

# DISEASES OF SPINAL CORD

THE MOST COMMONLY OBSERVED AND IMPORTANT DISORDERS OF SPINAL CORD CAN BE GROUPED IN TO FOLLOWING CLINICAL SYNDROMES:

1. PARAPLEGIA OR QUADRIPLEGIA, WITH SENSORY LOSS, DUE TO COMPLETE TRANSVERSE LESIONS OF SPINAL CORD
2. THE SYNDROME OF SUBACUTE OR CHRONIC SPINAL PARAPARESIS, WITH OR WITHOUT SENSORY CHANGES AND ATAXIA
3. THE SYNDROME OF SEGMENTAL SENSORY DISSOCIATION WITH BRACHIAL AMYOTROPHY (SYRINGOMYELIC SYNDROME)
4. VENTRAL CORD SYNDROME
5. CENTRAL CORD SYNDROME
6. HEMICORD SYNDROME (Brown-Séquard)
7. SYNDROME OF CONUS MEDULLARIS AND CAUDA EQUINA

## PARAPLEGIA OR QUADRIPLEGIA DUE TO COMPLETE TRANSVERSE LESIONS OF SPINAL CORD

-THIS SYNDROME IS BEST CONSIDERED IN RELATION TO TRAUMA WHICH IS THE MOST FREQUENT CAUSE, BUT IT OCCURS ALSO AS RESULT OF INFARCTION, HEMORRHAGE AND WITH RAPIDLY ADVANCING COMPRESSIVE, DEMYELINATIVE OR INFLAMMATORY LESIONS

-CLINICAL PICTURE – PARAPLEGIA OR QUADRIPLEGIA WITH SPHINCTERIC PARALYSES AND SENSORY LOSS BELLOW THE LEVEL OF LESION

## Spinal cord trauma

-PENETRATION OF SPINAL CANAL BY MISSILE IS THE COMMON CAUSE IN WARTIME

-VERTICAL COMPRESSION OF SPINAL COLUMN TO WHICH IS ADDED THE IMMEDIATE EFFECT OF ANTERO OR RETROHYPERFLEXION IS THE USUAL MECHANISM IN CIVILIAN LIFE

The resultant tearing of spinal ligaments permits the dislocation of an upper vertebra anteriorly on the one bellow, often with fracture of vertebral body or pedicles/. The posterior part of fractured body is displaced backward and compresses the cord. Less severe degrees of anteroflexion injury produce only dislocation.

Vulnerability to the effects of anteroo and retrohyperflexion injuries increased by the presence of cervical spondylosis, or congenitally stenosis of the spinal canal

The most vertebral injuries occurred at the levels of C1, C2, C4-C6, Th1, L2

**-WHIPLASH OR RECOIL INJURY (car accident)- flexion/retroflexion; Occipitounchal muscles and other supportive structure of the neck and head are affected much more than spinal cord or roots.**

**Pathology of spinal cord injury- As a result of squeezing of spinal cord there is destruction of gray and white matter and a variable amount of hemorrhage. These changes are maximal at the level of injury and one or two segments above and bellow it. This is traumatic necrosis of the spinal cord**

### **THE SUBSEQUENT EFFECTS OF ACUTE TRANSVERSAL LESION:**

**WHEN THE SPINAL CORD IS SUDDENLY AND COMPLETELY SEVERED IT IS EVIDENT THAT:**

**-THE STAGE OF SPINAL SHOCK-**

- 1.ALL VOLUNTARY MOVEMENT IN PARTS OF THE BODY BELLOW THE LESIONS IMMEDIATELY AND PERMANENTLY LOST**
- 2.ALL SENSATION FROM THE LOWER PARTS IS ABOLISHED**
- 3. REFLEX FUNCTION IN ALL SEGMENTS IN ISOLATED SPINAL CORD IS SUSPENDED. THE LAST EFFECT, CALLED SPINAL SHOCK INVOLVES TENDON AS WELL AS AUTONOMIC REFLEXES / ATONIC BLADDER, ATONIC BOWEL (PARALYTIC ILEUS), GASTRIC DILATATION, LOSS OF GENITAL REFLEXES AND VASOMOTOR CONTROL/. IT LASTS FOR WEEKS TO MONTHS AND IT IS SO DRAMATIC.**

**-THE STAGE OF HEIGHTENED REFLEX ACTIVITY**

**USUALLY, AFTER A FEW WEEKS, GRADUALLY THE REFLEX RESPONSE BECOME HYPERACTIVE AND THE BLADDER BECOMES SPASTIC. ALSO AUTONOMIC FUNCTIONS / VASOMOTOR AND SWEATING REACTIONS/ BECOME HYPERACTIVE.VARYING DEGREES OF HIGHTENED FLEXOR REFLEX ACTIVITY MAY LASTS FOR A YEARS.**

**“MASS REFLEX”**

**After several months the withdrawal reflexes become greatly exaggerated to the point of flexor spasms and my be accompanied by profuse sweating, piloerection, automatic emptying bladder or rectum. It may be evoked by stimulation of the skin or interoceptive stimulus and this is the “mass reflex”**

### **CLINICAL EFFECTS:**

**THE IMMEDIATE EFFECT OF TRANSVERSAL LESION DEPENDS ON ITS LEVEL**

**-C1-C3-RESPIRATORY PARALYSIS AND DEATH**

**-C4-C5- COMPLETE PARALYSIS OF ARMS AND LEGS**

**-C5-C7-PARALYSIS OF THE LEGS, AND ARMS CAN STILL BE ABDUCTED AND FLEXED**

**-LESIONS OF THORACIC CORD-PARAPLEGIA**

## **-BELLOW L1-CAUDA EQUINA SYNDROMA**

**THE LOSS OF MOTOR FUNCTION AT THE TIME OF INJURY IS ACCOMPANIED WITH SUPPRESSION OF REFLEX ACTIVITY BELLOW THE LESION, LOSS OF SENSATION BELLOW THE LEVEL CORRESPONDING TO SPINAL CORD LESIONS, AND ATONIC BLADDER AND BOWEL, GASTRIC ATONY.**

**-ACUTE CENTRAL SPINAL CORD SYNDROME-** It is the most prevalent of the partial cord syndromes. It is associated with cervical trauma (retroflexion injuries of the head and neck, infarction due to compression of the vertebral artery at medullary –cervical region), there is disproportionate damage to the central gray matter of the cord. The destruction of gray matter (motor and sensory neurons) may leave an atrophic, areflexic paralysis and a segmental analgesia and thermhypoesthesia, clinically simulating syringomyelic syndrome. The acute central cord syndrome affects motor function in the upper limbs more than in the lower ones. The paresis is often denser distally in extremities than proximally.

**-THE GREATEST RISK TO PATIENTS WITH SPINAL CORD INJURY IS THE FIRST WEEK TO 10 DAYS**

**-ANY RESIDUAL SYMPTOMS PERSISTING AFTER 6 MONTHS ARE LIKELY TO BE PERMANENT**

### **TREATMENT**

**-ORTHOPEDIC- IF EXIST SPINE FRACTURE AND DISLOCATION**

**-IMMEDIATE ADMINISTRATION OF HIGH DOSES OF CORTICOSTEROIDS (1000MG LEMOD SOLU IV –V DAYS)**

**-IN PATIENTS WITH COMPLETE SPINAL CORD LESIONS THE PREVAILING OPINION IS AGAINST LAMINECTOMY**

**-SYMPTOMATIC TREATMENT-MANAGEMENT OF BLADDER AND BOWEL DISTURBANCES, CARE OF SKIN, TREATMENT OF SPASTICITY AND FLEXOR SPASMS, PHYSIOTHERAPY**

## **NONTRAUMATIC TRANSVERSE MYELOPATHIES**

**An acute complete or nearly incomplete transverse cord lesion in the absence of trauma should lead to a consideration on the following:**

- TUMOR WITH CORD COMPRESSION**
- HEMORRHAGE INTO THE SPINAL CORD (Hematomyelia) AVM OR EPIDURAL OR SUBDURAL HEMORRHAGE OR VENOUS COMPRESSION OF THE LOWER CORD BY DURAL FISTULA OR AVM**
- ACUTE INFLAMMATORY NECROTIZING OR DEMYELINATIVE MYELOPATHY**
- EPIDURAL ABSCESS**

- **ISCHEMIC INFARCTION OF THE CORD**-due to occlusion in major segmental artery arising from vertebral artery (supplying the cervical cord) or the aorta (supplying the thoracic and lumbar cord). Dissection aortic aneurysm, arteritis, atherosclerosis of the collateral arterial vessels are the usual causes.

## **THE SYNDROME OF SUBACUTE OR CHRONIC SPINAL PARAPARESIS, WITH OR WITHOUT SENSORY CHANGES AND ATAXIA**

### **HEREDITARY SPASTIC PARAPARESIS (HSP)**

- Heterogeneous group of conditions with “pure” and “complicated” forms
- The pathologic involvement of lateral corticospinal tracts may present clinically as an upper motor neuron disorder
- In all forms of HSP beside degeneration of corticospinal tracts there is degeneration of posterior columns of the spinal cord
- The most common inheritance mode is autosomal dominant (AD) , approximately in 70-85% of HSP patients
- AD HSP is classified as type I or type II based on age of onset before and after age 35 years; in the more common type I onset of symptoms usually occurs in childhood or young adulthood; the clinical characteristics of type I and II are similar
- The key clinical features are progressive lower extremity spasticity and weakness, but gradually the upper extremities become involved too; pes cavus is seen in more than 50%; late features include loss of vibratory sensation in the legs and urinary urgency
- Autosomal recessive (AR) HSP often has an earlier age of onset and more rapid progression
- Rare forms include X-linked HSP (complicated HSP), which manifests more extensive CNS involvement.
- Linkage analyses has been described in autosomal dominant and recessive and X-linked forms, with evidence of genetic heterogeneity
- Mutations have been identified in four genes and molecular diagnosis is possible for mutations in these known genes
- In autosomal dominant HSP mutations of spastin gene have been found /SPG4/ with gene location in the chromosome 2p21-24; mutations accounted for around 40-50% of all AD HSP families
- Autosomal recessive form of HSP, is due to mutations in the paraplegin gene /SPG7/ with gene location in the chromosome 16q24.3
- X linked HSP is due to mutations of myelin protelipid protein (L1CAM) in the chromosome Xq22

**Treatment-** at present for HSP is limited to symptomatic care to reduce spasticity (walkers, wheelchairs, antispasticity drugs)

## **CERVICAL SPONDYLOSIS WITH MYELOPATHY**

**-Most frequently observed myelopathy**

**-Degenerative disease of middle and lower vertebrae in combination of degenerating and bulging disc(s), vertebral exostosis, thickening of the posterior longitudinal and yellow ligaments, often accompanied by congenitally narrowing of spinal canal compromise the cervical cord and roots by compression and possibly by reduction of the blood supply**

**-Most frequently we observed intervertebral disk protrusion (IDP), with prolapsus nucleus pulposus through the perforated fibrous ring; the IDP is usually in posterolateral direction, causes isolated root compression; central disc protrusion can cause spinal cord compression with spastic paraparesis, posterior column syndrome and bladder dysfunction**

**Clinically the syndromes consists:**

- 1) Painful, stiff neck with restricted movements**
- 2) radicular pain and numbness and reduced reflexes in arm**
- 3) symmetric or asymmetric spastic paraparesis and ataxia**
- 4) the most frequent locations are the C6/C7 and C5/C6 levels**

**The condition is often chronic and MRI, or CT myelography, EMG are necessary for diagnosing.**

**Treatment**

- 1) Conservative treatment**

- rest and relief of strain of the cervical vertebral column using a neck collar for period of 2-3 weeks**
- careful manual or mechanical traction to extend the intervertebral space**
- Cryotherapy of PVM**
- Drugs (muscle relaxants, anti-inflammatory analgetics)**

- 2) Surgical decompression-two techniques are commonly recommended-ventral discectomy and dorsolateral approach with foramenotomy**

**Criteria for surgical treatment:**

- Acute central IDP with symptoms of spinal cord compression**
- Lateral IDP with functionally significant radicular paresis without improvement over period of 3 weeks**
- Severe radicular pain syndrome unresponsive to treatment**

## **RADIATION MYELOPATHY**

Iatrogenic disease, as sequela of radiation therapy for tumors in these regions  
Pathologically there is coagulation necrosis of both gray and white matter extending overall segments of the cord and corresponding with level of the irradiation zone

### **Transient myelopathy**

Early type, appearing 3-6months after radiotherapy-characterised by parasthesias in extremities  
The sensory abnormalities disappear after a few months

### **Delayed progressive radiation myelopathy**

Most common complication, first appear after 6 months, usually between 12 and 15 months, occasionally after several years  
Clinically the onset is insidious, with sensory symptoms in extremities weakness of one or both legs;  
The neurological disturbance may take the form of Brown-Sequard syndrome, but later the syndrome is that of a transverse myelopathy with spastic paraplegia, sensory level on the trunk and sphincter disturbance  
The CSF is usually normal

This complication can be avoided if the total dose of radiation kept 6000 rads and if it is given over a period of 30-70 days (each daily fraction does not exceed 200 rads)

Treatment-Symptomatic- a number of case reports remark temporary improvement in neurological function after the administration of corticosteroids

## **MYELITIS**

1. Myelitis due to filterable viruses
  - Polyomyelitis, Coxsackie virus, echovirus
  - Herpes zoster
  - Rabies
  - HTLV 1,HTLV 3
  
2. Myelitis secondary to bacterial, fungal and parasitic disease of the meningeas and spinal cord
  - A/ Syphilitic myelitis
  - B/ Pyogenic suppurative myelitis
    - Subacute meningomyelitis
    - Acute epidural abscess and granuloma
    - Abscess of spinal cord
  
  - C/ Tuberculous myelitis

- Pott's disease
- TBC meningomyelitis
- Tuberculoma of spinal cord

**D/ Parasitic or fungal infections producing epidural granuloma, meningitis, meningomyelitis and abscess**

**E/ Myelitis unknown etiology**

- Postinfectious and postvaccinal
- Acute and chronic relapsing MS
- Necrotic or degenerative

### **CLINICAL CHARACTERISTICS:**

- Acute (develop rapidly, reach the peak of severity within days)
- Subacute (evolves over period of 2 to 6 weeks)
- Chronic (evolves over period more than 6 weeks)

### **ACUTE POLYOMIELITIS**

Prototype of disorder with acute LMN dysfunction. The disease is caused by poliovirus, belonging to Enterovirus and family of Picovirus. Immunization has eradicated the disease. The viruses of polio have affinity for neurons of anterior horn.

Only small proportion of people who are exposed in endemic regions develop either minor illness (a gastrointestinal influenza like illness) or the major illness. Clinically the major illness resembles aseptic meningitis and 50% of patient's progress to paralytic disease within 2-5 days. This phase characterized localized fasciculation severe myalgia and focal asymmetric paralysis. Any muscle can be affected, but leg, arm respiratory and bulbar muscles are most frequently affected. Most patients eventually make significant improvement (80% by 6 months)

**Diagnosis-** The CSF- pleocytosis with polymorphonuclear (acute stage), and lymphocytes (later). The CSF protein show mild to moderate increase. The CSF poliovirus specific immunoglobulin M antibody test enables to accurate immunological diagnosis. Stool cultures are positive in nearly 90%, by the tenth day of

**Treatment-** consists mainly of general supportive care, with close monitoring of respiratory function

**Vaccination-** The trivalent oral polio vaccine (TOPV)

### **PROGRESSIVE POSTPOLIOMYELITIS MUSCULAR ATROPHY (PPMA)**

Many years after recovery from poliomyelitis some patient's experiences progressive weakness called PPMA. The etiology has not yet been established Persistent infection with defective poliovirus is possible as well as persistent immune-mediated mechanism

## **Characteristic features of PPMA**

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### **Medical history**

- recovery for acute poliomyelitis
- A long stable course, at least 10 years

### **Signs and symptoms**

- progressive weakness, usually in previously affected muscles
- muscle pain and arthralgia

### **Laboratory studies**

- EMG identify evidence of previous polio infections
- No test is specific for PPMA

### **Diagnosis**

- Exclusion of other treatable disease

### **Treatment**

- symptomatic and supportive care
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## **HERPES ZOSTER**

It is caused by varicella zoster virus and probably represents reactivation varicella virus infection that has been latent in sensory ganglia following the primary infection with chickenpox. Pathologically there is an intense inflammation in two or three dorsal root or cranial nerve ganglia and in corresponding posterior and anterior roots, adjacent meninges and gray matter of the spinal cord on the one side. Myelitis is rare complication. The CSF contains 10-100 cells, mainly lymphocytes and a slightly increased protein. Clinical features- radicular pain, vesicular cutaneous eruption involving one or more dermatomes of one side of the body, and in some cases sensory and motor deficits in the segments bearing skin lesions. Any part of the body may be affected, but thoracic lesions are the most frequent. Involvement of multiples dermatomes should always suggest an underlying immunocompromised condition.

Treatment- Acyclovir (800mg five times daily for 7 days), analgetics.

## **HUMAN T-CELL LEUKEMIA VIRUS TYPE I (HTLV1)-TROPICAL SPASTIC PARAPARESIS**

HTLV1 causes chronic progressive myelopathy/meningomyelitis involving predominantly thoracic spinal cord/, in the Caribbean, South Africa Central and South America, and Japanese population.

This retrovirus produced insidiously spastic paraparesis, with paraesthesia and pain in the legs, and bladder dysfunction.

Usually these symptoms begin after 30 years. Routes of HTLV1 transmission are breast milk, sexual intercourse and exposure to contaminated blood products.

The definitive diagnosis of HAM requires HTLV-1 positive serology in blood and CSF.

Treatment- At present no antiviral agents effectively treat TSP

## **HIV ASSOCIATED MYELOPATHIES**

Spinal cord is frequent in HIV infection. A unique HIV associated vacuolar myelopathy is the chief cause of myelopathy This type of degeneration is seen at autopsy in over 20% of AIDS patients. Neurological examinations reveals spastic

paraparesis, gait ataxia, sensory and bladder dysfunctions. HIV-related vacuolar myelopathy is seldom associated with abnormalities on MRI.

A number of other myelopathies may also occur with HIV infection, including infectious myelopathies (cytomegalovirus, herpes simplex –2, HTLV1, Treponema pallidum, and epidural abscesses), vascular myelopathies, epidural and intramedullary tumors and a demyelinating myelopathy

## **Myelitis secondary to bacterial, fungal and parasitic disease of the meninges and spinal cord**

### **Syphilitic myelitis**

- Chronic meningoradiculitis (Tabes dorsalis)
- Chronic meningomyelitis
- Meningovascular syphilis
- Gummatous meningitis including chronic spinal patchy meningitis

Treponema pallidum is recognized cause of a wide range of neurological syndromes. Syphilis may be divided into several clinical stages. The primary syphilis is characterized by primary skin lesions (10 days to 10 weeks). In the secondary stage haematogenous dissemination of T. pallidum may occur. During this stage lymphocytic meningitis may occur. Even without of antibiotic therapy, the clinical signs resolve spontaneously and leaving patient in a latent stage of infection. Tertiary may develop in about one-third of untreated patients. This include gummatous syphilis, cardiovascular and various forms of tertiary neurosyphilis. Early phases of this type of disease include meningeal and meningovascular forms and parietic and tabes dorsalis may develop in later phases.

Tabes dorsalis- Disease presents after 15-20 years of primary infection.

-The clinical hallmark is the symptom of lightning pains, often in the legs, autonomic dysfunction and sensory ataxia. In addition the classical triad of signs includes Argyl-Roberson pupils, areflexia and loss of the proprioceptive sense. Visceral crises sometimes leading to misdiagnosis of acute abdomen. In 5-10% CSF is normal (so-called burned out tabes). If it is abnormal we noticed pleocytosis, increased protein, especially gamma globulin, presence of antibodies.

-Pathologic study reveals dorsal ganglion loss, degeneration of the dorsal funiculi of the sc secondary to lumbosacral radiculitis

-Diagnosis is based of history of primary and secondary syphilis, clinical characteristic, laboratory testing for reagin and treponemal antibodies (VDRL).

Treatment-Penicillin G, 18-24 million units IV 14 days, or erythromycin and tetracycline in penicillin sensitive patients 0.5g every 6 hours 20-30 days.

## **Pyogenic or suppurative myelitis**

The inflammatory reaction of spinal meninges is only one manifestation of generalized disease process. The spinal lesions may be involve primary pia-arachnoid (leptomeningitis), dura (pachimeningitis) or the epidural space (abscess or granuloma); in the later events damage to the spinal cord is due to compression and ischemia

**Spinal epidural abscess-** An injury to the back, often trivial, or other wound infection or bacteriemia, may permit seeding of the spinal epidural space or of vertebral body. Staphylococcus aureus is the most frequent etiologic agent. At first suppurative process is accompanied only by fever and pain in the back, followed several several days by radicular pain. Headache and nuchal rigidity are frequently present. After that period of time there is onset of rapidly progressive paraparesis to paraplegia associated with sensory loss in lower part of the body, sphincter paralysis. Percussion of the spine elicits tenderness over the site of infection.

**Diagnosis-** The CSF contains a small number of white cells, the protein content is high but glucose is normal, there is a dynamic block. MRI of thoracic segments or thoracolumbal segments or spinal cord are necessary. The other diagnostic way is myelography.

**Treatment-** If not treated surgically by laminectomy and drainage at earliest possible moment, the spinal cord lesions which is due in part to ischemia become more or less irreversible. Antibiotic therapy may also given.

**Subacute pyogenic infections and granulomatous infections (TBC, fungal)** The clinical picture is less dramatic and local and radicular pain is absent. The clinical and spinal fluid findings call for MRI or myelography

Treatment depends on the nature of underlying disease and general condition of the patient

**Subdural bacterial infections** also occur and are virtually indistinguishable from epidural ones. A clue may be provided by MRI, contrast myelography.

**Spinal cord abscess** is very rare. The symptoms are indistinguishable from those of epidural abscess. In some instances the patients was known to have had a systemic bacterial infections, septicemia or endocarditis.

## **TUBERCLOUS MYELITIS**

Solitary tuberculoma of the spinal cord is very rare. Tuberculous osteitis of the spine with kyphosis (Pott's disease) is more frequent and in those patients granulation tissue may extrude from infected vertebra and gives rise to an epidural abscess that compress the cord (Pott's paraplegia). The diagnostic procedures as well as treatment is the same as in tuberculous meningitis

## **Myelitis unknown etyology**

These disorders take the form of leukomyelitis based either on demyelination or necrosis of the tracts of spinal cord

Postvaccinal myelitis as a form of acute disseminated encephalomyelitis (ADEM)

The common precedents are viral infections (EBV, CMV, measles, rubella, chickenpox), certain vaccinations

The autoimmune reactions is postulated

Spinal cases are with clinical picture of transversal myelitis ; the neurological symptoms progress for several days after which remain stationary and then recede slowly. In most cases there are other neurological signs pointing lesions of cerebellum, brainstem, optic nerve. in cerebral cases , death may occur within days.

CSF- pleocytosis (lymphocytes and other mononuclear cells), normal or slightly raised proteins, normal glucose value. There is always uncertainty whether the illness is the opening phase of multiple sclerosis

### **DEMYELINATIVE MYELITIS**

One third of patients with multiple sclerosis, including older adults exhibit spinal form of the disease /symmetric or asymmetric quadriparesis or paraparesis with hyperreflexia and sensory ataxia/. This form share many of the properties of postinfectious type, except that this myelitis tend to evolve slowly over period of 1-3 weeks or longer

### **ACUTE NECROTIZING MYELITIS**

Rare disorder that present with at acute onset of paraplegia or quadriplegia , sensory loss and sphincter paralysis. The neurological signs may erupt in hours (myelomalacia or hematomyelia). A few or several hundred mononuclear cells and increased protein may be found in CSF. Autopsy of these patents showed necrotizing hemorrhage leucomyelitis. The combination of spinal cord necrosis and optic neuritis appears to correspond with Devic syndrome.

### **SUBACUTE NECROTIZING MYELITIS**

/Paraneoplastic myelitis/

Subacute progressive paraparesis, bladder and anal sphincter disturbances sensory deficits but no pain are typical clinical presenting symptoms. If symptoms ascending, respiratory weakness may result. The CSF shows mononuclear pleocytosis and occasional red blood cells with moderate elevation of protein. MRI will show an increase signal on T2 images. The pathology find the underlying necrosis of myelon at thoracic levels. This rare paraneoplastic syndrome has been found with bronchial carcinoma, Hodgkin's lymphoma, breast carcinoma. There is no therapy.

## **VASCULAR DISEASE OF THE SPINAL CORD**

The spinal cord is uncommon site of vascular disease. The spinal arteries are not susceptible to arteriosclerosis . Very rarely spinal cord is affected in lupus erythematosus.

### **INFARCTION OF THE SPINAL CORD (MYELOMALACIA)**

Cervical part of the spinal cord is the best-protected area from disorders pertaining to spinal circulation.

This is also the case with posterior areas of most areas of the spinal cord.

The ventral region of midthoracic, thoracolumbal and lumbosacral areas is most vulnerable. One of the main reasons is that the anterior longitudinal trunk is formed by only few ventral radiculospinal arteries, quite often almost the entire thoracolumbar part of the spinal cord is supplied by one single (Adamkiewitz) artery.

Arterial hypotension most frequently lead to spinal cord ischemia (hemorrhage, ischemic heart disorders, cardiac arrest) In these instances spinal infarcts may appear on the border between cervicothoracic and thoracolumbar regions (at the level of T4segment), as well as in the transverse sense-in the central part of the spinal cord.

Certain diseases, and iatrogenic damage to the aorta, which give off most of the segmental arteries, may result in the ischemia of the thoracolumbar part of the spinal cord.

Disecting aneurysm of the aorta rise to number neurological syndromes:

- Paralysis of the sphincters and both legs with sensory loss below T6
- Obstruction of brachial artery with sensorimotor neuropathy
- obstruction of a common carotid artery with hemiplegia

#### **Syndrome of anterior longitudinal trunk (Anterior spinal artery syndrome)**

Ischemia usually affects the ventral two thirds of the spinal cord. There are spastic paraparesis below the lesion, flaccid paralysis at lesion level, loss of pain and temperature sensation below the lesion (deep sensation spared) and bladder and bowel paralysis.

#### **Syndrome of posterior longitudinal trunk**

Occlusion of the posterior longitudinal trunks is very rare. It has been observed following intrathecal injection of certain medicaments or radiopaque. In these patients ischemic lesions affect dorsal white columns, dorsal horns and dorsolateral funiculus.

As a consequence, dyaesthesia and pain appear first, which are followed by loss of almost all modality of sense, dysfunctions of sphincters and spastic paraparesis. The patients recover satisfactorily.

Treatment of all forms of spinal cord infarction can only be symptomatic. After 10-14 days more active rehabilitation can be started.

#### **HEMORRHAGE INTO THE SPINAL CORD (HEMATOMYELIA)**

Hemorrhage into the spinal cord is also rare. The apoplectic onset of onset of symptoms that involve both motor and sensory tracts associated with blood and xantochromia in spinal fluid are the features of hematomyelia.

Etiology-vascular malformations, administration of anticoagulants

Diagnostic procedures- selective spinal angiography make it possible to distinguish between the several types of vascular malformations and hemangioblastomas and to localize them to the spinal cord, epidural, or subdural space or the vertebral body.

#### **SYRINGOMYELIA AND SYRINGOBULBIA**

Syringomyelia as a term used to describe a disease characterized by eccentric fluid cavities within spinal cord. The term is derived from syrinx a name given to

a hollow fluid-filled cavity. Syringomyelic cavity usually include central gray matter but extend into the anterior or posterior horns and often affected root entry zone white matter on that side. There are different mechanisms for development of this cavity. Syringomyelic cavity usually incorporates the central canal. Syrinxes are most common in cervical and upper thoracic spinal cord and often extend rostrally up to the second cervical segment. Less common is extension caudal into caudal thoracic and lumbar spinal cord.

**Classification (Barnett et al):**

**Type 1-Syringomyelia with obstruction of foramen magnum and dilatation of the central canal**

**A/ With type of I Chiari malformation**

**B/ With other obstructive lesions of the foramen magnum**

**Type 2- Syringomyelia without obstruction of foramen magnum (idiopathic type)**

**Type III-Syringomyelia with other disorders of spinal cord**

**A/ Neoplastic- intramedullary tumors, especially gliomas and ependymomas, may affect the same region of the cord as syrinxes. MRI can now probably differentiate neoplastic from developmental cavity**

**B/ Traumatic myelopathy**

**C/ Spinal arachnoiditis and pachymeningitis**

**Type IV-Pure hydromyelia with or without hydrocephalus**

**Clinical features varies depends not only on the extent of syrinx, but also on the associated pathological changes (Chiari malformation). Symptoms usually begin in the adult age period (35 to 45 years)**

**The onset is insidious and the course is irregularly progressive**

**The cardinal features are segmental weakness and atrophy of the hands and arms with loss of tendon reflexes and segmental anesthesia of the dissociated type (loss of pain and thermal sensation and preservation of the sense of touch) over the neck, shoulders and arms. Finally there is weakness and ataxia of the legs from involvement of the corticospinal tracts and posterior columns in cervical cord. Pain is a symptom in about half of our patients with types I and II. The pain is unilateral, burning aching quality, mostly on the border of areas of sensory impairment.**

**SYRINGOBULBIA- is the bulbar equivalent of syringomyelia. Usually the two coexist. Cavity is located most often in the lateral tegmentum of the tegmentum of the medulla, but it may extend into the pons.**

**Clinical features-the symptoms are usually unilateral- nystagmus, analgesia thermoanaesthesia of the face, wasting and weakness of the tongue palatal and cord paralysis (dysarthria, dysphagia, hoarseness)**

**When Chiari malformation is associated with syringomyelia clinical features are nystagmus, cerebellar ataxia, external neck pain, hydrocephalus, prominent corticospinal and sensory tract involvement craniocervical malformations.**

**Treatment- It is important to differentiate between cavitations that communicate with the fourth ventricle and subarachnoid space and those that not! MRI is**

necessary for every case. Cine MRI with presaturation bolus tracking may be helpful in analyzing CSF dynamics.

Therapy attempts to at least stop the process by surgical decompression of the sc and shunting the content of syrinx either into subarachnoidal space at the peritoneum. The second type of intervention seems to be superior.

The choice of the best neurosurgical procedure depends on the anatomic situation around the craniocervical junction and the brain stem as well as the type of spinal cavitations.

Rarely the decompression of the foramen magnum is only the first step of the therapy.

When the ventricles are enlarged a ventriculoatrial shunt may be necessary. In addition symptomatic treatment have to been taken in consideration.

## **SPINAL ABNORMALITIES WITH MYELOPATHY**

**Anomalies of Craniocervical Junction-** Congenital fusion of the atlas and foramen magnum are the most common. Whenever the anteroposterior diameter of the canal behind the odontoid process was less than 19.0 mm, there were signs of spinal cord compression

**Platybasia and basilar invagination-** Refers to a flattening of the base of the skull (the angle formed by intersection of the plane of the clivus and the plane of the anterior fossa is greater than 135).

**Basilar impression or invagination** means upward bulging of the margins of the foramen magnum; if the occipital condyles, which bear the thrust of the spine, are displaced above the plane of the foramen magnum, basilar invagination is present. Each of these abnormalities (congenitally or acquired) give rise to characteristic shortness of the neck and combination of spinal and cerebellar signs

**Abnormalities of odontoid process – Atlantoaxial dislocation-** atlas displaced anteriorly in relation to the axis cause acute or chronic spinal cord compression depends on the type of abnormalities- congenital or the result of injury. Rheumatoid arthritis is another cause

**Lumbar spinal canal stenosis** occurs in middle or old age, either with straightforward radicular symptoms or in the form of cauda equina syndrome. This occurs especially when the patient stands or walks, because of increased lordotic compression – intermittent claudication of cauda equina; Symptoms are: pain in the buttocks and legs numbness and weakness of the legs. Symptoms quickly relieve after rest in the ventral position. Stress-dependent relative ischemia of the cauda equina is possible additional factor. The final diagnosis is based on radiological documentation using CT or water-soluble myelography; MRI imaging visualizes the soft tissue abnormalities Treatment- In less severe cases-conservative; Patients with severe neurological loss or with resistant pain syndrome usually undergo decompressive laminectomy overall several lumbar segments

## **SUBACUTE COMBINED DEGENERATION /VITAMIN B12 DEFICIENCY/**

Long standing deficiency of vitamin B 12 (cobalamin) has two major effects: 1) a pernicious anemia and 2) a degeneration of the posterior and lateral columns of spinal cord (and sometimes brain, peripheral nerves, and optic nerve) which may occur independently and precede the hematological effects. The neurological disease traced to a failure of cobalamin dependent enzyme-methyl-malonyl-CoA mutase, which is essential for the maintenance of myelinated fibres.

**Clinical features**-The patients first notice general weakness and paraesthesias / they are localized in distal parts of all four limbs. As the illness progresses, the gait becomes unsteady. If the disease remains untreated an ataxic paraplegia with variable degrees of spasticity may develop. At first the tendon reflexes are usually absent, but with progression of disease become exaggerated, the plantar response becomes extensor. Loss of vibration as well as position sense is the most consistent sign. The signs are usually limited to the legs.

Demyelination followed by axonal degeneration seems to affect the most heavily myelinated fibres first which may explain why lesions appear first in the posterior columns and later in lateral columns. They tend to appear in midthoracic levels. The defect of cutaneous sensation /impaired tactile sensation, pain, thermal perception over the limbs, in distal distribution/ implicating peripheral neuropathy.

The pathological changes in the brain are similar to those in SC. Changes appear most marked in the corpus callosum, and the frontal and parietal white matter. A variety of symptoms occur, consisting of disorders of mood, agitation, confusion, incontinence. Optic neuropathy due to vitamin B12 deficiency is rare. It is of interest that neuropsychiatric disorders due to vit. B12 deficiency occur commonly without anemia.

**Diagnostic procedures**- Diagnosis of megaloblastic anemia, if it is absent the most reliable procedure is the measurement of the serum vit. B12 level, assays from the antibodies from intrinsic factor.

**Treatment**- Cyanocobalamin or hydroxycobalamin 1000ug per day first two weeks im. Thereafter this dose is repeated twice per week for a year, and then monthly for the remainder of the patient's life. A neurological symptoms and signs may improve during the first 3-6 months, and then at a slower tempo during the year or longer.

## **Childhood and adult spinal muscular atrophies (SMA)**

The group of hereditary motor neuron disorders in which is a progressive degenerative process of motoneurons of the spinal cord or caudal parts of brain stem or both

SMA type	age of onset	survival and prognosis	Inheritance	Defective gene
Infantile SMA (Werdnig – Hoffmann)	Birth-6mos	Deaths by 2yrs old	AR	SMN, NAIP genes *

Intermediate SMA	Before 18mos	No walking, survive to childhood	AR	SMN , NAIP genes
Juvenile(Kugelberg-Welander)	After 18mos	Survive to adulthood	AR	SMN, NAIP genes
Adult onset SMA	After 20 years	Slow progression	AR/AD	Unknown
X-linked Bulbospinal SMA (Kennedy's Disease)	Only males After age 30 yrs	Slow progression	X linked recessive,	Abnormal CAG expansion in gene encoding androgen receptor protein

\*SMN and NAIP genes are located at chr.5; More than 98% SMA I,II have deletion exons 7and 8 of SMN genes

### Clinical features of SMA

**Infantile SMA -Type I (Werdnig –Hoffmann)-It is apparent even before birth-decreased fetal movements late in the third trimester. Babies are weak and floppy, limb weakness is severe, generalized, stretch reflexes are absent, bulbar muscle weakness makes feeding laborious, intercostal muscles are also severe weakened and lead to respiratory insufficiency.**

**Intermediate SMA –Type II- the distribution pattern and progression of weakness are similar to that SMA I but the clinical course is slower.**

**Juvenile-Type III (Kugelberg-Welander). and adult onset SMA- Slowly progressive limb-girdle wasting and weakness , more severe in girdle muscles. Reflexes reduced or absent. Fasciculation are visible than in other types.**

**In adult onset sometimes the weakness is generalized or distal. The clinical course of adult onset SMA is slower than juvenile SMA. EMG is necessary to differentiate the two conditions Usually patients with SMA type III require wheelchair by time they reach their mid-30s yrs.**

**Bulbospinal SMA (Kennedy's Disease)- Slowly progressive limb-girdle muscle weakness, moderate bulbar dysfunction, prominent fasciculations at the limb, tongue, facial muscles; endocrine abnormalities.**

## **OTHER ANATOMICAL SYNDROMES OF SPINAL CORD**

### **1. Central cord syndrome**

**The central cord syndrome affects motor function in the upper limbs more than in the lower ones. The paresis is often denser distally in extremities than proximally. A good method for remembering this syndrome is the MUD (motor>sensory>upper>lower and distal > proximally)**

**Anatomically, this syndrome is explained by somatotopic organization of the long tracts of sc.**

### **2. Brown-Sequard syndrome**

**a. Hemicord syndrome**

**b. ipsilateral spastic paresis and loss of deep sensation below lesions with contralateral superficial sensory loss (dissociated)**

- c. possibly radicular pain and flaccid paresis ipsilaterally at the level of lesion

### **3. Conus medullaris syndrome (in cord from S3 down)**

- a. Lesions at the level of the first lumbar vertebra
- b. Saddle anesthesia
- c. Bladder and rectum paralysis (bladder overflow-incontinence, faecal incontinence)
- d. Anal and bulbocavernosus reflexes are absent

### **4. Epiconus syndrome**

- a. Spinal cord transection at the spinal level of thoracolumbar junction
- b. Paresis or paralysis of hip extension, knee flexion and or of feet and toes
- c. Absence of ankle jerks
- d. Disorder of sensation from L4 down
- e. Paralysis of sphincters

### **5. Cauda equina syndrome**

- a. Multiple root lesions below level of body of second lumbar vertebra
- b. Saddle anesthesia
- c. Bladder and bowel paralysis (in complete cauda equina syndrome)
- d. Segmental paresis below knees, possibly knee flexors and buttock muscles
- e. Segmental sensory disturbance in legs below knees and in feet
- f. Absence of ankle jerks