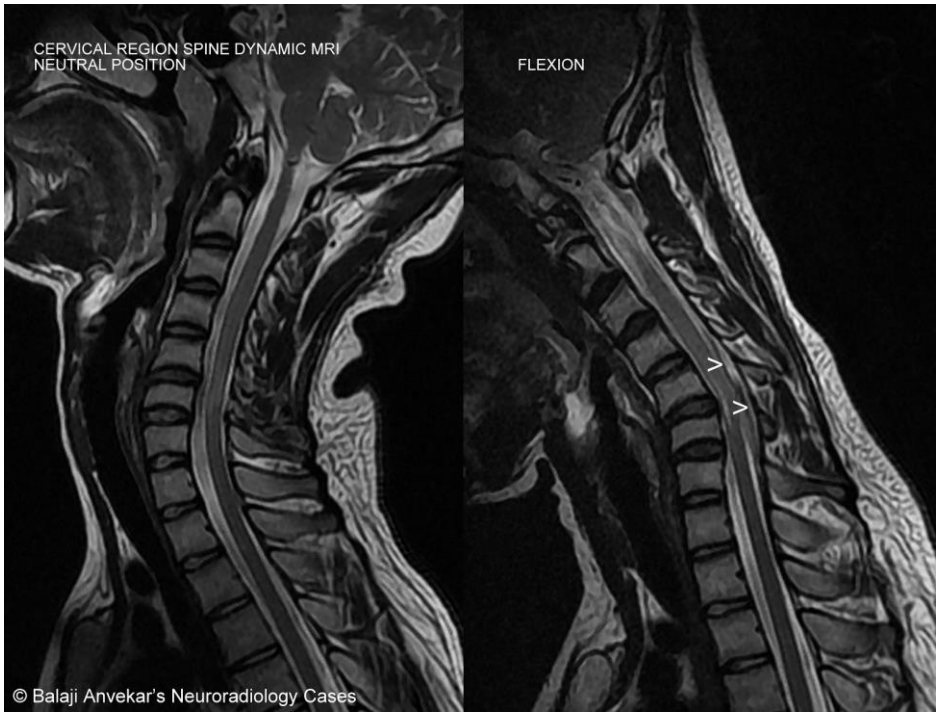
Toxic neuropathy

Toxic neuropathy refers to neuropathy caused by drug ingestion, drug or chemical abuse, or industrial chemical exposure from the workplace or the environment.

Diagnosis

MRI

The typical clinical features and dynamic MRI study when the neck is flexed confirm the diagnosis of Hirayama disease.



Nerve conduction studies (NCS)

These show reduced amplitude of compound muscle action potential (CMAP) in left median and ulnar nerve probably due to severe atrophy of the tested muscle. Sensory NCS was normal.

Electromyography (EMG)

It shows incomplete recruitment pattern with no evidence of positive sharp wave, fasciculation, fibrillation, and no spontaneous insertion activity.

Blood investigations

Among normal findings are-

- Complete blood count
- Sedimentation rate
- Renal, liver, and thyroid function tests
- Creatine kinase
- Vitamin B12 and vitamin D3 level

Negative results are seen for-

Vasculitis screening

- Rheumatoid factor, antinuclear antibody, extractable nuclear antigens, and antiphospholipid antibody

Viral serology

- Human immunodeficiency virus (HIV), hepatitis B, and hepatitis C

Treatment

The treatment of Hirayama disease depends on prevention of repeated cervical spine injury during flexion of neck. A cervical collar which is used to restrict cervical spine to flex or hyperextend, is of much help.

Homoeopathic treatment

The Homoeopathic treatment depends on constitutional symptoms and remedies indicated for acute and chronic nerve injuries. The following are the remedies most indicated in treating Hirayama disease-

abrot. Acon. AESC. agar-ph. agar. All-c. aloe alum-p. alum-sil. Alum. alumn. Am-c. ambr. aml-ns. ammc. Anac. Ang. ant-c. aphis Apis Arg-met. arg-n. ARN. ars-i. ars-s-f. ARS. arum-t. arund. asar. atro. aur-ar. aur-i. aur-s. aur. bapt. bar-m. bell-p. BELL. berb. beryl. Bism. borx. bov. brach. brom. bry. bufo cact. calc-i. calc-p. Calc-s. Calc. Camph. cann-i. cann-s. carb-ac. carb-an. carb-v. carbn-o. Carbn-s. carl. Caust. cere-b. Cham. CHEL. chen-a. Chin. chinin-ar. chinin-s. CIC. cimic. cina clem. coc-c. Cocc. cod. coff. colch. coloc. com. CON. corn. croc. Crot-c. Crot-t. cupr-s. Cupr. Cur. cycl. DIG. dios. dol. Dros. dulc. elaps euph. eupi. ferr-m. ferr. Gels. gins. Glon. Gran. graph. grat. Guaj. ham. hell. helon. hep. hipp. hura hydr-ac. hydroph. hyos. HYPER. Ign. Iod. Ip. jatr-c. kali-ar. kali-bi. KALI-C. kali-i. kali-n. kali-s. Kalm. Lach. lact. laur. Led. lil-t. linu-c. Lyc. lyss. m-ambo. m-aust. macro. mag-c. mag-p. mand. mang. med. meny. Merc-c. merc-i-f. merc-sul. Merc. mez. mim-p. mosch. nat-ar. nat-c. Nat-m. nat-p. NAT-S. Nit-ac. nit-s-d. nux-m. Nux-v. ol-an. Op. paeon. par. peti. Petr. Ph-ac. Phos. phys. phyt. pic-ac. Picro. Plat. PLB. psil. psor. rad-br. rheum Rhod. Rhus-t. Ruta sabad. sabin. samb. santin. sarcol-ac. sars. SEC. SEL. seneg. Sep. Sil. spig. spong. squil. STANN. Staph. stict. Stram. stront-c. STRY. sul-ac. sul-i. Sulph. sumb. symph. syph. tab. tarent. tell. ter. Thuj. til. tub. valer. verat-v. verat. xan. xanth. zinc-m. Zinc.

Short repertory of Hirayama disease

BACK - BENDING - forward - agg. mang. pic-ac.

BACK - BENDING - head - forward - agg. - Cervical region cimic. lyss. rad-br.

BACK - INJURIES - Cervical region ARN. HYPER. mez. rhus-t.

BACK - INJURIES – Spine acon. Apis Arn. bell-p. Calc. caust. cimic. Con. HYPER. ign. Led. NAT-S. Nit-ac. Rhus-t. Ruta Sil. symph. tell. Thuj. zinc.

BACK – INJURIES arg-n. Arn. HYPER. kali-c. nat-s. phys. xanth.

EXTREMITIES - CONVULSION – clonic Ars. atro. bell. brom. carb-ac. carbn-o. coc-c. cocc. cupr-s. cupr. dol. ign. nux-m. op. phos. Picro. Plb. SEC. Stram. STRY. sul-ac.

EXTREMITIES - CONVULSION – Fingers agar. am-c. arn. ars. Bell. bry. Calc. camph. cann-s. cham. CHEL. Cic. cina clem. cocc. coff. Cupr. dros. ferr. hell. Ign. iod. ip. kali-n. lach. lyc. merc-c. mosch. nat-m. nux-v. phos. plb. rheum rhus-t. santin. Sec. spig. stann. staph. sulph. tab. verat.

EXTREMITIES - CONVULSION – Forearms aphis apis chen-a. sec. zinc-m. zinc.

EXTREMITIES - CONVULSION - Hands - taking hold of something ambr. Dros. stry. sulph.

EXTREMITIES - CONVULSION - Hands - writing agg. sil.

EXTREMITIES - CONVULSION – Hands acon. ambr. anac. apis arum-t. bar-m. Bell. bism. calc. Camph. cann-s. carb-v. carbn-s. caust. cic. cina coloc. cupr. dros. graph. Iod. kali-bi. kali-

i. Merc. mosch. nat-m. paeon. plat. plb. rheum Sec. Stram. Stry. sul-ac. tab. verat. zinc-m. Zinc.

EXTREMITIES - CONVULSION - Upper limbs - extending to - Finger acon.

EXTREMITIES - CONVULSION - Upper limbs - extending to - up and down after exertion caust.

EXTREMITIES - CONVULSION - Upper limbs - working hard with hands amel. agar.

EXTREMITIES - CONVULSION - Upper limbs acon. agar-ph. agar. am-c. apis ars. arum-t. BELL. bry. camph. cann-i. carb-ac. caust. Cham. chinin-ar. Cic. cina Cocc. croc. Crot-c. cupr-s. cupr. hydr-ac. hyos. Ign. Iod. Ip. jatr-c. kali-bi. kali-i. linu-c. lyc. lyss. m-ambo. meny. Merc-c. nat-m. nit-ac. Op. phos. Plat. plb. rheum ruta sabad. samb. Sec. Sil. squil. staph. Stram. STRY. sul-ac. sul-i. Sulph. tab. verat-v. verat.

EXTREMITIES - CONVULSION - Wrist, 8-11 h Nat-m.

EXTREMITIES - EMACIATION - Fingers - First lach. thuj.

EXTREMITIES - EMACIATION - Fingers - Tips ars.

EXTREMITIES - EMACIATION - Fingers lach. Sil. thuj.

EXTREMITIES - EMACIATION - Forearms Phos. PLB.

EXTREMITIES - EMACIATION - Hands ars. chin. cupr. graph. mez. ph-ac. Phos. PLB. SEL. sil.

EXTREMITIES - EMACIATION - Shoulders Plb. sumb.

EXTREMITIES - EMACIATION - Thumbs - Balls PLB.

EXTREMITIES - EMACIATION - Thumbs thuj.

EXTREMITIES - EMACIATION - Upper arms - Inner side plb.

EXTREMITIES - EMACIATION - Upper arms Nit-ac. plb.

EXTREMITIES - EMACIATION - Upper limbs ars. carb-an. chin. cupr. graph. Iod. Lyc. PLB. sel. syph. thuj.

EXTREMITIES - EMACIATION - Wrists PLB. sel.

EXTREMITIES - WEAKNESS - Upper arms - Deltoid stann.

EXTREMITIES - WEAKNESS - Upper arms - exertion agg.; after Arg-met.

EXTREMITIES - WEAKNESS - Upper arms - extending to - Hand crot-t.

EXTREMITIES - WEAKNESS - Upper arms - flexion agg. phyt.

EXTREMITIES - WEAKNESS - Upper arms - morning - waking; on arg-met.

EXTREMITIES - WEAKNESS - Upper arms - motion - agg. grat. phyt.

EXTREMITIES - WEAKNESS - Upper arms - motion - amel. arg-met.

EXTREMITIES - WEAKNESS - Upper arms - night - sleep; during sep.

EXTREMITIES - WEAKNESS - Upper arms - paralytic Arg-met. bell. ferr. kali-n.

EXTREMITIES - WEAKNESS - Upper arms - raised, when grat.

EXTREMITIES - WEAKNESS - Upper arms - rising agg. thuj.

EXTREMITIES - WEAKNESS - Upper arms – sudden mang.

EXTREMITIES - WEAKNESS - Upper arms - waking agg.; after arg-met.

EXTREMITIES - WEAKNESS - Upper arms - writing agg. carl. cic. con.

EXTREMITIES - WEAKNESS - Upper arms acon. Arg-met. arg-n. bell. brach. bry. calc. carl. cham. cic. clem. colch. crot-t. cycl. gins. grat. guaj. jatr-c. kali-n. lact. mang. merc-c. mim-p. nat-m. Phos. phyt. sep. sil. stann. Sulph. Thuj.

EXTREMITIES - WEAKNESS - Upper limbs - clenching hands, on chin.

EXTREMITIES - WEAKNESS - Upper limbs - cold, from exposure to Rhod.

EXTREMITIES - WEAKNESS - Upper limbs - convulsions; after CIC.

EXTREMITIES - WEAKNESS - Upper limbs - exertion; after slight Cic. Lach. Stann.

EXTREMITIES - WEAKNESS - Upper limbs – morning carbn-s. dulc. Kali-c. lyc. Nux-v. peti. sil. sulph. valer.

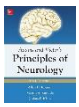
EXTREMITIES - WEAKNESS - Upper limbs - taking hold of something arn. ARS. bov. carb-v. cina colch. nat-m. sil.

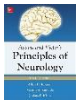
EXTREMITIES - WEAKNESS - Upper limbs - writing agg. acon. agar. brach. carb-v. Caust. cocc. kali-c. lyss. merc-i-f. mez. sabin.

EXTREMITIES - WEAKNESS - Upper limbs abrot. Acon. AESC. agar. All-c. aloe alum-p. alum-sil. Alum. alumn. Am-c. aml-ns. ammc. Anac. Ang. ant-c. Apis arg-met. arn. ars-i. ars-s-f. Ars. arund. asar. aur-ar. aur-i. aur-s. aur. bapt. BELL. berb. beryl. Bism. borx. bov. brach. brom. bry. bufo cact. calc-i. calc-p. Calc-s. Calc. camph. carb-v. Carbn-s. Caust. cere-b. cham. Chel. Chin. chinin-ar. chinin-s. CIC. cod. coff. coloc. com. CON. corn. Crot-c. Crot-t. Cupr. Cur. DIG. dios. dros. dulc. elaps euph. eupi. ferr-m. ferr. Gels. gins. Glon. Gran. graph. grat. Guaj. ham. hell. hep. hipp. hura hydroph. hyper. ign. Iod. kali-ar. kali-bi. KALI-C. kali-i. kali-n. kali-s. Kalm. Lach. lact. laur. led. lil-t. Lyc. lyss. m-aust. macro. mag-c. mag-p. mand. mang. med. Merc-c. merc-i-f. merc-sul. merc. mez. nat-ar. nat-c. Nat-m. nat-p. nat-s. nit-ac. nit-s-d. Nux-v. ol-an. op. par. peti. Petr. Ph-ac. Phos. phyt. plat. plb. psil. psor. Rhod. Rhus-t. ruta sabad. sabin. sarcol-ac. sars. sec. seneg. Sep. Sil. spig. spong. STANN. Staph. stict. stram. stront-c. sul-ac. Sulph. sumb. tab. tarent. ter. Thuj. til. tub. valer. verat. zinc.

GENERALS - INJURIES – Nerves all-c. arn. bell-p. helon. hyper. led. meny. ph-ac. phos. xan.

Bibliography


Chapter 39. Degenerative Diseases of the Nervous System > Diagnosis of ALS Adams & Victor's Principles of Neurology, 10e... The early clinical picture of motor system disease is closely simulated by a centrally placed cervical spondylotic bar or ruptured cervical disc, but with these conditions there is usually pain in the neck and shoulders, limitation of neck movements, and sensory changes, and the lower motor neuron...


Chapter 44. Diseases of the Spinal Cord > Cervical Dural Sac Myelopathy (Hirayama Disease) Adams & Victor's Principles of Neurology, 10e... This unusual myelopathy has usually been considered in discussions of the motor neuron disorders because of its characteristic features of chronic wasting of one or both hands and forearms without sensory changes or long tract signs. It appears, however, that the damage in this disease is from...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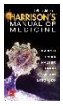
Peripheral Neuropathies, Including Guillain-Barré Syndrome > INTRODUCTION Harrison's Manual of Medicine, 19e... sensory loss With upper motor neuron findings Consider: motor neuron disease Without upper motor neuron findings Consider: progressive muscular atrophy, juvenile monomelic amyotrophy (Hirayama’s disease), multifocal motor neuropathy, multifocal acquired motor axonopathy...


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